ISSN 2218-5836 (online)

World Journal of *Orthopedics*

World J Orthop 2016 July 18; 7(7): 406-462

Published by Baishideng Publishing Group Inc

WJO World Journal of Orthopedics

A peer-reviewed, online, open-access journal of orthopedics

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2015-2018

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WJO World Journal of Orthopedics

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Monthly Volume 7 Number 7 July 18, 2016

NAME OF JOURNAL
World Journal of Orthopedics

ISSN
ISSN 2218-5836 (online)

LAUNCH DATE
November 18, 2010

FREQUENCY
Monthly

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PUBLISHER
Baishideng Publishing Group Inc
8226 Regency Drive,
Pleasanton, CA 94588, USA
Telephone: +1-925-223-8242
Fax: +1-925-223-8243
E-mail: bpgoffice@wjgnet.com
Help Desk: http://www.wjgnet.com/esps/helpdesk.aspx
http://www.wjgnet.com

PUBLICATION DATE
July 18, 2016

World Journal of Orthopedics

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Help Desk: http://www.wjgnet.com/esps/helpdesk.aspx
DOI: 10.5312/wjo.v7.i7.406

World J Orthop 2016 July 18; 7(7): 406-417
ISSN 2218-5836 (online)

REVIEW

Prenatal diagnosis and assessment of congenital spinal anomalies: Review for prenatal counseling

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Author contributions: Upasani VV, Ketwaroo PD and Glotzbecker MP contributed to patient chart review, manuscript preparation, manuscript editing; Estroff JA, Warf BC and Emans JB contributed to manuscript preparation, manuscript editing.

Conflict-of-interest statement: None of the authors on this manuscript have any direct or indirect conflicts of interest or financial disclosures related to the contents of this article.

Manuscript source: Invited manuscript

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Received: February 3, 2016
Peer-review started: February 14, 2016
First decision: March 21, 2016
Revised: May 3, 2016
Accepted: May 31, 2016
Article in press: June 2, 2016
Published online: July 18, 2016

Abstract

The last two decades have seen continuous advances in prenatal ultrasonography and *in utero* magnetic resonance imaging. These technologies have increasingly enabled the identification of various spinal pathologies during early stages of gestation. The purpose of this paper is to review the range of fetal spine anomalies and their management, with the goal of improving the clinician's ability to counsel expectant parents prenatally.

Key words: Prenatal ultrasound; *In utero* magnetic resonance imaging; Prenatal counseling; Congenital spinal anomalies

Core tip: Advances in prenatal ultrasonography and *in utero* magnetic resonance imaging have given clinicians powerful tools to identify spinal pathologies during early stages of gestation. Prenatal counseling requires a "team" of appropriate specialists requiring coordination and cross-communication from multiple surgical and medical disciplines. The orthopedic clinician should critically assess the accuracy of the diagnosis, the likely natural history of the deformity, what treatments may be required, and what the impact of the anomaly will be on the child's growth and eventual adult function.

Upasani VV, Ketwaroo PD, Estroff JA, Warf BC, Emans JB, Glotzbecker MP. Prenatal diagnosis and assessment of congenital spinal anomalies: Review for prenatal counseling. *World J Orthop* 2016; 7(7): 406-417 Available from: URL: http://www.wjgnet.com/2218-5836/full/v7/i7/406.htm DOI: http://dx.doi.org/10.5312/wjo.v7.i7.406

INTRODUCTION

The last two decades have seen continuous advances in prenatal ultrasonography and *in utero* magnetic resonance imaging (MRI). These technologies have increasingly enabled the identification of various spinal pathologies during early stages of gestation. The purpose of this paper is to review the range of fetal spine anomalies and their management, with the goal of improving the clinician's ability to counsel expectant parents prenatally.

PRENATAL COUNSELING

The interdependent roles of radiologist and counseling clinicians

The availability of detailed information about skeletal and visceral anomalies allows more accurate fetal diagnosis but also renders the job of the counseling clinicians more complex. Because so many skeletal anomalies are associated with either other anomalies or genetic syndromes, prenatal counseling requires a "team" of appropriate specialists. Armed with prenatal imaging, counseling clinicians are obligated not just to give the family their best estimate of a diagnosis, but also to put the fetus's condition into perspective and attempt to provide some over-arching outlook for the family. This may require coordination and cross-communication among specialists from multiple surgical and medical disciplines. Faced with a fetal spine anomaly, the clinician should critically assess the accuracy of the diagnosis, the likely natural history of the deformity, what treatments are likely, and what the impact of the anomaly will be on the child's growth and eventual adult functioning. This estimate will be easy for an isolated hemivertebra but becomes more complex for multiple vertebral anomalies, spinal dysraphism, myelodysplasia or segmental spinal dysgenesis. Experience with the accuracy of previous prenatal diagnoses makes a more accurate prognosis possible. Co-consultation with other medical or surgical specialists is critical when more than one organ system is involved.

IMAGING PRINCIPLES

Prenatal 2-D and 3-D ultrasound for spinal anomaly detection

While some anomalies in the spina bifida spectrum result in open neural tube defects and therefore may be initially detected with elevated maternal serum alpha fetoprotein, skin covered (closed) spinal dysraphism and other spinal anomalies such as diastematomyelia, vertebral segmentation anomalies, sacral agenesis, spinal dysgenesis, spondylothoracic or spondylocostal dysplasia, or some skeletal dysplasias may be detected only by prenatal imaging. Routine fetal sonography has not been mandated in the United States; however, most pregnant women have at least one detailed anatomical evaluation usually occurring between 18-24 wk of gestation. More recently, routine first trimester ultrasound and maternal serum screening has been offered to patients between 11 and 14 wk of gestation with the goal of measuring the fetal nuchal translucency thickness, nasal bone length and ductus venosus waveform. Imaging at this early time point in the hands of an experienced practitioner may allow for detection of fetal structural anomalies, such as neural tube defects[1]. One metric that has been used in this timeframe is biparietal diameter, which has been shown to measure less than the 5th percentile in 50% of spina bifida patients[1].

Neural element involvement in cases of spinal dysraphism may only be detectable by ultrasound with a high frequency linear transducer[2]. However, more typical obstetrical ultrasound transducers (6-8 MHz) may still detect neural tube defects or abnormal limb positioning, which can serve as an early indicator of neurological dysfunction. Three-dimensional ultrasound of the fetal spine can have great diagnostic benefit, as it allows the operator to manipulate data in any plane after the completion of the exam, particularly if fetal positioning during the optimized 2-D acquisition is challenging[2]. In many cases, however, the presence of the obligatory cranial anomalies associated with an open neural tube defect, described in further detail below, are more readily observed than the spinal defect itself. In general, ultrasonography is able to detect vertebral segmentation, laminar segmentation or rib developmental anomalies as well as gross alignment abnormalities. Congenital fusion or spinal stenosis on the other hand may be better appreciated with MRI.

Prenatal MRI of spinal anomalies

MRI can be used prenatally to better delineate involvement of neural elements associated with osseous spinal anomalies[3]. This can be important for delivery planning, decisions regarding pregnancy interruption, and in some cases, preparation for *in utero* surgery. Patients with closed neural tube defects have a better postnatal prognosis, including superior bladder functionality and lower risk of scoliosis, than their open neural tube defect counterparts, and MRI may more clearly distinguish these two populations[2,4]. While it is not typically utilized for screening purposes, in some cases, fetal MRI may also demonstrate unsuspected spinal anomalies in

fetuses imaged for evaluation of other organ systems.

Fetal MRI provides superior soft tissue contrast. However, fetal movement can make MR imaging in standard planes challenging, and fast pulse sequences such as steady state free precession, half-fourier acquisition single-shot turbo spin echo, fast T1-weighted gradient echo, and echo planar imaging are often utilized. Thin slices of 2-4 mm thickness can minimize the detrimental effects of fetal movement on image quality while maximizing detail of the spinal and cranial anomalies[2,3]. Standard fetal MRI is performed at 1.5 or 3 Tesla field strength. Gadolinium-based contrast agents are not administered to pregnant women, given their ability to cross the placenta and the potential toxicity of unchelated gadolinium to the developing fetus[5]. MRI can be used to approximate the location of the conus, and identify a lipoma or other soft tissue mass within the spine, however can often not detect a syrinx, fatty filum or be used to assess spinal alignment.

Postnatal imaging of spinal anomalies

Additional imaging in the postnatal period can be useful in evaluating the newborn with vertebral anomalies noted on pre-natal imaging. Plain radiographs (AP and lateral of the entire spine including the ribs), should be obtained early, optimally in the first 2 mo, as the bony details of a prenatally-noted anomaly are more evident before further ossification of the vertebra, and other anomalies not seen prenatally can sometimes be detected. All congenital vertebral anomalies have an increased risk (approximately 15%) of intraspinal anomalies. Neonatal spinal ultrasound performed before extensive laminar ossification has occurred (6-12 wk) will show major intraspinal anomalies and tethering[6]. MRI of course better demonstrates intraspinal anomalies[7-9], but generally requires a general anesthetic or heavy sedation which has theoretical deleterious effects in the young child[10-12]. MR can sometimes be accomplished with a "feed and wrap" technique in the infant without general anesthesia or sedation. This technique is used to soothe the child in an attempt to obtain the imaging without requiring sedation. When and whether all vertebral anomalies should be evaluated postnatally with MRI for intraspinal anomalies is debated. Certainly any patient with clinical signs of intraspinal anomaly (extremity deformity or bladder function suggestive of neurologic abnormality) or about to have surgery of the spine should have a whole spine MR. Otherwise the MR may be deferred until an older age when easier to perform or may possibly never be needed if the anomaly is minor without neurologic symptoms. Evaluation of the neonatal spine is typically performed with ultrasound and radiography, though MRI sometimes plays a role as well. While radiographs may readily identify bony abnormalities in the case of vertebral segmentation or formation, ultrasound or MRI better assesses the relationship to neural elements.

CONGENITAL SPINAL ANOMALIES

Chiari malformation

Chiari malformations (CM) describe a heterogeneous group of conditions that are likely embryologically unrelated. Types Ⅰ and Ⅱ CM both involve displacement of the cerebellum with (type Ⅱ) or without (type Ⅰ) displacement of the lower brainstem caudal to the foramen magnum[13]. The resulting compression of these structures can affect neurologic function and impede the normal flow of cerebrospinal fluid. CM type Ⅱ is thought to be due to intrinsic structural anomalies of the brain and spinal cord during fetal development in relation to an accompanying neural tube defect[14,15]. Genetic, nutritional, and environmental factors have all been demonstrated to play a role in the evolution of these defects.

Type Ⅰ CM involves extension of the cerebellar tonsils into the foramen magnum and may be discovered incidentally, often later in childhood or even adulthood and often does not require further treatment. Type Ⅱ CM (previously referred to as Arnold-Chiari malformation) is certainly the more commonly diagnosed prenatal condition of the two. Affected fetuses have a small posterior fossa with hindbrain malformation consisting of displacement of the lower brainstem and cerebellar tonsils and vermis through an enlarged foramen magnum, tectal beaking, medullary kink, enlarged massa intermedia, callosal dysgenesis, and towering cerebellum through an enlarged tentorial hiatus[16,17]. This is almost always found in association with a myelomeningocele. Chiari Ⅲ malformation is serious though fortunately rare and may be diagnosed prenatally after identification of an occipital encephalocele containing herniated cerebellum in addition to the findings of Chiari Ⅱ. Chiari Ⅳ malformation is also extremely rare and demonstrates cerebellar hypoplasia with normal positioning of the cerebellar tonsils.

Prenatal findings: Ultrasonographic identification of CM *in utero* can provide valuable information regarding associated conditions. For example, ventriculomegaly is the most common prenatal finding associated with Type Ⅱ CM and can be associated with neurologic dysfunction, structural abnormalities of the skull, and poor fetal outcome. Biconcave frontal bones resulting in the "lemon sign" on ultrasound (Figure 1) are often seen, though are not specific to an open neural tube defect; 1%-2% of normal fetuses may have this finding, and it may resolve over time even in patients with open neural tube defects[2]. Curved, diminutive appearance of the cerebellum due to effacement of the cisterna magna results in the typical "banana sign" on ultrasound (Figure 2), which has higher specificity for the diagnosis and is 99% sensitive. When combined, the lemon and banana signs have a > 99% sensitivity for CM type Ⅱ prenatally[16]. In addition, there is an almost invariable

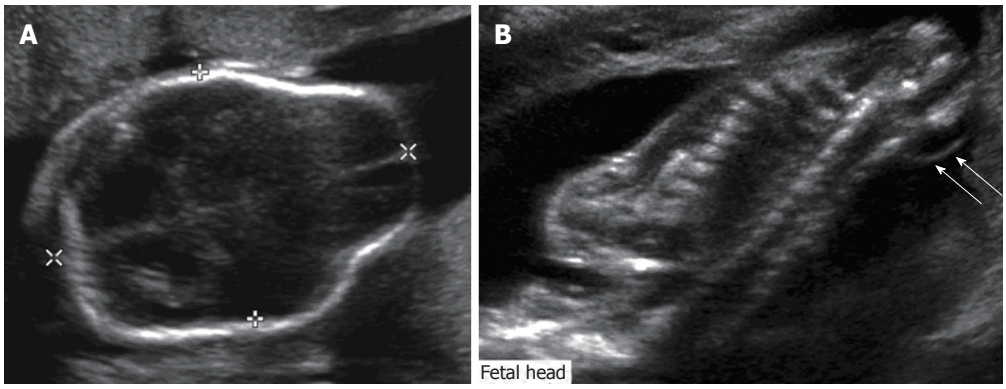

Figure 1 Fetal ultrasound. Grayscale ultrasound images of the fetal head (A) and spine (B) demonstrate ventriculomegaly as well as a lemon-shaped configuration of the fetal head due to bifrontal concavity, typical of Chiari II malformation. There is an associated myelomeningocele (arrows).

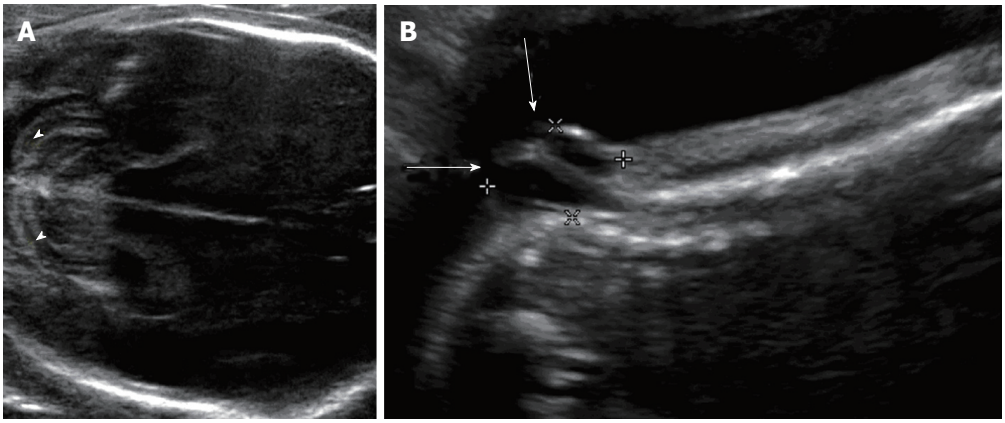

Figure 2 Chiari ultrasound. Grayscale ultrasound images of the 23 wk fetal head (A) and spine (B) demonstrate curved, "banana-shaped" appearance of the fetal cerebellum (arrowheads) typical of Chiari II malformation, as well as associated lower sacral myelomeningocele (arrows).

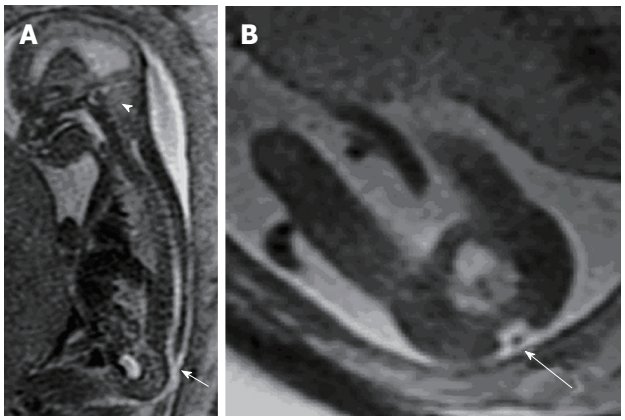

Figure 3 Fetal magnetic resonance imaging. A: Sagittal T2 weighted fetal MR demonstrates a small posterior fossa (arrowhead) with crowding of the cerebellar tonsils and hindbrain; B: Evaluation of the spine demonstrated a lumbosacral myelomeningocele.

association with myelomeningocele. While CM type Ⅱ is typically diagnosed with prenatal ultrasound, fetal MRI (Figure 3) allows for confirmation, further evaluation if ultrasound is technically limited, and evaluation of associated intracranial findings such as callosal agenesis and hydrocephalus[18].

Postnatal evaluation and treatment: For all types of CM, findings are usually first diagnosed with MRI, which demonstrates the classic findings described above. Neurosurgical consultation is warranted in patients with CM to consider decompression of the hindbrain or treatment for hydrocephalus. Intervention for Chiari type Ⅰ in the infant should generally not be entertained without a compelling clinical indication, such as lower cranial nerve dysfunction (*e.g*., apnea or dysphagia) or hydromyelia (syringomyelia). Such is uncommon in young infants. The classic symptoms, suboccipital and posterior neck pain exacerbated by activities involving a valsalva maneuver, cannot be appreciated in babies, and in very young children operative intervention for pain symptoms alone should be approached with caution. This is a dynamic malformation resulting from a relative deficit in the posterior fossa volume, which can change with further growth and development. In other words, for very young children, the malformation can resolve spontaneously over time. Furthermore, infants undergoing posterior fossa decompression for Chiari are more likely to have recurrence from scarring and bone regrowth, requiring repeat surgery in the future.

Patients with Chiari Ⅰ malformation can present with hydromyelia (syringomyelia) even without other

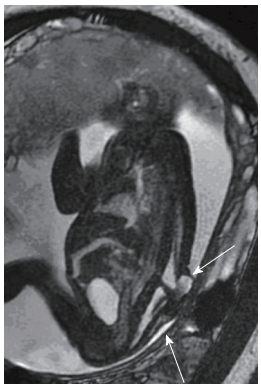

Figure 4 Myelomeningocele magnetic resonance - T2 weighted magnetic resonance image of a 26 wk fetus demonstrates a large thoracolumbar myelomeningocele (arrows) covered by skin.

symptoms. Hydromyelia, or expansion of the central canal of the spinal cord (not unlike the ventriculomegaly of hydrocephalus), results from disturbed CSF flow and pulsatility at the level of the craniocervical junction in these patients. This commonly presents as progressive scoliosis, which is often "atypical" (*e.g.*, levoscoliosis, young age, or male gender), and the diagnosis is made by MRI as part of the scoliosis workup. Chiari decompression (suboccipital craniectomy, C1 laminectomy, and duraplasty) results in resolution of the hydromyelia in most cases. If the scoliosis is not yet severe, the curve progression can be arrested or even reversed without orthopedic intervention. Untreated Chiari I with hydromyelia can lead to indolently progressive neurological deterioration involving both upper and lower motor neuronal dysfunction.

Infants with myelomeningocele and the type Ⅱ Chiari malformation are quite distinct from those with Chiari I. The latter is rarely associated with hydrocephalus, while the former most commonly is. Furthermore, the type Ⅱ malformation involves more than mechanical constriction of the hindbrain structures. This is a diffuse neuronal migrational anomaly affecting the entire central nervous system, with implications for neurocognitive and motor development in addition to the possibility of intrinsic brainstem dysfunction apart from the effects of mechanical compression.

The initial approach to treating symptoms of hindbrain dysfunction (*e.g.*, stridor, dysphagia, apnea) or expanding hydromyelia in these infants is treatment of the accompanying hydrocephalus, which can now often be successfully accomplished endoscopically without shunt implantation[19,20]. Only when this has been satisfactorily accomplished should hindbrain decompression be considered, and at times recovery of lower cranial nerve functions is not realized even with that intervention.

In contrast to the Chiari Ⅰ malformation, infants with Chiari Ⅱ have an enlarged foramen magnum and the torcular and transverse sinuses are typically very low-lying. Hence, the need for suboccipital craniectomy is minimal and associated with higher risk, and the key part of the decompression is comprised of cervical laminectomies and an expansion duraplasty for dorsal decompression of the hindbrain seated in the cervical spinal canal.

Myelomeningocele

Myelomeningocele is a primary neural tube defect associated with failure of neural tube closure at some point along the developing spinal cord that results in a segmental spinal cord malformation with absence of the overlying dura, lamina, soft tissues and skin (Figure 4). There is typically a membrane covering the defect that traps the CSF[18]. It is one of the most common spinal anomalies, with a worldwide incidence of 1 in 1000 births[21]. A genetic predisposition to this condition has been identified in some cases, and supplemental maternal folic acid beginning prior to conception and extending through the first month of pregnancy decreases its incidence. Because of this, the incidence in a given population is variable depending on maternal access to prenatal care and nutrition in addition to genetic factors.

The laminar defect, referred to as spina bifida, can also be seen in isolation or in the context of other skin covered anomalies of the spinal cord (Figure 5). These are often referred to as "spina bifida occulta" (as opposed to the "spina bifida aperta" of myelomeningocele). Spina bifida occulta lesions, such as filum lipoma, lipomyelomeningocele, and terminal myelocystocele are thought to be anomalies of "secondary neurulation" from errors in differentiation and migration of the caudal cell mass that occur after primary neurulation is complete. Thus, the general term, "spina bifida" is quite nonspecific, as is the term "dysraphism", and can lead to confusion.

Prenatal findings: Myelomeningocele is often first suspected when routine screening during pregnancy reveals elevated levels of maternal serum alpha-fetoprotein[22]. Targeted ultrasound also plays an important role in the early detection and delineation of neural tube defects. Myelomeningocele is virtually always accompanied by the Chiari Ⅱ malformation, described above. Patients with myelomeningocele may have an additional spinal cord anomaly, such as a split cord malformation (diastematomyelia), warranting close attention to the spine in both pre- and post-natal populations for pre-surgical planning of myelomeningocele repair[16].

Prenatal treatment: While early post-natal closure of the myelomeningocele to prevent infection is the gold standard, recent advances in fetal surgery have demonstrated that prenatal open or endoscopic repair of the neural tube defect may result in reversal of the Chiari Ⅱ malformation, reduced incidence of

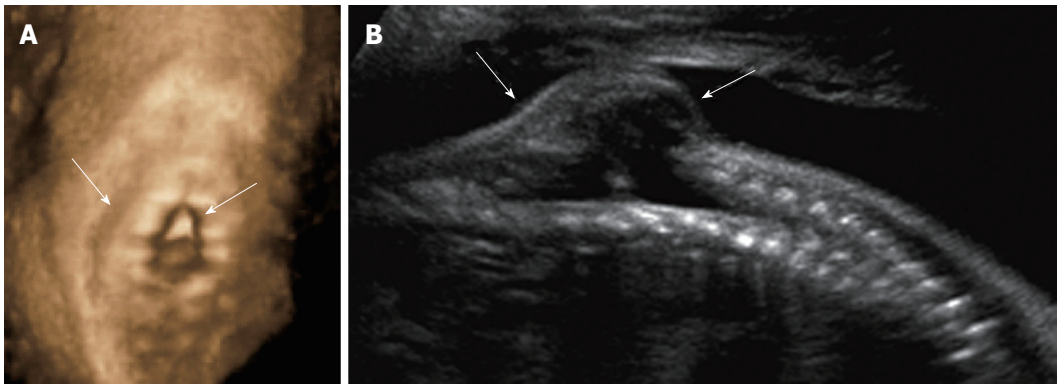

Figure 5 Spinal dysraphism ultrasound. Coronal 3D (A) and sagittal 2D (B) ultrasound images of the 26 wk fetal spine demonstrate dysraphism of the thoracolumbar junction (arrows), in the region of known myelomeningocele.

hydrocephalus, and slightly reduced severity of postnatal neurologic sequelae[23,24]. The relative value of this intervention, however, must be considered within the context of maternal and fetal risk, including the risk of prematurity and its attendant sequelae, fetal demise, and uterine dehiscence. There also appears to be an increased risk of early symptomatic spinal cord tethering and intraspinal dermoid cyst formation. In general the prevalence of orthopedic manifestations of spina bifida, including gait abnormalities, foot deformities (equinovarus, cavovarus, calcaneovalgus, vertical talus), hip dislocations, and abnormalities in the rotational alignment of the lower extremities have not been significantly impacted by prenatal interventions[21].

Postnatal evaluation and treatment: The most common early intervention aside from the initial myelomeningocele closure is the management of hydrocephalus, which is required in around two-thirds of the patients. Placement of a ventricular shunt had been the only effective treatment for these patients since the early 1960's; but, it has recently been demonstrated that the majority (around 75%) can be successfully treated with a minimally invasive endoscopic procedure that combines endoscopic third ventriculostomy with choroid plexus cauterization[20,25].

The primary goals of orthopedic interventions should be to maximize function and mobility while minimizing deformity and pain. Treatment recommendations are based on the functional motor level of the lesion[26,27]. In patients with a thoracic or upper lumbar functional level, the goals of treatment are to maintain spinal balance, a level pelvis, mobile hips and knees, and plantigrade feet. Most of these patients will require surgical stabilization of the spine to treat scoliosis and kyphosis[28-31]. Patients with a mid-lumbar functional level usually have an intact quadriceps muscular function and have the potential to be limited community ambulators with braces and assistive devices. These patients often have weak hip abductors and develop significant genu valgum and knee flexion contractures. Muscle transfers and osteotomies to correct coronal and axial plane abnormalities play an important role in keeping these patients ambulatory[28,32]. Patients with low lumbar or high sacral function typically ambulate using ankle foot orthoses. Hip instability in this group should be aggressively managed to achieve concentric hip reduction and coverage[33]. Foot reconstruction procedures to correct cavovarus deformity are indicated to maintain a supple foot that can be braced[34,35].

Tethered spinal cord

Tethered spinal cord syndrome is a clinical entity in which spinal cord function is compromised by inappropriate attachment of and traction upon the cord, which appears to be associated with compromised tissue perfusion. Tethering of the cord is associated with a variety of etiologies, including scar formation from prior surgery (such as myelomeningocele repair) or an anomaly of secondary neurulation resulting in inappropriate conus traction from a filum lipoma or lipomyelomeningocele[36,37]. Symptoms of spinal cord tethering may also result from a split cord malformation (diastematomyelia) or from a dermal sinus tract that extends intra-dural and is attached to the dorsal spinal cord.

Prenatal findings: *In utero* diagnosis of a tethered spinal cord is usually based on direct visualization of an abnormally caudal position of the conus medullaris, or by identification of a lipoma within the spinal canal. A low-lying conus terminates below the upper endplate of L3 in a child. Though normative values in the fetus have yet to be fully established, prenatal ultrasound has demonstrated that the conus has normally ascended to the L1/L2 level by the 40th postmenstrual week (Figure 6)[38]. A tethering lesion may also result in lack of normal nerve root motion with the CSF pulsations during neonatal imaging. The diagnosis of tethered spinal cord can occasionally be made prenatally with ultrasound and fetal MRI, though postnatal diagnosis is far more common, and may be heralded by the

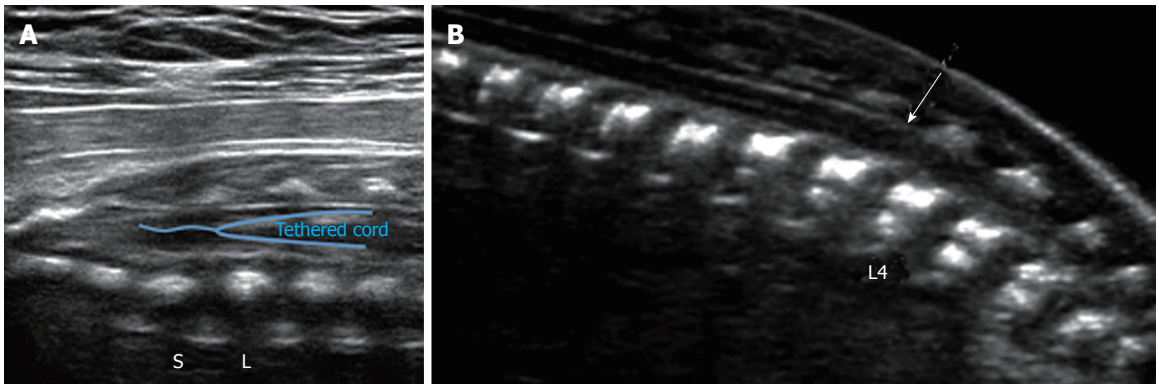

Figure 6 Spinal tether ultrasound. Grayscale ultrasound image (A) of the lumbosacral spine in a 29 wk fetus demonstrates a low-lying conus medullaris, extending below the lumbosacral junction (L/S). No evidence of associated fatty filum terminale was seen in this case; B: Grayscale ultrasound image of the lumbosacral spine in a 21 wk fetus demonstrates a low-lying conus medullaris (arrow), extending below the inferior endplate of L3. No evidence of associated fatty filum terminale was seen in this case.

presence of a sacral dimple or soft tissue mass, or after evaluation for symptoms related to cord tethering, such as bowel and bladder dysfunction, lower extremity orthopedic deformity, or ambulatory disturbance[17].

Postnatal evaluation and treatment: Strictly speaking, "tethered spinal cord" is a clinical rather than radiological diagnosis, and the role of a radiologist is typically to identify possible reasons for clinical symptoms of tethering. Ultrasound can be used to identify tethering up to approximately 6 mo of age, though earlier imaging is technically easier when performed before the posterior spinal elements have ossified. If a spinal tether is not suspected until the child is older, MRI is required for diagnosis.

The clinical manifestations of tethering lesions due to abnormal traction on the conus vary[39]. These include pain, neurogenic bladder dysfunction, progressive foot deformities, scoliosis, and the development of motor or sensory deficits in the lower extremities. It may be identified (as is also the case with hydromyelia) in cases where an MRI is ordered for a rapidly progressive or atypical scoliosis. The characteristic physical exam findings may include the presence of a hairy patch (associated with split cord malformations), sacral dimple, or subcutaneous lipoma in the lumbosacral region[40].

Tethered spinal cord is often released surgically as a prophylactic measure in very young children because of the anticipated risk of progressive neurologic sequelae over time. In older children, symptomatic tethering of the spinal cord may occasionally be present even if the conus is normally positioned[39], and a coexisting filum lipoma is often seen[41]. Clinical indications for tether release include back or leg pain, progressive deterioration of motor or sensory function (such as a change in gait), neurogenic bladder dysfunction, constipation, progressive orthopedic deformities, or spasticity. Rarely, patients with dermal sinus tracts may develop recurrent meningitis[42]. Although residual deficits in urologic or neurologic function may persist, these patients can have an excellent quality of life. Patients with the recent onset of clinical manifestations such as pain, constipation, or urinary incontinence often experience resolution of their symptoms.

Diastematomyelia

Diastematomyelia is a rare congenital anomaly that results in a longitudinal split of the spinal cord[43], usually occurring at the level of the upper lumbar vertebrae. The genesis of this anomaly is thought to occur very early in gestation during gastrulation and prior to neural tube closure. The two hemicords are typically separated by a fibrous, cartilaginous, or osseous septum and reside in two separate dural tubes (type I split cord malformation) (Figure 7). Type II split cord malformations have both hemicords within a single, non-duplicated, dural tube[44]. Each hemicord usually contains a central canal, one dorsal horn and one ventral horn. The two hemicords typically reunite caudally, though two coni medullarae may be seen in diplomyelia, an embryologically distinct entity[45]. Diastematomyelia is typically associated with vertebral segmental anomalies.

Prenatal findings: Though the diagnosis may go unrecognized by prenatal ultrasound in some cases, the presence of a septum between the two hemicords, which appears as an echogenic structure extending from the posterior elements to the posterior aspect of a vertebral body, has high specificity for diastematomyelia[17,18,43]. The spinal canal may also be widened in the coronal plane[41]. The mean gestational age at diagnosis is 21.5 wk, and approximately one third of patients have associated spinal dysraphism[43]. Patients with isolated diastematomyelia have a far better postnatal prognosis than those with associated open neural tube defect, so identifying any of these findings prenatally is critical for family counseling. Prenatal MRI can be used to confirm the diagnosis and look for associated findings,

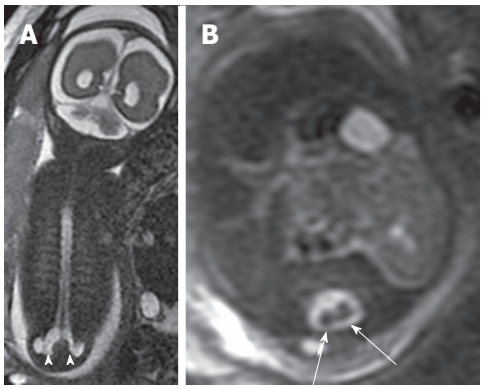

Figure 7 Lumbosacral myelomeningocele. Coronal (A) and axial (B) T2 weighted magnetic resonance images of a 26 wk fetus demonstrate complex spinal anomalies with a lumbosacral myelomeningocele (arrowheads) and diastematomyelia (arrows).

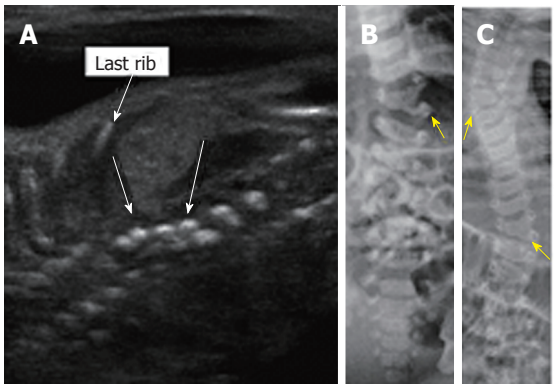

Figure 8 Thoracic hemivertebrae. Parasagittal grayscale ultrasound image (A) of a 20 wk fetus acquired with a high frequency linear transducer demonstrate unpaired echogenic structures at the thoracolumbar junction representing hemivertebrae (arrows). Postnatal frontal radiographs of the same patient's spine at 0 d (B) and 3 years of age (C) demonstrate left thoracolumbar and right midthoracic hemivertebrae.

as some patients have associated meningocele or myelomeningocele, and 75% have tethering lesions[17].

Postnatal evaluation and treatment: The clinical manifestations of diastematomyelia are very similar to those of a tethered cord. Cutaneous lesions, especially a patch of hypertrichosis, are often found over the affected area of the spine and the neurologic symptoms include motor or sensory deficits, scoliosis, incontinence and pain. Postnatal neurologic deficits may present as the child learns to walk, at which time evaluation with MRI is appropriate. MRI is used to confirm or make the diagnosis. In some cases, it may be found during screening MRI for early onset or atypical scoliosis.

If an associated myelomeningocele or secondary tethering lesion such as filum lipoma is not present, the role of prophylactic operative intervention is debated, but is certainly indicated for new or progressive symptoms. When indicated, surgical intervention includes decompression of the neural elements with excision of the interposing tissue and reconstruction of the duplicated dural sacs.

Vertebral formation/segmentation anomalies

Vertebrae typically develop between the sixth to eighth weeks of gestation when two lateral chondrification centers unite to form the primary ossification center of the vertebral body[46]. A disruption in this process can lead to a failure of vertebral formation or segmentation (Figure 8). Although the exact etiology for this phenomenon is unknown, some attribute vertebral formation or segmentation anomalies to disruption of the vascular supply to that portion of the vertebral column[47]. Vertebral anomalies may occur in multiple areas within the spine and are often associated with anomalies in the extremities and other organ systems, including cardiac, genitourinary, gastrointestinal, and pulmonary[48]. They are broadly classified into categories including failure of formation or failure of segmentation.

Prenatal findings: Ultrasound can be used to detect a vertebral anomaly as early as the twelfth week of gestation[49]. In particular, 3-D ultrasound can have a dramatic effect on ease of visualization of these lesions, particularly in skeletal rendering mode[2]. Failures of formation, including hemivertebrae (Figure 9) and wedge vertebrae, may be identified with prenatal ultrasound or less commonly by MRI by recognizing abnormal spinal curvature and unpaired vertebral ossification centers. Failures of segmentation, including block vertebrae, unilateral bar vertebrae, and atlanto-occipital fusion are primarily related to failure of intervertebral disc formation and may present prenatally with fetal kyphoscoliosis[50].

If a spine anomaly is identified, the lower extremities should be evaluated in detail for positioning and functionality. Identification of vertebral anomalies also warrants close inspection of the rest of the fetus, particularly to evaluate for VACTERL association [vertebral anomalies, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies/rib fusions (Figure 10), limb anomalies][16]. Amniocentesis can also provide valuable information regarding any chromosomal abnormalities that may be present.

Postnatal evaluation and treatment: Counseling and management of these patients depends on the gestational age at diagnosis, location and number of vertebral anomalies, and whether there is involvement of multiple organ systems. An isolated hemivertebra typically has a very good prognosis with conservative management and close follow-up. On the other hand, neonates with multiple vertebral anomalies as well as other organ system involvement are typically at higher risk for premature delivery and perinatal morbidity and mortality. Postnatal screening for associated intraspinal, cardiopulmonary, or urologic abnormalities is critical. In patients less than 3 mo of age, a spinal ultrasound may be used to identify a low-lying conus, diplomyelia, diastematomyelia or syringomyelia[51]. Intraspinal

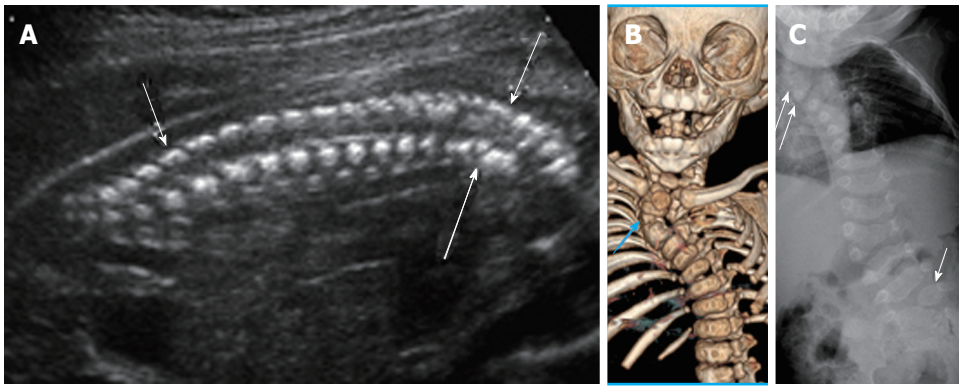

Figure 9 Cervicothoracic Hemivertebrae. Grayscale ultrasound image (A) of a 21 wk fetus demonstrate multiple hemivertebrae (arrows) and cervicothoracic kyphosis. Six months postnatal multidetector unenhanced computed tomography of the thorax with volume rendering (B) as well as frontal radiograph of the spine (C) demonstrate multiple vertebral segmentation anomalies, including right upper thoracic and left lumbosacral hemivertebrae (arrows), resulting in congenital cervicothoracic dextroscoliosis.

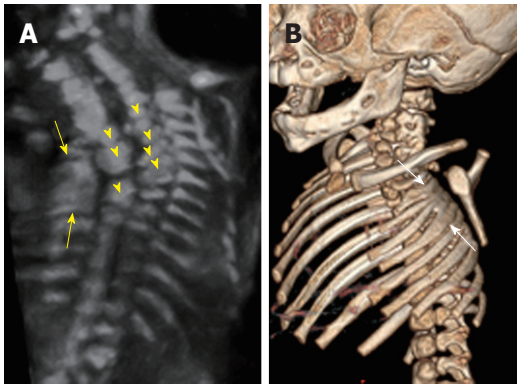

Figure 10 Rib fusions. Grayscale 3D ultrasound image (A) of a 21 wk fetus demonstrate fusion of multiple left sided ribs (arrows) as well as multiple segmentation anomalies (arrowheads) resulting in congenital cervicothoracic kyphoscoliosis. Postnatal multidetector unenhanced computed tomography (B) of the thorax with volume rendering demonstrates bony fusion of multiple left sided ribs (arrows) as well as multiple segmentation anomalies and cervicothoracic dextroscoliosis.

anomalies may be detected in up to 37% of patients with congenital scoliosis[52].

Congenital curves are typically inflexible and unresponsive to bracing[53]. Surgical management is indicated for documented deformity progression, and for anomalies at high risk for progression. Published data have given some insight into the relative progression of commonly encountered malformations[54]; however many of the deformities do not follow the simplified classification scheme, and serial radiographs are required during growth to understand and document progression. Additionally, the location of the vertebral anomaly also plays an important role in determining treatment. For example, small deformities at the lumbosacral or cervicothoracic junction can cause marked imbalance and large compensatory curves. Additionally the treatment of thoracic insufficiency syndrome, often seen in patients with multiple rib fusions or in patients with spondylocostal dysplasia, has recently been described as a surgical indication in patients with congenital vertebral abnormalities to prevent cardiopulmonary comorbidities and allow for symmetric development of the chest and spine[55,56].

Lumbosacral agenesis

Lumbosacral agenesis or caudal regression syndrome is a rare condition with an incidence of 1 in 25000 births. It is technically an anomaly of the caudal cell mass/filum terminale and is strongly associated with maternal hyperglycemia. Most commonly, this is seen in fetuses of mothers with preexisting uncontrolled diabetes (Figure 11) and is associated with vertebral, limb, genitourinary, and anal anomalies[39].

Prenatal findings: Structurally, these patients have varying degrees of coccygeal, sacral, and lumbar agenesis with the abnormality beginning caudally and progressing cranially. The conus medullaris may have a truncated appearance in these patients both by MRI and ultrasound, and the diagnosis is usually first suspected when screening ultrasound demonstrates sacral agenesis. Recent literature suggests that conus morphology can also be evaluated in these patients prenatally with ultrasound utilizing high frequency linear transducers. Evaluation for Currarino triad is warranted in all patients with lumbosacral agenesis, which in addition to sacral anomalies, is comprised of anal atresia and a presacral mass (either teratoma or meningocele)[16].

Postnatal evaluation and treatment: The clinical manifestations of sacral agenesis can vary depending on the extent of lumbosacral root involvement[57]. Most patients have neuropathic bladder and are at increased risk for recurrent urinary tract infections and incontinence. Poor gastrointestinal motility and imperforate anus are also commonly seen. Neurologic involvement includes lower extremity muscle weakness and altered sensation[58]. Equinovarus, lower extremity contractures, kyphoscoliosis and hypoplastic pelvis with

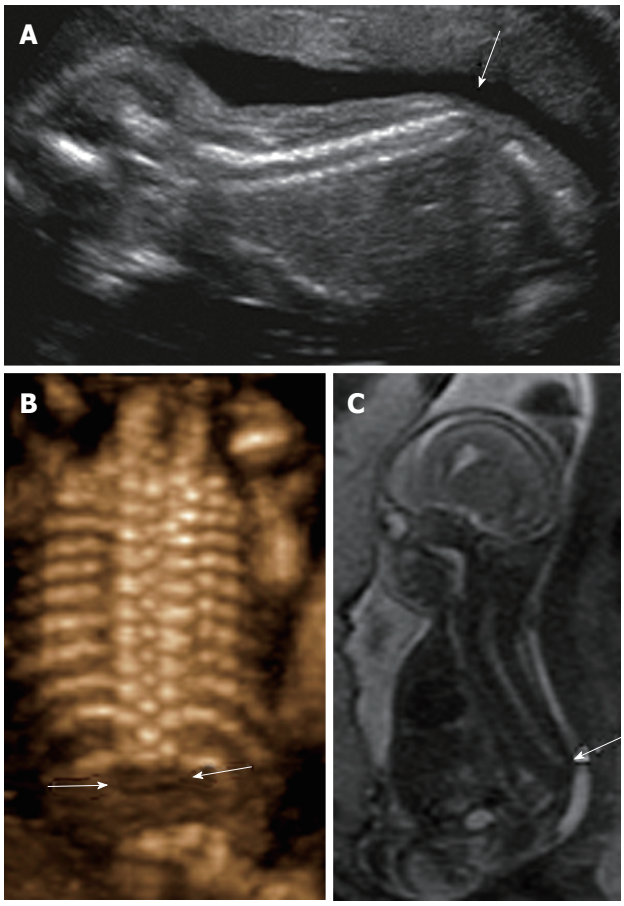

Figure 11 Sacral agenesis. 2D sagittal grayscale ultrasound (A), coronal 3D ultrasound (B), and parasagittal T1 weighted image (C) of a 19 wk fetus demonstrate absence of the fetal spine caudal to T12 (arrows), consistent with sacral agenesis/caudal regression syndrome. The mother was known to have type 1 diabetes, a risk factor for this condition.

hip instability are common orthopedic manifestations of this condition.

CONCLUSION

In summary, advances in prenatal ultrasonography and *in utero* MRI have given clinicians powerful tools to identify spinal pathologies during early stages of gestation. Prenatal counseling requires a "team" of appropriate specialists requiring coordination and cross-communication from multiple surgical and medical disciplines. The orthopedic clinician should critically assess the accuracy of the diagnosis, the likely natural history of the deformity, what treatments may be required, and what the impact of the anomaly will be on the child's growth and eventual adult function.

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P- Reviewer: Singh K, Teli MGA, Zaminy A **S- Editor**: Ji FF
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World Journal of Orthopedics

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DOI: 10.5312/wjo.v7.i7.418

World J Orthop 2016 July 18; 7(7): 418-425
ISSN 2218-5836 (online)

ORIGINAL ARTICLE

Basic Study

Evaluation of bone remodeling in regard to the age of scaphoid non-unions

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Author contributions: All authors made substantive intellectual contributions to this study, whether in the contributions to conception and design (Rein S, Hanisch U, Schaller HE, Zwipp H, Rammelt S, Weindel S), or acquisition of data (Rein S, Schaller HE, Weindel S), or analysis and interpretation of data (Rein S, Hanisch U, Schaller HE, Zwipp H, Rammelt S, Weindel S), or drafting and revising the manuscript critically (Rein S, Hanisch U, Schaller HE, Zwipp H, Rammelt S, Weindel S).

Institutional review board statement: This study was approved by the University of Tübingen Institutional review board (approval number: 367/2007A).

Informed consent statement: All patients gave their informed consent prior to surgery.

Conflict-of-interest statement: This study was financially supported by Zimmer Company, Münsingen, Switzerland. It is declared that the company was not involved in planning the experiment, and did not take part in data collection, analyses, interpretation of data or writing of the manuscript. The authors declare that they have no competing interests and disclose any financial conflicts of interest that may influence interpretation of this study and/or results.

Manuscript source: Invited manuscript

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Received: January 22, 2016
Peer-review started: January 22, 2016
First decision: March 1, 2016
Revised: March 26, 2016
Accepted: May 10, 2016
Article in press: May 11, 2016
Published online: July 18, 2016

Abstract

AIM: To analyse bone remodeling in regard to the age of scaphoid non-unions (SNU) with immunohistochemistry.

METHODS: Thirty-six patients with symptomatic SNU underwent surgery with resection of the pseudarthrosis. The resected material was evaluated histologically after staining with hematoxylin-eosin (HE), tartrate resistant acid phosphatase (TRAP), CD 68, osteocalcin (OC) and osteopontin (OP). Histological examination was performed in a blinded fashion.

RESULTS: The number of multinuclear osteoclasts in the TRAP-staining correlated with the age of the SNU and was significantly higher in younger SNU (P = 0.034; r = 0.75). A higher number of OP-immunoreactive osteoblasts significantly correlated with a higher number of OC-immunoreactive osteoblasts (P = 0.001; r = 0.55). Furthermore, a greater number of OP-immunoreactive osteoblasts correlated significantly with a higher number of OP-immunoreactive multinuclear osteoclasts (P = 0.008; r = 0.43). SNU older than 6 mo showed a significant decrease of the number of fibroblasts (P = 0.04). Smoking and the age of the patients had no influence on bone remodeling in SNU.

CONCLUSION: Multinuclear osteoclasts showed a significant decrease in relation to the age of SNU. However, most of the immunhistochemical findings of bone remodeling do not correlate with the age of the SNU. This indicates a permanent imbalance of bone formation and resorption as indicated by a concurrent increase in both osteoblast and osteoclast numbers. A clear histological differentiation into phases of bone remodeling in SNU is not possible.

Key words: Bone remodeling; Histology; Immunohistological staining; Scaphoid non-union; Scaphoid; Wrist joint

Core tip: The bone remodeling in regard to the age of scaphoid non-union is investigated with immunohistochemistry. Multinuclear osteoclasts showed a significant decrease in relation of the age of scaphoid non-union, but smoking and the age of the patients had no influence on bone remodeling. Most of the immunhistochemical findings of bone remodeling do not correlate with the age of the scaphoid non-unions, which indicates a permanent imbalance of bone formation and resorption.

Rein S, Hanisch U, Schaller HE, Zwipp H, Rammelt S, Weindel S. Evaluation of bone remodeling in regard to the age of scaphoid non-unions. *World J Orthop* 2016; 7(7): 418-425 Available from: URL: http://www.wjgnet.com/2218-5836/full/v7/i7/418.htm DOI: http://dx.doi.org/10.5312/wjo.v7.i7.418

INTRODUCTION

The scaphoid is the most commonly fractured carpal bone[1-3]. Scaphoid non-union (SNU) occurs in approximately 5% to 13% of treated scaphoid fractures and in an unknown number of unrecognized fractures[2,4-6]. The reasons for this high non-union rate are multifactorial including fracture location and vascularity, failure of recognizing the fracture and inadequate initial treatment[7,8]. Additionally, scaphoid fractures heal by intramembranous ossification, which leaves the scaphoid without protective fracture callus against potentially disruptive forces. Progressive osteoarthritis, so called scaphoid non-union advanced collapse, inevitably develops in all cases with untreated SNU over time[9].

Fracture healing consists of a regulatory circuit, which requires the proliferation and differentiation of osteoblasts and osteoclasts for bone regeneration and remodeling, together with formation of new blood vessels for bone vascularisation and a myriad of intercellular interactions and molecular communications to coordinate this complex process[10]. Osteoblasts produce organic components of the extracellular matrix, regulate the mineralisation of the osteoid and therefore are essential for bone formation[11,12]. Osteoclasts are responsible for bone resorption by removing mineralized matrix and breaking up the organic bone. Both osteoclasts and activated macrophages show a high expression of tartrate resistant acid phosphatase (TRAP) and glycoprotein CD 68. TRAP is synthesized as latent proenzyme and activated by proteolytic cleavage and reduction[13,14]. Osteocalcin (OC) is an extracellular matrix protein produced by osteoblasts, which constitutes 2% of the total protein content in bone. It is distributed in cement lines of both cortical and trabecular bone[15,16]. OC is thought to have a role in the early stages of bone healing and is a marker for bone formation[17,18]. Osteopontin (OP) is a non-collagenous extracellular matrix protein and is biosynthesized by osteoblasts, osteoclasts, osteocytes, activated fibroblasts, hypertrophic chondrocytes and cemented lines[16]. It is a multifunctional protein that is involved in several aspects of bone turnover and remodeling as well as fracture healing[16,19].

A recent study found significant less bone remodeling in SNU older than a mean age of 45 mo[20]. However, conventional histological investigation is not sufficient to analyze bone remodeling, because staining of the tissue is unspecific. Immunohistochemistry (IHC) using specific markers of bone resorption and bone formation is helpful to shed further light on the process of bone remodeling in SNU. It is hypothesized that there would be differences in numbers of immunohistochemically stained cells that would correlate with the age of the SNU. This difference in staining may lead to the identification of more bone formation (increase in osteoblasts) or bone resorption (increase in number of osteoclasts) over time and may generate more information about the development of SNU. Therefore the aim of this study was to evaluate bone remodeling of SNU with immunohistochemical markers in regard to the age of the fracture.

MATERIALS AND METHODS

Ethics

The study was conducted in accordance with the Helsinki Declaration. The local ethics committee review

board approved the study (367/2007A).

Patients

Thirty six male patients with a mean age of 26 (SD 12; range: 12-56) years at the time of injury were included in this study. Sixteen right and 20 left wrists were injured. The mean time between injury and surgery for non-union was 22 (SD 27; range: 4-144) mo. Six SNU were localised in the proximal third, 27 in the middle third, and one in the distal third of the scaphoid, respectively. In two patients, exact localisation of SNU was not defined. No additional surgery was performed during the follow-up period in 25 cases. However, one patient received a vascularised bone graft from the distal radius, one patient a four corner fusion, one patient a denervation of the wrist, and four patients another kind of wrist surgery in the postoperative follow-up. The data of the longer postoperative period in four patients were not available. Seventeen patients were non-smokers, 16 patients were smokers, but in three patients it was unclear, whether they are smokers or non-smokers.

Only patients, who stated a defined date of trauma having a symptomatic SNU, were included in this study. Exclusion criteria were unclear date of trauma, prior surgical treatment or associated adjacent injuries of the wrist as well as relevant underlying clinical diseases as diabetes mellitus or vascular disorders. Delayed fracture healing was defined between 4 to 6 mo. If no stable ossification was seen after 6 mo, the term non-union was used[21].

Histological examination

During surgery the SNU was resected completely, whereas resection sides showed healthy bone verified by macroscopic bleeding. Autologous cancellous bone was interposed in the former SNU gap and compression osteosynthesis using a Herbert screw was performed[22]. Specimens were immediately fixed in 4% neutral buffered (pH = 7.4) formaldehyde solution for 24 h at 4 ℃, decalcified with diaminoethanetetraacetic acid and embedded in paraffin.

Sections of 2 μm were cut on a Leica rotation microtome (RM2055, Wetzlar, Germany) and mounted on silane-coated slides for conventional staining, enzyme- and IHC. HE-staining was performed in all specimens for morphological evaluation. Subsequently, the tissue sections were stained with TRAP, CD 68 (working dilution: 1:150, monoclonal, clone: KP-1, mouse anti-human, Dako, Glostrup, Denmark), OC (working dilution: 1:250, monoclonal, clone: OCG-3, mouse anti-human, Zytotec, Berlin, Germany) and OP (working dilution: 1:300, polyclonal, rabbit antisera, Chemicon, Temecula, Canada). Blocks and slides were stored at room temperature.

The mounted sections were dehydrated beginning with xylol in decreasing concentrations. Sections were then rehydrated with distilled water and pretreated according to the individual instructions from the suppliers of the used primary antibodies.

No special pretreatment was necessary for CD 68 and OC. For OP, specimens were treated with trypsine (pH = 6.0) for 30 min. After washing in phosphate-buffered saline solution (PBS, pH = 7.4), endogenous peroxidase activity was blocked in all sections with 1% hydrogen peroxide for 5 min. Nonspecific electrostatic protein charging was blocked with blocking reagent (Dako, Glostrup, Denmark) for 10 min at room temperature. Sections were incubated with respective normal sera (Linaris, Wertheim, Germany) for an hour at room temperature and then incubated overnight at 37 ℃ with primary antibodies. Biotinylated secondary antibodies were added for 30 min at 37 ℃, followed by an avidin-biotin-enzyme complex for 30 min at 37 ℃ (Vectastain ABC-HRP kit, Linaris, PK-4000, Wertheim-Bettingen, Germany) at room temperature. The peroxidase activity was visualized with 3'-3'-diaminobenzidine. Then counterstaining with hematoxylin was performed. Sections were washed thoroughly three times in PBS for 5 min after each step. Finally, sections were dehydrated and covered with Entellan (Merck, Darmstadt, Germany). Control procedures, *i.e.,* identical staining without adding primary antibodies, were performed in parallel. Then counterstaining with hematoxylin was performed.

Histopathological examination of the stained tissue sections was performed using an Olympus BHS light microscope in the transmitted mode at final magnifications of 40 ×, 100 ×, 200 × and 400 ×. One section in each staining per subject was analysed. Total cell counts were counted at an original magnification of 100 × in 10 subsequent adjacent visual fields, representing the whole width of the non-union. Only fibroblasts were counted in 5 subsequent visual fields, because the volume of fibroblast tissue was not big enough for 10 visual fields in most cases. All specimens were blinded for cell counts.

Histopathological analysis was centered on osteopathological criteria including determination of chondrocytes and extracellular matrix (ECM) at the non-union gap, osteoblasts, osteoclasts, osteocytes, osteoid and cement lines of the underlying bone, mesenchymal cells and ECM in resorptive bone cysts as well as cysts containing fibrous or fibrocartilage tissue and typical hyaline cartilage in the OC and OP staining[23].

Morphological analysis and cell counting

Morphological analysis was first performed with the HE-staining. With the help of the following criteria the different cell types were identified: Osteoblasts, which are mononuclear cells, were counted if they lined up the external surface of bone trabeculae and the surface of Haversian canals (Figure 1)[23,24]. Osteocytes were counted if they were embedded into the mineralized bone matrix (Figure 1). Cells with two to fifteen nuclei in small resorptive excavations (Howship's lacunae) on the bone surface were counted as multinuclear osteoclasts (Figure 2).

Mononuclear osteoclast precursors could only be

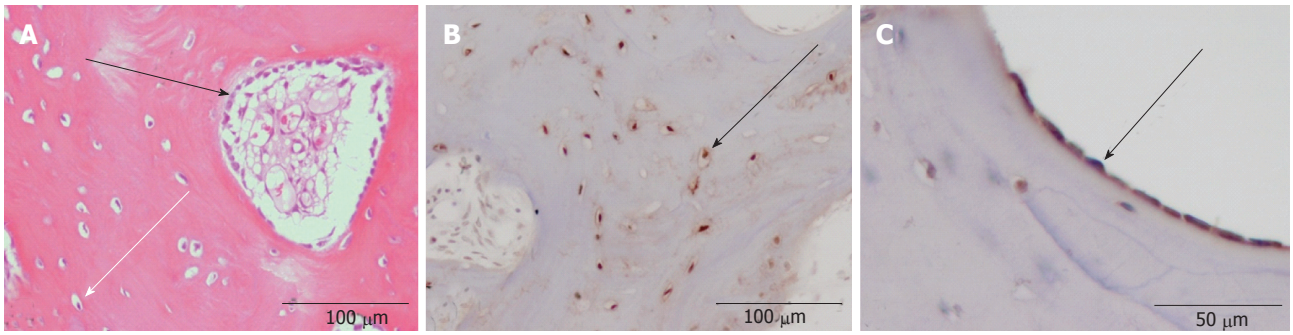

Figure 1 Morphological analysis. A 7-mo-old SNU is shown (A-C). A: HE-staining shows osteoblasts (black arrow) and osteocytes (white arrow) in the underlying sclerosed bone; B: OC-staining shows osteocytes (arrow) in detail; C: OP-staining shows the osteoblasts (arrow) lining a Haversian vessel in detail. Original magnification 200 × (A, B), 400 × (C). SNU: Scaphoid non-unions; OP: Osteopontin; OC: Osteocalcin.

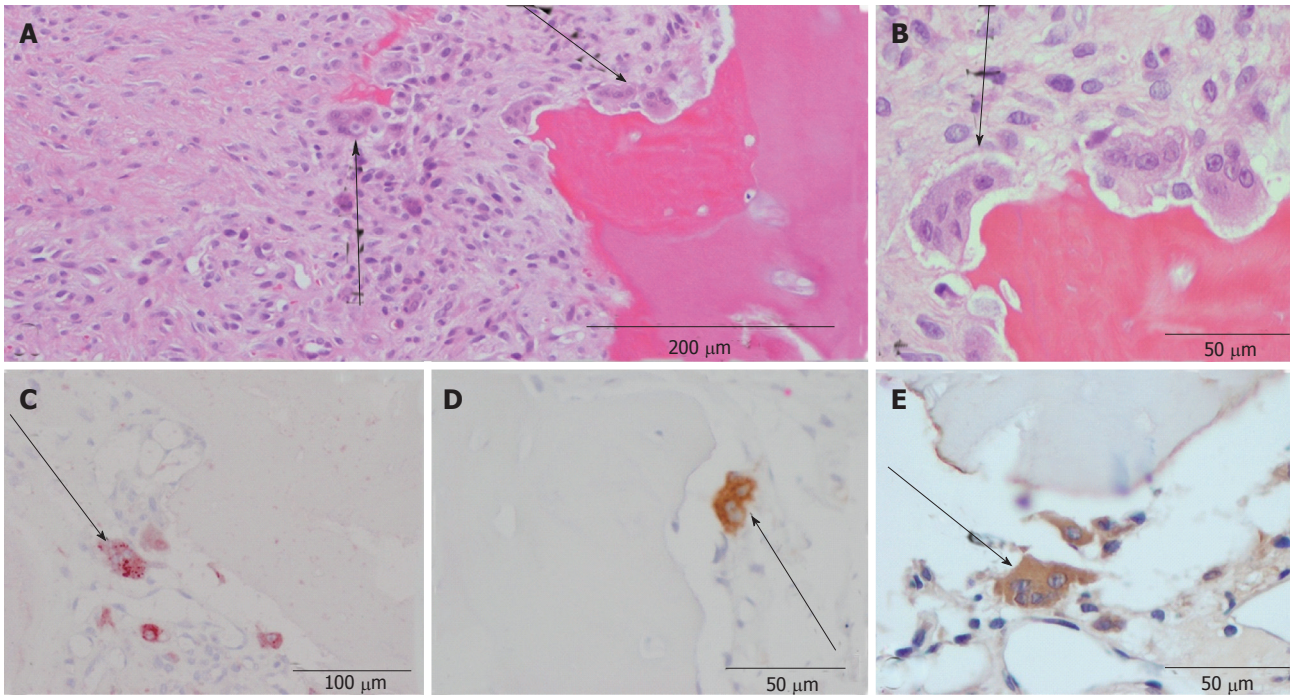

Figure 2 Osteoclasts. Examples for multinuclear osteoclasts as seen in the HE-staining (A and B), the TRAP (C), CD 68 (D) and OP (E) staining. (A-D) show osteoclasts (arrows) in a 10-mo-old SNU, whereas (E) shows an osteoclast in a 7-mo-old SNU. Original magnification 100 × (A), 200 × (C), 400 × (B, D, E). SNU: Scaphoid non-unions; OP: Osteopontin; TRAP: Tartrate resistant acid phosphatase.

identified in the TRAP and CD 68 staining and were counted as mononuclear positive-stained cells in these two stainings. Fibroblasts were counted as cells located in the non-union gap in the HE-staining (Figure 3).

Multinuclear osteoclasts were counted in the TRAP staining, with CD 68 and OP-IHC. Osteoblasts and osteocytes were counted with OP- and OC-IHC, respectively.

Data analysis

Statistical analysis was performed with a two sided *t* test in order to analyse the occurrence of OC- and OP-positive cells in the different parts of the SNU with a level of significance of $P \leqslant 0.05$.

The two-sided Pearson correlation analysis was used to investigate the linear relationship with regard to age of SNU and patients, and counted cell numbers. Correlation analysis was performed with Spearman's rho coefficient with a significance level of $P \leqslant 0.05$. The influence of smoking has been investigated with the Kruskal-Wallis test followed by the Mann-Whitney test with a level of significance of $P \leqslant 0.05$. Statistical analysis was performed with the computer program SPSS (Version 11.5, Chicago, United States).

RESULTS

Immunohistochemical findings

Table 1 gives an overview over the markers that could reliably and reproducibly be detected. Negative procedures without antibodies showed no staining. OP was immunolocalized within chondrocytes and ECM of the non-union, osteoclasts, osteoblasts, osteoid and osteocytes of the underlying bone as well as the

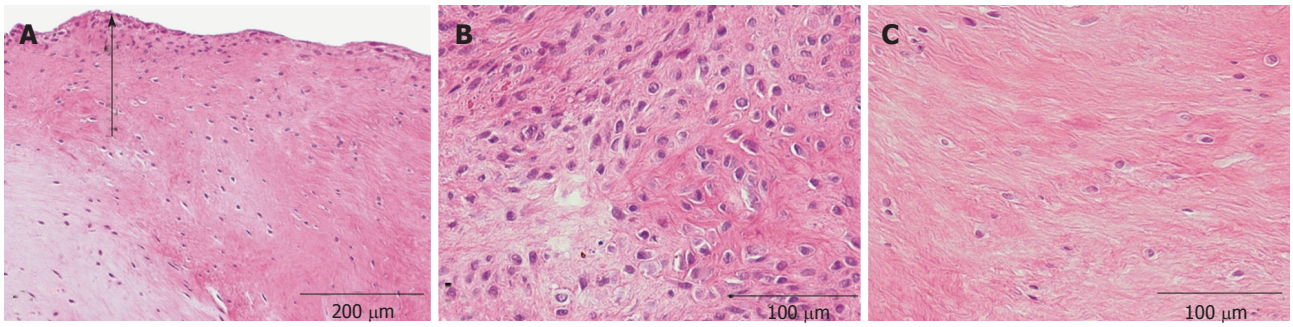

Figure 3 Non-union tissue in the hematoxylin-eosin staining. A: Non-union tissue with fibrocartilage covered by a layer of synovium-like lining cells (arrow) of a 60-mo-old SNU; B: Fibrous tissue rich in fibroblasts of a 10-mo-old SNU. Note the contrast in cell number as compared to the 60-mo-old SNU in A; C: Fibrous tissue with few fibroblasts, same patient as in A. Note the contrast in cell number as compared to the 60-mo-old SNU. Original magnification 100 × (A), 200 × (B, C). SNU: Scaphoid non-unions.

Table 1 Summary of the histological features with the range of time of their appearance

Histological feature		Immunohistochemical findings												
		Osteocalcin							OP					
		Yes (*n*)	Range of time (mo)		No (*n*)	Range of time (mo)		Not assessable (*n*)	Yes (*n*)	Range of time (mo)		No (*n*)	Range of time (mo)		Not assessable (*n*)
Non-union	Chondrocytes	1	12	12	35	4	144	-	25	4	144	11	6	60	-
	ECM	0	-	-	36	4	144	-	34	4	144	2	15	24	-
Underlying bone	Osteoblasts	31	6	144	4	4	22	1	34	4	144	2	9	15	-
	Osteoclasts	2	8	29	33	4	144	1	33	4	85	2	9	15	1
	Osteocytes	32	6	144	4	4	15	-	34	4	144	2	9	15	-
	Osteoid	13	7	28	23	4	144	-	32[1]	4	85	4[1]	14	144	-
	Cement lines	33	4	144	3	7	15	-	8	7	144	28	4	85	-
Resorptive bone cysts	Mesenchymal Cells	-	-	-	15	4	60	21	11	4	24	5	9	60	20
	ECM	-	-	-	15	4	60	21	15	4	60	1	15	15	20
Fibrous bone cysts	Mesenchymal cells	1	15	15	12	7	54	23	10	7	54	7	4	28	19
	ECM	1	15	15	12	7	54	23	12	7	54	5	4	24	19
Hyaline cartilage	Typical	33	4	144	2	9	12	1	34	4	144	1	15	15	1

The underlying bone's osteoid showed significantly more often OP-immunoreactivity in younger (18.5 ± 17.9 mo) than in older SNU (50.5 ± 62.7 mo) ([1]P = 0.02). ECM: Extracellular matrix; OP: Osteopontin; SNU: Scaphoid non-unions.

hyaline cartilage. Cement lines of newly formed lamellar bone only stained positively for OP in 8 out of 36 cases (Table 1). OC showed immunoreactivity in cement lines, osteocytes, osteoblasts and hyaline cartilage (Table 1). Resorptive and fibrous bone cysts showed immunoreactivity for OP in most cases but not for OC (Table 1). Enzyme-histochemical staining against TRAP specifically stained osteoclasts and mononuclear precursors indicating bone resorption during the remodeling process (Figure 2C). The macrophage marker CD 68 was detected in mononuclear and multinuclear macrophages or osteoclasts (Figure 2D). Mononuclear macrophages/osteoclast precursors and multinuclear osteoclasts stained positively for CD 68 in 32 out of 36 cases, whereas in only 8 out of 36 cases osteoclasts were stained positive for TRAP.

Osteoid showed immunoreactivity for OP in 32 younger SNU (18.5 SD 17.9 mo) with a range of age between 4 to 85 mo, whereas there was no immunoreactivity for OP in 4 older SNUs (50.5 SD 62.7 mo) with a range of age between 14 to 144 mo. The difference between the two groups was statistical significant (P = 0.02; Table 1).

Cell counting and correlation analysis

Single results of the cell counting are presented in Table 2. The number of multinuclear osteoclasts in the TRAP-staining correlated with the age of the SNU and was significantly higher in younger SNU (P = 0.034; r = 0.75; Figure 4). All other correlations in regard to the age of the SNU showed no significant results.

A higher number of OP-immunoreactive osteoblasts significantly correlated with a higher number of OC-immunoreactive osteoblasts (P = 0.001; r = 0.55; Figure 5). Furthermore, a greater number of OP-immunoreactive osteoblasts correlated significantly with a higher number of OP-immunoreactive multinuclear osteoclasts (P = 0.008; r = 0.43; Figure 6).

Table 2 Results of the cell counting are shown as mean with standard deviation

Single results of the cell counting					
Histological feature	**HE**	**TRAP (*n* = 8)**	**CD 68 (*n* = 32)**	**OP (*n* = 36)**	**OC (*n* = 36)**
Osteoblasts	N/A	N/A	N/A	23.4 ± 12	25.6 ± 19.5
Osteoclasts					
Uninuclear	N/A	9 ± 6.5	4.6 ± 5.1	N/A	N/A
Multinuclear	N/A	2.3 ± 0.8	1.8 ± 1.2	1.1 ± 0.8	N/A
Osteocytes	N/A	N/A	N/A	38.2 ± 11.2	50.8 ± 16.4
Fibroblasts	285 ± 181	N/A	N/A	N/A	N/A

TRAP: Tartrate resistant acid phosphatase; CD 68: Cluster of differentation 68; OC: Osteocalcin; OP: Osteopontin; N/A: Not analysed.

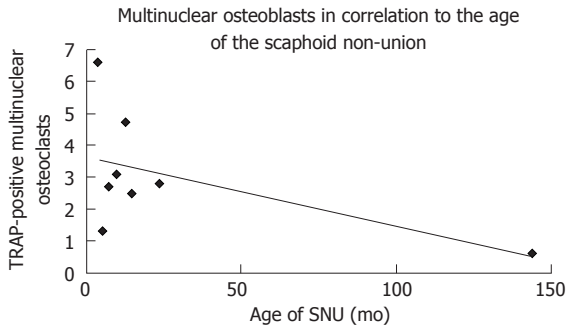

Figure 4 Multinuclear osteoclasts in correlation to the age of the scaphoid non-unions. The number of multinuclear osteoclasts in the TRAP-staining was significantly higher in younger SNU (P = 0.034; r = 0.75). SNU: Scaphoid non-unions; TRAP: Tartrate resistant acid phosphatase.

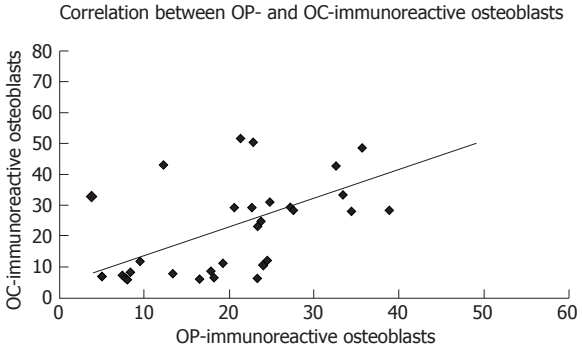

Figure 5 Correlation between osteoblasts in the osteopontin- and osteocalcin- immunohistochemistry. A greater number of OC-immunoreactive osteoblasts correlated significantly with a greater number of OP-immunoreactive osteoblasts (P = 0.001; r = 0.55). OC: Osteocalcin; OP: Osteopontin.

A mean of 285 (SD 181) fibroblasts were counted in the 36 investigated SNU in the HE staining. A mean of 457 (SD 175) fibroblasts were counted in SNU (n = 4) up to 6 mo old. In contrast, a mean of 264 (SD 173) fibroblasts were measured in SNU (n = 32) older than 6 mo. This was a significant decrease of fibroblasts in SNU, which are older than 6 mo (P = 0.04). However, no significant correlations have been found between the age of the patients and all investigated cell types. Furthermore, no significant differences have been observed between smokers and non-smokers for all investigated cell types.

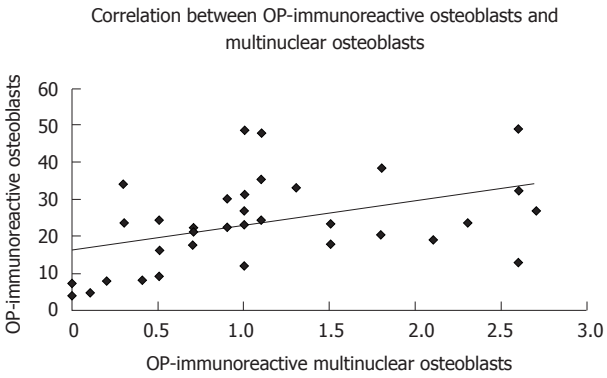

Figure 6 Correlation between osteopontin-immunoreactive osteoblasts and multinuclear osteoclasts. A greater number of OP-immunoreactive osteoblasts correlated significantly with a greater number of OP-immunoreactive multinuclear osteoclasts (P = 0.008; r = 0.43). OP: Osteopontin.

DISCUSSION

A recent study has shown that significant less bone remodeling takes place in older SNU with a mean age of 45 mo compared to a mean age of 18 mo[20]. However, these results were based on conventional HE-staining. Several bone-specific extracellular matrix proteins may be used to assess bone remodeling[16]. OC is reportedly the most specific noncollagenous bone matrix protein, being expressed by osteoblasts and osteocytes[25]. In the present study, we demonstrate specific staining of osteoblasts, osteocytes, cement lines, hyaline cartilage and in some cases osteoid (n = 13). OP is reportedly expressed by osteocytes, osteoblasts, and their precursors, osteoclasts, hypertrophic chondrocytes, and cement lines[15,16]. We have seen specific staining of osteoblasts, osteoclasts, osteocytes, osteoid, chondrocytes, ECM of the non-union gap, and hyaline cartilage. OP interacts with osteoclasts, implicating it as a potentially important marker of bone resorption[26].

A greater number of OC-immunoreactive osteoblasts correlated significantly with a greater number of OP-immunoreactive osteoblasts. However, correlation analysis between the two markers showed no time-dependent significant differences. Furthermore, a higher number of OP-immunoreactive osteoblasts correlated significantly with a higher number of OP-immunoreactive multinuclear osteoclasts, indicating a

higher bone remodeling in younger SNU. These findings confirm the theory that bone remodeling is a balance between bone formation and bone resorption in which osteoblasts exhibit two opposite phenotypes. There is the osteogenic phenotype, which secretes bone matrix at the bone resorption site, and the osteoclastogenic phenotype, which supports osteoclast differentiation in the old bone area[11]. The close interplay between osteoblasts and osteoclasts during bone repair is well established[10-12,17,23,24].

A recent study has shown that cell viability and mineralization-positive colony forming units were significantly reduced in osteoblasts retrieved from non-union sites. This study identified a set of significantly down-regulated factors in those "non-union osteoblasts" that are involved in the regulation of osteoblast proliferation and differentiation[27]. This indicates that activity of osteoblasts in non-unions is altered, which could explain the lack of time-dependent changes in the OC- and OP-staining. However, another study could demonstrate, that OC-positive osteoblasts, which were taken from SNU, possessed osteogenic capability and could be stimulated by recombinant human bone morphogenetic protein-2 *in vitro*, resulting in significant increase in osteoblast differentiation and bone production[28].

The number of osteoclasts decreased significantly in older SNU, which could be shown in the TRAP-staining, but not in the CD 68 IHC. This could be explained by the fact that CD 68 is not a specific osteoclast marker but rather a marker for several cells of the monocyte/macrophage lineage.

It is known, that nicotine has a dose dependent negative effect on bone healing, resulting in ischemia, diminished osteoblast function and decreased expression of bone morphogenetic protein[29,30]. However, in this study nicotine abuse had no measurable influence on bone remodeling in SNU.

The high account of fibroblasts reflects a cell rich fibrous tissue in the non-union gap. We have seen a significant decrease of the count of fibroblasts in SNU older than 6 mo. Our explanation is that the instability in the non-union gap induces or provokes an activation of fibroblasts. For that reason, further research on this topic could be the investigation of proliferation with specific immunohistochemical markers, *e.g.,* Ki 67.

A greater number of OP-immunoreactive osteoblasts significantly correlated with a greater number of OC-immunoreactive osteoblasts and OP-immunoreactive multinuclear osteoclasts. Multinuclear osteoclasts show a significant decrease in older SNU. Fibroblasts showed a significant decrease in SNU, which are older than 6 mo. These results indicate a decreased bone remodeling in older SNU. On the other hand, permanent remodeling indicates mechanical instability and imbalance. Therefore most of the immunohistochemical markers of bone remodeling do not correlate with the age of the SNU. Smoking had no influence on bone remodelling in SNU.

ACKNOWLEDGMENTS

The authors thank Ursula Range for her advice with the statistical analysis, Suzanne Manthey and Doreen Küchler for histological preparation as well as Thomas Albrecht for preparation of the photographs.

COMMENTS

Background

The scaphoid is the most commonly fractured carpal bone, whereas non-union occurs in approximately 5% to 13% of treated scaphoid fractures.

Research frontiers

Conventional histological stainings are insufficient to analyze bone remodeling, because staining of the tissue is unspecific.

Innovations and breakthroughs

Immunohistochemistry using specific markers of bone resorption and bone formation is helpful to shed further light on the process of bone remodeling in scaphoid non-union.

Applications

Multinuclear osteoclasts, as a marker for bone resorption, showed a significant decrease in relation of the age of scaphoid non-union, but smoking and the age of the patients had no influence on bone remodeling. Most of the immunhistochemical findings of bone remodeling do not correlate with the age of the scaphoid non-unions (SNU), which indicates a permanent imbalance of bone formation and resorption.

Peer-review

This is a study on the bone remodeling in regard to the age of SNU with immunohistochemistry. The rationale for the study is appropriate and it is an interesting paper with a valuable contribution.

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P- Reviewer: Luo XH, Maia LP **S- Editor**: Qiu S **L- Editor**: A
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DOI: 10.5312/wjo.v7.i7.426

World J Orthop 2016 July 18; 7(7): 426-433
ISSN 2218-5836 (online)

ORIGINAL ARTICLE

Case Control Study

Computerized tomography based "patient specific blocks" improve postoperative mechanical alignment in primary total knee arthroplasty

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Author contributions: Vaishya R helped in performing the surgery and designing the research; Vijay V helped in data collection and preparation of the manuscript; Birla VP helped in data collection and statistics; Agarwal AK helped in preparation of manuscript.

Institutional review board statement: The study was approved by the IRB of the institution.

Informed consent statement: All the patients included in the study gave an informed consent for inclusion in the study.

Conflict-of-interest statement: No conflicts of interest to declare.

Data sharing statement: The authors are willing to share the data of the present study.

Manuscript source: Invited manuscript

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Received: January 11, 2016
Peer-review started: January 14, 2016
First decision: February 29, 2016
Revised: March 14, 2016
Accepted: May 7, 2016
Article in press: May 9, 2016
Published online: July 18, 2016

Abstract

AIM: To compare the postoperative mechanical alignment achieved after total knee arthroplasty (TKA) using computer tomography (CT) based patient specific blocks (PSB) to conventional instruments (CI).

METHODS: Total 80 knees were included in the study, with 40 knees in both the groups operated using PSB and CI. All the knees were performed by a single surgeon using the same cruciate sacrificing implants. In our study we used CT based PSB to compare with CI. Postoperative mechanical femoro-tibial angle (MFT angle) was measured on long leg x-rays using picture archiving and communication system (PACS). We compared mechanical alignment achieved using PSB and CI in TKA using statistical analysis.

RESULTS: The PSB group (group 1) included 17 females and seven males while in CI group (group 2) there were 15 females and eight males. The mean age of patients in group 1 was 60.5 years and in group 2 it was 60.2 years. The mean postoperative MFT angle measured on long-leg radiographs in group 1 was 178.23° (SD = 2.67°, range: 171.9° to 182.5°) while in group 2, the mean MFT angle was 175.73° (SD = 3.62°, range: 166.0° to 179.8°). There was significant improvement in postoperative mechanical alignment (*P* value = 0.001), in PSB group compared to CI. Number of outliers were also found to be less in group operated with PSB (7 Knee) compared to those operated with CI (17 Knee).

CONCLUSION: PSB improve mechanical alignment after total knee arthroplasty, compared to CI. This may lead to lower rates of revision in the PSB based TKA as compared to the conventional instrumentation.

Key words: Knee; Replacement; Arthroplasty; Patient specific jigs; Conventional jigs

Core tip: Computer tomography (CT) based patient specific blocks (PSB) can help restore the mechanical axis of the patients undergoing primary total knee replacement. In the present study, the PSB group had significantly better post-operative mechanical axis as compared to the conventional instrumentation group. CT based PSB holds promise to help in accurate restoration of the mechanical axis and might decrease the rates of revision after total knee arthroplasty.

Vaishya R, Vijay V, Birla VP, Agarwal AK. Computerized tomography based "patient specific blocks" improve postoperative mechanical alignment in primary total knee arthroplasty. *World J Orthop* 2016; 7(7): 426-433 Available from: URL: http://www.wjgnet.com/2218-5836/full/v7/i7/426.htm DOI: http://dx.doi.org/10.5312/wjo.v7.i7.426

INTRODUCTION

Total knee arthroplasty (TKA) has been a successful surgery providing excellent functional results in a patient with an advanced degenerative joint disease of the knee. It has been accepted that postoperative lower limb alignment is an important parameter for favorable functional outcomes in TKA[1]. Arthroplasty surgeons aim to achieve an average mechanical angle (*i.e.*, 180°) and any variation more than 3° may lead to poor functional results and early implant failure[2,3]. Therefore, postoperative mechanical alignment has been considered an important index for a successful surgery amongst arthroplasty surgeons.

The commonly used conventional instruments (CI) in TKA consist of intramedullary (I/M) femoral alignment guide and extramedullary (E/M) tibial alignment guide. Although these instruments are familiar to most surgeons and easy to use, they have demonstrated a limited degree of accuracy[4]. The CI used as I/M femoral guide is aligned on the basis of anatomical axis of the femur. However, the surgeon intends to have distal femur bone cut perpendicular to the mechanical axis (MA) of the femur. In this technique, the surgeon has to make an assumption about the femoral valgus angle which may lead to error in bone cuts, due to the discrepancy in the assumed and the real femoral valgus angle. Also, there could be errors in rotational alignment that is estimated using the epicondylar axis or the anteroposterior axis as popularized by Whiteside *et al*[5]. The epicondylar axis is hard to determine and can lead to significant inter as well as intra-observer variability[6-8]. The I/M guide is dependent on an exact fit of the rod in the femoral canal, which is not always obtained and on the position of the entrance hole in the femur, which is user dependent and may significantly alter the distal femoral mechanical angle[9-12]. Due to several inherent limitations of CI, there was a need for improvement, and this has prompted efforts to develop a more precise surgical technology for restoring an accurate mechanical axis after TKA.

This initiative led the development of computer-assisted surgery (CAS) techniques. Although, CAS has been shown to improve MA over CI, it is also associated with disadvantages like increased surgical time, expensive instrumentation, steep learning curve, higher number of personnel required and higher infection rates[13,14].

Patient specific blocks (PSB) were introduced as an alternative to CAS and CI, with the goal of improving postoperative alignment, implant positioning and overcoming the shortcomings associated with the CAS and CI. These blocks were manufactured before the surgery after a computerized tomography (CT) analysis (Figure 1). The primary aim of these blocks is to optimize the bone cuts so as to achieve accurate MA of the lower limb. There has been recent interest seen in the literature regarding the use and efficacy of patient specific instruments (PSI) in achieving the optimal MA after TKA. But, until now the published reports comparing PSI and CI in primary TKA vary regarding the actual advantages of PSB[15-19].

We undertook this study with the aim to compare the postoperative mechanical alignment achieved using PSI and CI in TKA to assess the efficacy of CT-based PSB in the achievement of anatomical alignment after primary TKA.

MATERIALS AND METHODS

This study was a prospective, comparative single study conducted over a period of one year. All the patients who came with severe degenerative arthritis of knee (Ahlback's grade 3 or 4) and planned for TKA during the period from November 2013 to October 2014 in one unit were included in this study. An approval from the institutional review board was taken. As the technique of PSB was still new and very few patients were aware of it, all the subjects were first explained and made familiar with the technique of PSB and its difference from CI for TKA. Patient were informed about the expected benefits and advantages of PSB, and also about the extra cost of 400$ per knee involved in using the PSB (cost of CT Scannogram and manufacturing of the blocks). Subjects who were ready to bear the extra cost, willing to undergo the CT scan, and gave consent were operated using PSB, and were named as group 1. Remaining subjects were conducted using CI and designated as group 2.

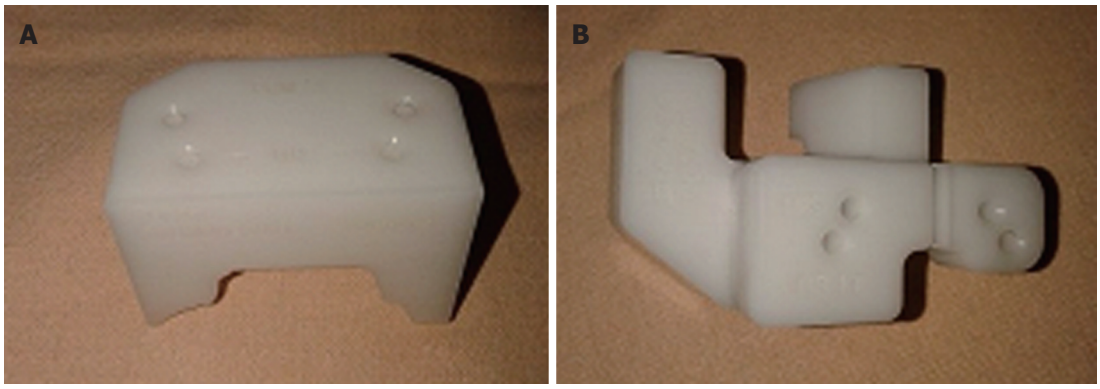

Figure 1 Patient specific blocks used in the present study (A: Femoral jig, B: Tibial jig).

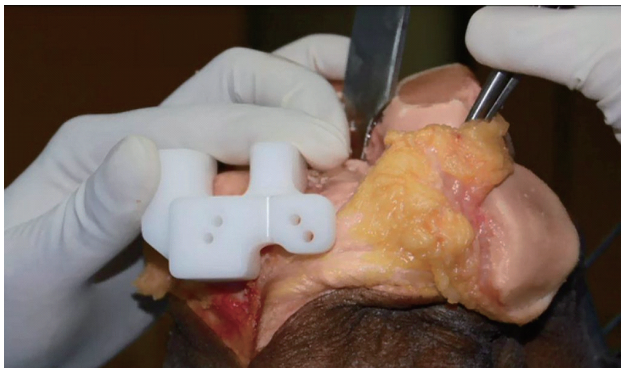

Figure 2 Tibial jig conforming to the proximal tibial anatomy and fiiting well to the bone. This jig is used for insertion of the proximal tibial pins followed by use of the conventional proximal tibial jig.

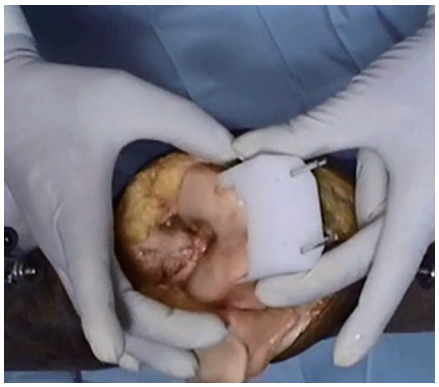

Figure 3 Femoral jig matching the contour of the anterior femoral cortex.

Each group included 40 knees. Most of the knees had varus deformity except six knees in each group having a valgus deformity. CT scan of group 1 subjects was done preoperatively in the same institute and data sent to PSB manufacturing unit. PSB manufactured from Stryker® was used for all subjects in group 1 (Figure 1). Stryker Scorpio™ posterior stabilized knee system was used in all the patients.

An anterior-posterior radiograph of the knee in standing and lateral positions was done preoperatively. Each of these patients was operated by the same surgeon (RV) in the same operating setup and using a similar protocol.

An anterior midline longitudinal incision with modified Insall's approach[20] was used in all the TKA and the majority of operating steps were same in both the groups, except that the alignment guides (PSB) were used in group 1 (Figures 2 and 3). In group 2, E/M alignment guide for tibia and I/M alignment guide for femur were used. The tibial alignment guide was adjusted to achieve 0º varus/valgus cut with 5º posterior slope, whereas on the femoral side 50 valgus (which was reduced to 30 valgus for valgus knees) and 30 external rotation was kept in the alignment guide. The decision to change the femoral valgus angle in the valgus knees in the CI group was taken based on standard protocol and was not based on any pre-operative femoral valgus angle calculation.

We aimed to achieve a rectangular gap of 20 mm which was equally symmetrical in both flexion and extension. All the modifications required in operating steps for proper bone cuts to achieve accurate alignment were done intraoperatively in group 2 patients performed with CI. Whereas, in group 1 patients all the planning was done preoperatively, which started with preoperative CT scannogram followed by an assessment of bone cut on virtual bone models and finally fabrication of blocks after approval from the operating surgeon (Figure 1). These blocks were then used intraoperatively to decide the pin position over which a conventional cutting jig was placed for planned bone cuts.

The primary aim of this study was to compare the postoperative mechanical alignment achieved with using PSI and CI in TKA. The MA was assessed by measuring the Mechanical femoral-tibial angle (MFT angle) on a long-leg radiograph of lower limb done on second follow-up visits, planned after one month of surgery, as most of the patients were able to stand erect with full knee extension and without any support (Figure 4). All the MFT angles were measured by a single individual of the operating unit (VB) who first underwent adequate training in the measurement of the MFT angle on long leg standing radiographs. Outliers of the mechanical axis after total knee arthroplasty were defined as those patients who had values of mechanical axis ± 3 degrees

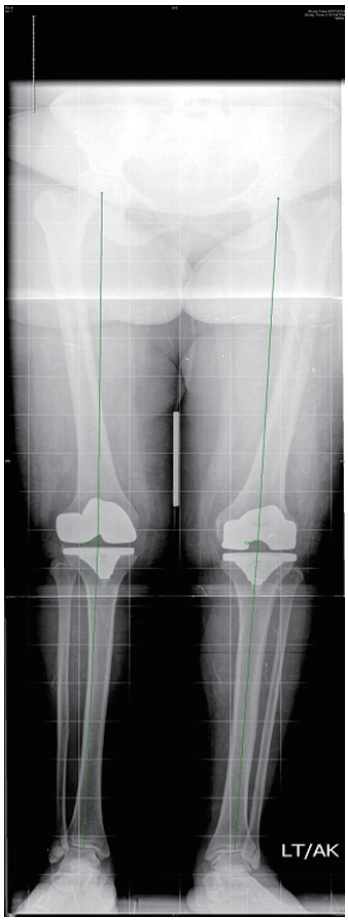

Figure 4 Postoperative weight bearing long leg radiograph measuring the hip-knee-ankle angle axis (mechanical axis).

from the neutral 180º. Thus, inter-observer and intra-observer variation were avoided. Data from both the groups was analyzed and compared using statistical analysis as mentioned below.

Statistical analysis

Statistical analysis was performed by the SPSS program for Windows, version 17.0. Continuous variables were presented as mean ± SD or median and categorical variables were presented as absolute numbers and percentage. Data were checked for normality before statistical analysis. Normally distributed continuous variables were compared using the unpaired *t* test, whereas the Mann-Whitney *U* test was used for those variables that were not normally distributed. Categorical variables were analyzed using either the χ^2 test or Fisher's exact test. For all statistical tests, a *P* value less than 0.05 were taken to indicate a significant difference.

RESULTS

The study group 1 included 17 females and seven males while in group 2 there were 15 females and eight males. The mean age of patients in group 1 was 60.5 years and in group 2 it was 60.2 years. In group 1, 16 patients had bilateral, and 8 had unilateral TKA. In group 2, 17 patients had bilateral, and 6 had unilateral TKA. No significant difference was found between the two groups concerning age, sex, and sidewise distribution. The mean postoperative MFT angle measured on long-leg radiographs in group 1 was 178.23° (SD = 2.67°, range - 171.9° to 182.5°) while in group 2, the mean MFT angle was 175.73° (SD = 3.62°, range - 166.0° to 179.8°) (Table 1).There was a significant difference in postoperative MA achieved in PSI (group 1) compared to CI (group 2) with p-value = 0.001. In group 1 there were 7 outliers while in group 2, there were 17 outliers (Figures 5 and 6). Thus, there was also increase in outliers in group 2 compared to group 1. There was a significant difference in outliers achieved in PSI (group 1) compared to CI (group 2) with p-value = 0.0147 (Figures 5 and 6).

Table 1 comparison of post-operative mechanical femoral tibial angle between the patient specific block group and the conventional group

	PSB group (n = 40) Mean ± SD	Conventional group (n = 40) Mean ± SD	Mean difference	*P* value
Postop MFT angle	178.23 ± 2.67	175.73 ± 3.62	2.497	0.001

Postop: Post operation; MFT: Mechanical femoro-tibial; PSB: Patient specific blocks.

DISCUSSION

The number of TKA annually is increasing every year, but the main concern for a surgeon and patient undergoing primary TKA remains long-term survival of the implant. Also, the increased morbidity, healthcare cost and complications associated with revision surgery have further guided the arthroplasty surgeons to strive for longer implant survival. One of the most important factors contributing to implant survival is accurate mechanical alignment. Various techniques like computer navigation, PSI, *etc.*, have been introduced to achieve precise mechanical alignment in TKA.

The importance of accurate postoperative mechanical alignment of lower limb for the successful outcome of TKA has been accepted widely and supported by several studies. As early as in 1977, Lotke *et al*[21] reported the importance of accurate implant positioning in the long term survival of knee implants. These findings were further corroborated by Ritter *et al*[2] who demonstrated an increased failure rate for knees in varus. Sharkey *et al*[3] concluded that most of the revisions which were performed within the first two years of primary surgery were due to errors in surgical technique. This background knowledge prompted the development and introduction of PSI with the aim of improving post-operative mechanical alignment while bypassing the drawbacks of CAS. Improvement in mechanical alignment with the use of PSI in TKA was first reported

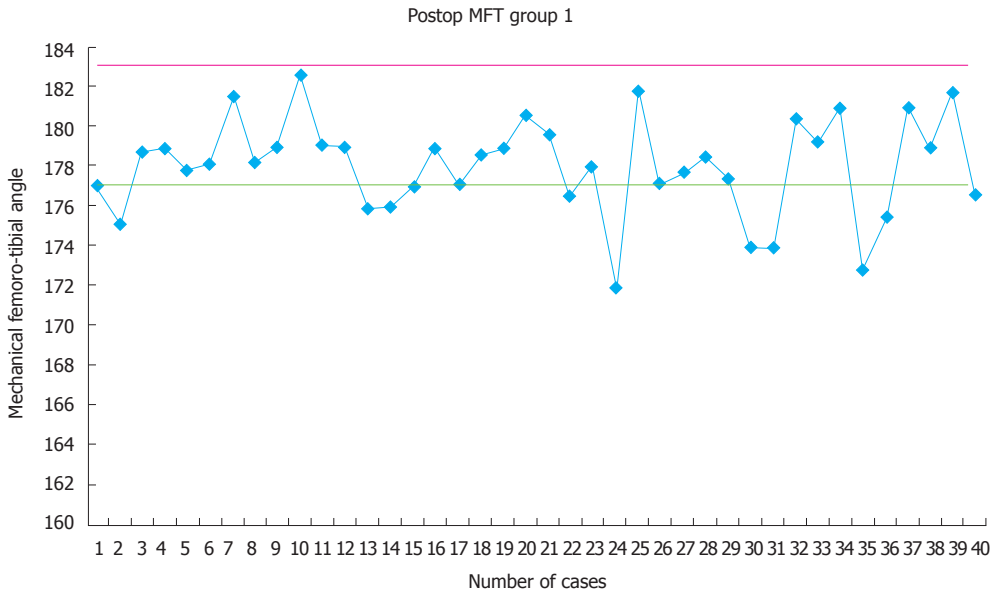

Figure 5 Scatter diagram depicting the distribution of the post-operative mechanical femoro-tibial angle (mechanical femoro-tibial angle) in the patient specific block group (group 1). MFT: Mechanical femoro-tibial angle.

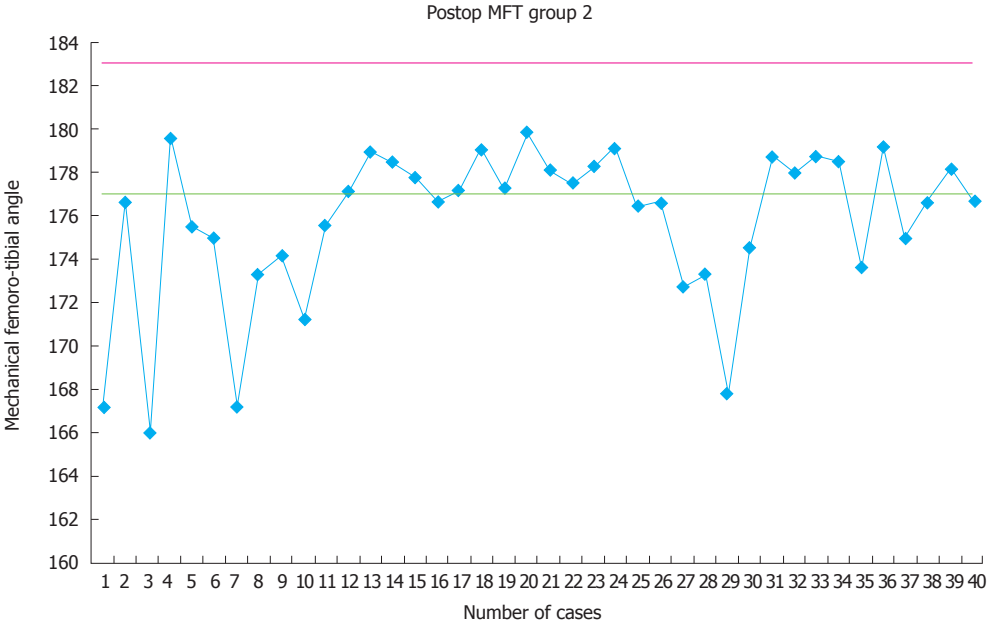

Figure 6 Scatter diagram depicting the distribution of the post-operative mechanical femoro-tibial angle (mechanical femoro-tibial angle) in the conventional instrument group (group 2). MFT: Mechanical femoro-tibial angle.

by Hafez *et al*[22] on cadaveric knees and plastic bones. Similar results were obtained from a study done by Ng *et al*[4] where they compared 569 TKA done with PSI to 155 TKA done with conventional instruments and found decreased a number of outliers with PSI. The numbers of outliers with any technique used in TKA remains an important observation and indicate the efficacy of that technique. We also found decreased outliers with the use of PSI, in our study. Heyse *et al*[23] and Renson *et al*[24] also noticed decreased the number of outliers with PSI (13%) compared to conventional jig (29%).

However, some authors found contradictory results in their studies. Abane *et al*[25] compared 70 TKA done with PSI to an equal number of TKA done with CI and could not find any significant improvement in the post operative MA with the use PSI over CI. Klatt *et al*[26] reported 4 patients operated with PSI showed malalignment in mechanical axis. From their small case series, they opined that this emerging technology ignores conventional principles of TKA and the software used unreliable at present. Chen *et al*[16] in their study of 60 patients found increased outliers with the use of PSI. Parratte *et al*[27] suggested that more refinement needed to be done in PSI to have better control of

Table 2 Comparison of computerized tomography based and magnetic resonance imaging based patient specific blocks

	Computerized tomography based blocks	Magnetic resonance imaging based blocks
Time for study	Lesser (approximately 5 min)	Longer (approximately 45 min)
Cost	Economical	Costlier
Radiation exposure	Uses ionizing radiation. But focused hip-knee-ankle computerized tomography scanogram reduces exposure to 5 mSv (equal to yearly background exposure)	Does not use ionizing radiation
Availability	Easily available	Not available at all centers
Patient turn over	Useful for high patient turnover centers	Not suitable for high patient turnover center
Contraindication	Can be used in patient with any metal prosthesis *in situ*	Cannot be used in patient with metal prosthesis or cardiac pacemaker *in situ*
Based on	Bony landmarks	Cartilage
Accuracy	Comparable	Comparable
Initial infrastructure set up cost	Lower as compared to MRI	High, non affordable for low volume centres
Claustrophobia	No contraindication	Can not be performed in claustrophobic patients

MRI: Magnetic resonance imaging.

the tibial rotation. The inferior results in these studies could have been due to the evolving technology of PSI at the time of these studies, the associated learning curve and different designs of the cutting jig used in the studies[16,25,26].

The other point of discussion in the use of PSI is the radiological method used for pre-operative planning. This technology utilizes advanced imaging techniques like magnetic resonance imaging (MRI) and CT scan to reproduce a 3D model of the patient's bone which is in turn used to create jigs which are used intra-operatively either to directly take bone cuts or to guide the insertion of pins. The debate regarding the best imaging modality is ongoing. In a systematic review of the use of CT *vs* MRI in PSI showed a decreased number of outliers in the CT group (12.5%) as compared to MRI (16.9%) group[28]. There are various advantages of the CT-based PSI over MRI-based (Table 2). The CT scan is associated with decreased cost and imaging times[28]. Moreover, it can be easily performed in patients with cardiac pacemakers and metallic implant *in situ*. The widespread availability of the CT scan in a developing country, also make it accessible to a larger profile of surgeons and patients. Even in high volume center, there is usually a single MRI machine which can further increase the waiting period of the patient. The accuracy of the CT-based PSI as compared to the MRI-based PSI have been found to be comparable[29,30]. The only disadvantage of the CT scan based PSI is the possibility of increased radiation exposure as compared to MRI. This has further been circumvented by the use of focused scans of the hip, knee and ankle, which further decreases the dosage of radiation to 5MSv, which is comparable to the background radiation or approximately 70 chest X-rays[31]. In the present study, CT-based PSI technique was used and found to provide a satisfactory outcome.

In the present study, we found more accurate alignment in TKA done by PSI compared to CI, with a statistically significant improvement in MFT angle in PSI group (P = 0.001). The number of outliers was also decreased with the use of PSI. The mean MFT angle measured postoperatively in group 1 was 178.23° and in group 2 it was 175.73°. The preoperative planning done in PSI group was the main reason for achieving accurate MA postoperatively. The CT scannogram of lower limb done preoperatively gives thorough information of actual bony anatomy of knee and limb alignment. The patient specific blocks thus produced fit to patient's bony anatomy accurately and thus lead to proper pin positioning and accurate bone cuts intraoperatively. Proper pin positioning, and accurate bone cuts can be attributed as the reasons for the improvement in mechanical alignment in TKA with PSI.

PSI avoids manual error involved in pin position with conventional instruments. The accuracy of the valgus cut angle depends on an accurate entry point and the snug fit of the intra-medullary alignment rod in the femoral canal. These two factors are circumvented by the use of PSI and hence may be responsible for better achievement of the distal femoral valgus angle. Moreover, the positioning of the extra-medullary alignment rod for the tibia depends on the surgeon's perception of centralizing the rod at the center of the ankle. This is subject to error due to the surgeon (difference in perception of the alignment) and also due to patient factors like excessive fat and soft tissue around the ankle, *etc*. With the use of PSI, both these factors are decided pre-operatively during planning and hence the manual error is minimized.

There were certain shortcomings of the present study. The study was not a randomized controlled trial. Our sample size was also small, but it can be considered a representative study and further studies should be done to define the exact efficacy of PSI in improving the overall mechanical axis.

Considering the current trends of joint replacement and also due to a consistent increase in the life expectancy of the population, there is going to be an exponential growth in the number of total knee arthroplasty being done. The focus is shifting to increase the long-term survival of the artificial joints in a cost efficient manner, to decrease the financial and health burden of revision

surgery. PSI technology is a step in this direction as it helps in improving the postoperative mechanical axis and also improve the operating room efficiency. This study defines that the PSI technology helps in significantly improving the postoperative mechanical alignment as compared to conventional technology and has future potential to provide long-term benefits to patients.

COMMENTS

Background

The zeal to achieve improved patient outcomes and longer implant survival has opened new avenues in the field of total knee arthroplasty. Patient specific jigs hold promise in achieving both these.

Research frontiers

Patient specific blocks have recently caught the attention of researchers with multiple studies showing good results. These blocks can be manufactured on the basis of the patient's magnetic resonance imaging (MRI) and computer tomography (CT) scan. It is of interest to see whether CT based jigs can achieve good results in the post operative outcomes and alignment of patients undergoing total knee arthroplasty.

Innovations and breakthroughs

The authors confirm that CT based blocks can achieve accurate restoration of the post operative mechanical axis and good post operative pain relief and rehabilitation. The CT scan based blocks hold some inherent advantages over the MRI based jigs and these have also been discussed in the present study.

Applications

CT based patient specific blocks hold promise for the future in the quest to improve implant longevity in total knee arthroplasty.

Peer-review

This manuscript is well organized, and the results are acceptable.

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P- Reviewer: Camera A, Fujimoto E **S- Editor**: Kong JX
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DOI: 10.5312/wjo.v7.i7.434

World J Orthop 2016 July 18; 7(7): 434-441
ISSN 2218-5836 (online)

ORIGINAL ARTICLE

Retrospective Cohort Study

Constrained fixed-fulcrum reverse shoulder arthroplasty improves functional outcome in epileptic patients with recurrent shoulder instability

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Author contributions: Thangarajah T contributed to data collection, data analysis, and manuscript preparation; Higgs D, Bayley JIL and Lambert SM carried out surgeries and critical analysis of manuscript.

Institutional review board statement: This retrospective study was undertaken using data from medical records only and thus was conducted without IRB approval.

Informed consent statement: Our retrospective study contained data from medical records.

Conflict-of-interest statement: The authors declare that they have no conflict of interest. No benefits in any form have been received or will be received from any commercial party related directly or indirectly to the subject of this article.

Data sharing statement: No additional data are available.

Manuscript source: Invited manuscript

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Received: January 12, 2016
Peer-review started: January 14, 2016
First decision: February 29, 2016
Revised: May 4, 2016
Accepted: May 7, 2016
Article in press: May 9, 2016
Published online: July 18, 2016

Abstract

AIM: To report the results of fixed-fulcrum fully constrained reverse shoulder arthroplasty for the treatment of recurrent shoulder instability in patients with epilepsy.

METHODS: A retrospective review was conducted at a single facility. Cases were identified using a computerized database and all clinic notes and operative reports were reviewed. All patients with epilepsy and recurrent shoulder instability were included for study. Between July 2003 and August 2011 five shoulders in five consecutive patients with epilepsy underwent fixed-fulcrum fully constrained reverse shoulder arthroplasty for recurrent anterior shoulder instability. The mean duration of epilepsy in the cohort was 21 years (range, 5-51) and all patients suffered from grand mal seizures.

RESULTS: Mean age at the time of surgery was 47 years (range, 32-64). The cohort consisted of four males and one female. Mean follow-up was 4.7 years (range, 4.3-5 years). There were no further episodes of instability, and no further stabilisation or revision procedures were performed. The mean Oxford shoulder instability score improved from 8 preoperatively (range, 5-15) to 30 postoperatively (range, 16-37) (P = 0.015) and the mean subjective shoulder value improved from 20 (range, 0-50) preoperatively to 60 (range, 50-70) postoperatively (P = 0.016). Mean active forward elevation improved from 71° preoperatively (range,

45°-130°) to 100° postoperatively (range, 80°-90°) and mean active external rotation improved from 15° preoperatively (range, 0°-30°) to 40° (20°-70°) postoperatively. No cases of scapular notching or loosening were noted.

CONCLUSION: Fixed-fulcrum fully constrained reverse shoulder arthroplasty should be considered for the treatment of recurrent shoulder instability in patients with epilepsy.

Key words: Arthroplasty; Dislocation; Epilepsy; Instability; Shoulder

Core tip: Epileptic patients with recurrent shoulder instability pose a significant challenge. We have reported the first series in the literature of patients with epilepsy-related recurrent shoulder instability to be treated with fixed-fulcrum constrained reverse anatomy arthroplasty. Our results suggest that it is successful in reducing pain and eliminating actual and perceived instability in this population. Contrary to previous reports there were no cases of glenoid loosening, implant failure or revision procedures. Postoperatively, there was a significant improvement in functional outcome, which was accompanied by a mean improvement of 25° in active external rotation and 29° in active forward flexion.

Thangarajah T, Higgs D, Bayley JIL, Lambert SM. Constrained fixed-fulcrum reverse shoulder arthroplasty improves functional outcome in epileptic patients with recurrent shoulder instability. *World J Orthop* 2016; 7(7): 434-441 Available from: URL: http://www.wjgnet.com/2218-5836/full/v7/i7/434.htm DOI: http://dx.doi.org/10.5312/wjo.v7.i7.434

INTRODUCTION

Epileptic seizures can cause shoulder dislocation and instability[1]. The incidence of dislocation during a seizure is approximately 0.6% but this is likely to be an underestimation since many may be undetected because medical management of the seizure often takes precedence[2,3]. Recurrent instability is common and occurs soon after the first dislocation, with anterior and posterior instability occurring equally[1]. Significant bone loss from the glenoid rim and fossa, and corresponding extensive humeral head fractures are held responsible for this and are recognized as being a hallmark of the condition[1,4,5]. To address this, the majority of non-arthroplasty ("conservative") surgical strategies focus on restoration and/or augmentation of the bony glenohumeral joint while also addressing capsular insufficiency and arthritis[6,7]. Despite technically satisfactory reconstruction ("preservative") procedures some patients still experience persistent instability and increasing arthritis symptoms. Further conservative reconstruction becomes an unenviable prospect due to poor bone stock, large joint surface defects, and rotator cuff musculotendinous and capsular insufficiency. The patients are often young, in education or seeking work, and find the prospect of living with a painful unstable shoulder unbearable. In this context, arthrodesis has been reported to be a successful treatment strategy but the limitation in range of movement that inevitably results means that it is not suitable for all patients[8]. Constrained arthroplasty may therefore represent an alternative treatment option.

The Bayley-Walker shoulder (Stanmore Implants Worldwide Ltd, United Kindom) was specifically conceived for the treatment of patients with difficult shoulder reconstruction problems such as advanced rotator cuff arthropathy and tumours[9,10]. The device is a constrained fixed-fulcrum reverse anatomy prosthesis comprising a large-pitched, hydroxy-apatite-coated titanium glenoid screw with a 22 mm CoCrMo alloy head that forms a constrained "snap-fit" articulation (to increase stability) with an UHMWPE liner encased in a tapered titanium alloy humeral component giving 60° of intrinsic motion in any direction[10]. The center of rotation is placed medially and distally to the axis of the normal shoulder, which increases the lever arm of the deltoid, but to a lesser degree than most existing non-linked reverse anatomy prostheses[9]. These features make the Bayley-Walker prosthesis a potential treatment option for recurrent shoulder instability in patients with epilepsy who have sufficient glenoid bone stock for secure primary fixation of the glenoid component. There are no reports of this management strategy in the current published literature.

The aim of this retrospective study was to report the results of fixed-fulcrum fully constrained reverse shoulder arthroplasty (FF-RSA) for the treatment of recurrent shoulder instability in patients with epilepsy.

MATERIALS AND METHODS

Between July 2003 and August 2011 five shoulders in five consecutive patients with epilepsy underwent FF-RSA for recurrent instability. Cases were identified using a computerized database and all clinic notes and operative reports were reviewed. The mean duration of epilepsy in the cohort was 21 years (range, 5-51) and all patients suffered from grand mal seizures. The index dislocation occurred a mean of 15 years (range, 2-38) before FF-RSA surgery. All cases were performed by the senior authors (J I L Bayley, Deborah Higgs, and Simon M Lambert). Mean age at the time of surgery was 47 years (range, 32-64) and the cohort consisted of four males and one female. All patients had anterior instability. Three patients had bilateral symptoms. The dominant shoulder was affected in three cases. FF-RSA was performed after an average of two previous

Table 1 Patient details

Case	Gender	Age	Previous stabilisation procedures	Duration of follow-up (yr)	Additional procedures
1	Male	41	Putti-Platt procedure; Allograft humeral head reconstruction; Coracoid transfer; Revision allograft humeral head reconstruction	4.3	-
2	Female	64	Coracoid transfer	4.6	-
3	Male	48	No previous stabilisation procedures	4.8	-
4	Male	32	Putti-Platt procedure; Bankart repair; Revision Bankart repair; Allograft humeral head reconstruction; Humeral head resurfacing	5.0	Examination under anaesthesia and arthrocentesis due to persistent pain
5	Male	51	No previous stabilisation procedures	-	-

stabilisation procedures (range, 0-5) in all but two patients in whom the procedure was used as primary treatment (Cases 3 and 5). All patients had had an onset of instability that coincided with a seizure. In all cases subsequent dislocations occurred during normal activities or further seizures.

The indication for surgery was severe pain and recurrent or persistent (*i.e.*, the shoulder was never able to be actively centralised) instability with a non-functioning rotator cuff. The glenoid bone stock was judged on computer tomography (CT) to be sufficient for primary implantation of the glenoid screw component. Neurological advice was sought preoperatively in all cases to optimise the treatment of the epilepsy. Detailed patient data are presented in Table 1.

Surgical technique

The deltopectoral approach was used in all patients. The glenoid was exposed and a Bayley-Walker uncemented glenoid screw was inserted using standard instrumentation. The humeral canal was prepared, and a Bayley-Walker humeral stem was inserted. A simple sling was used for 6 wk postoperatively during which period passive rotation at waist level was permitted; active scapular postural and motion exercises were encouraged. An active anterior deltoid activation programme was initiated after the first phase when osseo-integration of the glenoid screw was considered likely to have been achieved.

Assessment of radiological and functional outcome

Preoperative and postoperative radiographic imaging was performed in all cases and included anteroposterior and axillary views. Scapular notching was classified by the size of the defect on the anteroposterior radiograph using the four-part grading system devised by Sirveaux *et al*[11]. Humeral loosening was assessed from anteroposterior radiographs as described by Boileau *et al*[12].

Preoperative and postoperative clinical outcome measures included active forward elevation, active external rotation, and the Oxford Shoulder Instability score (OSIS)[13]. Range of movement was assessed by the operating surgeon and/or an orthopaedic resident. The OSIS is a 12-item questionnaire that places a significant emphasis on the impact the patient's shoulder instability has on their lives, making it a highly discriminant tool to assess the efficacy of interventions used to treat it. In addition, all patients were assessed using the subjective shoulder value (SSV). The SSV can be used as a supplementary tool to traditional, more complex outcome measures and may be used in conjunction with other scores to assess the patients' outcome. It has also been suggested to be a more sensitive measure of shoulder function in patients with instability[14].

Statistical analysis

The paired *t* test was used to compare OSIS and SSV before and after surgery. A P value of < 0.05 was considered significant. The SPSS software package, version 22 (SPSS Inc, an IBM Company, Chicago, Illinois) was used to analyse data.

RESULTS

Mean follow-up was 4.7 years (range, 4.3-5 years). One patient was deceased (Case 5) and was therefore exempt from functional outcome analysis. There were no further episodes of instability or persistence of apprehension, and no further stabilisation procedures and no revision procedures were performed. All patients were on medical treatment for their epilepsy and had been reviewed by a neurologist preoperatively. No cases of scapular notching or loosening of either the humeral or glenoid component were noted.

Mean active forward elevation improved from 71° preoperatively (range, 45°-130°) to 100° postoperatively (range, 80°-90°) (P = 0.418). Mean active external rotation improved from 15° preoperatively (range, 0°-30°) to 40° (20°-70°) postoperatively (P = 0.221). Clinical outcome following constrained fixed-fulcrum reverse shoulder arthroplasty can be found in Table 2.

The mean OSIS improved from 8 preoperatively (range, 5-15) to 30 postoperatively (range, 16-37) (P = 0.015). The mean SSV improved from 20 (range, 0-50)

Table 2 Clinical outcome following constrained fixed-fulcrum reverse shoulder arthroplasty

Case	Active forward elevation in degrees (preop/postop)	Active external rotation in degrees (preop/postop)	Oxford shoulder instability score (preop/postop)	Subjective shoulder value (preop/postop)
1	130/80	30/25	5/32	0/60
2	45/80	20/20	15/37	50/70
3	60/90	10/70	6/33	20/60
4	50/150	0/45	7/16	10/50

Preop: Pre operation; Postop: Post operation.

preoperatively to 60 (range, 50-70) postoperatively (P = 0.016). Following surgery all patients reported less pain and avoided fewer activities due to the fear of a further dislocation. An improvement in dressing and washing was also noted in all cases. The results from the OSIS are summarized in Table 3.

One patient (Case 4) complained of persistent pain and underwent arthrocentesis to exclude an infective cause. No organisms were isolated and the patient reported an excellent outcome at the latest follow-up characterized by an improvement in functional outcome.

Case presentations

Case 1: A 41-year-old male with epilepsy was evaluated for a painful and unstable glenohumeral joint eight years after the index dislocation. Four previous stabilisation procedures had been performed, but due to persistent symptoms FF-RSA was undertaken. Following surgery, there was a reduction in pain and an improvement in range of movement due to a stable glenohumeral joint. No prosthetic complications were noted at the latest follow-up (Figure 1).

Case 2: A 64-year-old female epileptic with a 38-year history of glenohumeral instability was reviewed for an unstable shoulder and severe pain. Preoperative CT demonstrated advanced osteoarthritis with subchondral cysts, subchondral sclerosis, and narrowed joint space. FF-RSA was carried out successfully with no implant-related complications. At the latest follow-up, instability had been eliminated and there was a concomitant reduction in pain, and an improvement in function.

Case 3: A 48-year-old male with epilepsy was assessed for a painfully unstable glenohumeral joint two years after the first dislocation. No previous stabilisation procedures had been performed. FF-RSA was undertaken in the setting of an inactive rotator cuff and functioning deltoid. Postoperatively, there were no episodes of instability and the patient was satisfied with the outcome (Figure 2).

Case 4: A 32-year-old epileptic male was reviewed following an 11-year history of glenohumeral instability. Five previous surgeries had been undertaken, including a humeral head resurfacing arthroplasty for dislocation arthropathy. FF-RSA was considered due to ongoing symptoms. At the time of surgery, the humeral head resurfacing implant was loose and there were arthritic changes on the glenoid. One year following surgery the patient complained of persistent pain and so arthrocentesis was undertaken. This did not demonstrate any infective cause and the pain eventually settled (Figure 3).

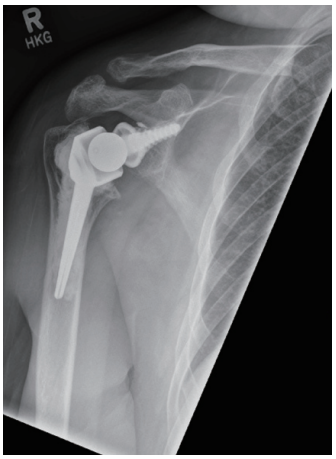

Figure 1 Anteroposterior radiograph of a 41-year-old right hand-dominant male with 4 previous stabilizations, 4 years after a right Bayley-Walker fixed-fulcrum constrained reverse shoulder arthroplasty.

Case 5: A 51-year-old epileptic male was evaluated following a 16-year history of recurrent glenohumeral instability. No previous stabilisation procedures had been undertaken. At the time of surgery, a full-thickness rotator cuff tear was noted. No implant-related complications were detected following surgery. However, the patient later died of an unrelated medical condition.

DISCUSSION

Shoulder instability can be a significant problem in patients with epilepsy. Management is challenging as seizures exert considerable forces on surgical repairs. It is therefore imperative that a neurologist is involved preoperatively so that medical treatment can be optimized. Patients with epilepsy are prone to recurrence and often undergo multiple operations to achieve a stable shoulder joint[1]. With each successive procedure the risk of complications increases and the chance of a successful outcome is reduced. Only a small proportion of cases remain refractory to conventional surgery but these patients often have distorted anatomy and multiple soft tissue defects that make further reconstruction challenging and fraught with complications. To address this, Thangarajah *et al*[8] reported the outcome of six epileptic patients with recurrent instability followed-up for a mean of 39 mo who were treated with glenohumeral arthrodesis. Mean age of the cohort was 31 years. An overall improvement

Table 3 Change in the Oxford Shoulder Instability score following constrained fixed-fulcrum reverse shoulder arthroplasty

Item	No. of patients reporting an improvement	No. of patients reporting worsening	No. of patients reporting no change
No. of dislocations	4	0	0
Dressing oneself	4	0	0
Worst pain	4	0	0
Interference with work	4	0	0
Avoidance of activities due to fear of dislocation	4	0	0
Prevented activities of importance	4	0	0
Interference with social life	3	0	1
Interference with sport/hobbies	1	0	3
Frequency with which patient thinks about their shoulder	3	0	1
Willingness to life heavy objects	1	0	3
Base-line level of pain in shoulder	4	0	0
Avoidance of certain positions when sleeping	2	0	2

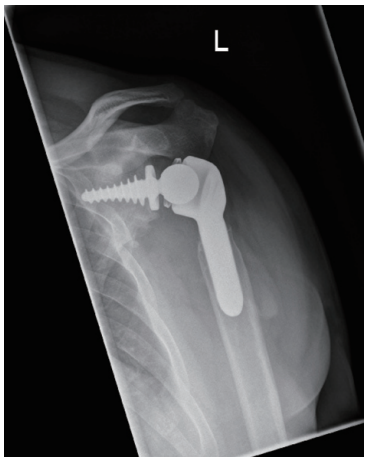

Figure 2 Anteroposterior radiograph of a 48-year-old left hand-dominant male, one year after a left Bayley-Walker fixed-fulcrum constrained reverse shoulder arthroplasty.

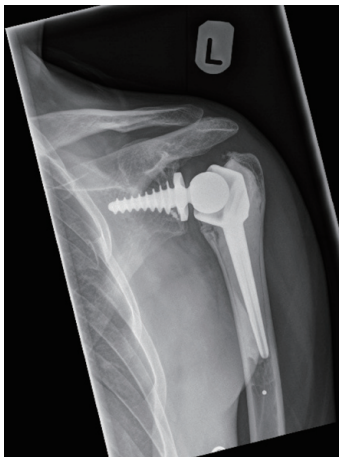

Figure 3 Anteroposterior radiograph of a 32-year-old right hand-dominant male with 5 previous stabilizations, 3 years after a left Bayley-Walker fixed-fulcrum constrained reverse shoulder arthroplasty.

in functional outcome was noted, predominantly due to the reduction in pain and elimination of instability, but due to the restriction in range of movement and limitation in function it could not be recommended ubiquitously. In this context, constrained shoulder arthroplasty could be a potential alternative but there are no studies examining this strategy in an epileptic population.

Constrained shoulder arthroplasty is characterised by mechanical coupling of the humeral and glenoid components around a fixed center of rotation (hence the terminology "fixed-fulcrum" total shoulder arthroplasty). FF-RSA was initially considered as a potential solution for rotator cuff arthropathy but early designs were associated with loosening and implant failure, with relatively poor ranges of motion compared with the emerging anatomical designs[15,16].

The Bayley-Walker shoulder replacement is a constrained FF-RSA device that has been used for complex reconstruction problems in which the rotator cuff is absent (proximal humeral tumour excision), severely deficient (rotator cuff arthropathy), or likely to be compromised with the passage of time (post-traumatic arthropathy, particularly when associated with disruption of the coraco-acromial arch)[9,10]. Its design includes a conical shaped glenoid screw that reduces the strain placed on the implant, and "snap-fit" components that enhance stability and place the center of rotation medially and distally to the axis of the normal shoulder in order to increase the lever arm of the abductors[9,17].

Post *et al*[18] reported the results of 43 constrained total shoulder replacements performed in 42 patients. Follow-up was for a minimum of 27 mo. Indications for surgery included osteonecrosis, complex fractures, juvenile rheumatoid arthritis, and failed arthroplasty. Twelve material failures were noted in 22 stainless steel implants but only two were found in 21 cobalt-chromium prostheses. Glenoid loosening was not encountered. Traumatic dislocation occurred in four cases. Revision surgery was undertaken in 13 patients and was due to further traumatic episodes causing dislocation and material failure. Maintenance of the glenoid vault was found to be essential for secure fixation of the glenoid component and so the authors discouraged the use of constrained arthroplasty in conditions that caused excessive bone loss such as

tumours and severe osteoporosis. Coughlin *et al*[19] evaluated the results of 16 semi-constrained total shoulder arthroplasties performed for intractable pain and severe degenerative disease. Follow-up was for a mean of 31 mo, with one failure due to mechanical loosening. An improvement was noted in total range of motion. Griffiths *et al*[10] reviewed a series of 68 consecutive patients who underwent replacement of the proximal humerus for tumour using a massive endoprosthesis. The mean age of the group was 46 years and follow-up was for a mean of 5 years 11 mo. An unconstrained endoprosthesis was implanted into the first 64 patients and a custom-made constrained (Bayley-Walker) reverse polarity fixed-fulcrum implant linked to a massive proximal humeral endoprosthesis was used in the remaining four. No dislocations were noted in the group with a constrained implant at a mean of 14.5 mo following surgery. This was in contrast to the unconstrained group who had a dislocation rate of 25.9%. No cases of glenoid loosening were identified in the cohort.

Reverse total shoulder arthroplasty (RTSA) has been established as an effective treatment for rotator cuff arthropathy in an older patient population with low functional demands[20]. Few studies have examined its use in patients under the age of 60 years with those that do reporting serious complications in the short-term such as the need for revision and dislocation[21,22]. Muh *et al*[21] retrospectively reviewed 66 patients with a mean age of 52 years who underwent RTSA. Prior surgical procedures were common with 67% of the cohort having at least one prior intervention. At a mean follow-up of 36.5 mo there was an improvement in range of movement and functional outcome. Ten complications were identified including five dislocations that required revision in three cases and three infections that required further surgery. Scapular notching was found in 43% of patients. Sershon *et al*[22] evaluated 35 patients with a mean age of 54 years that underwent RTSA for a range of indications such as rheumatoid arthritis, failed rotator cuff repair, instability sequelae and cuff tear arthropathy. Of these, 83% had previous surgery averaging 2.5 procedures per patient. At a mean follow-up of 2.8 years there was an improvement in range of movement and functional outcome. Six patients had major complications including three dislocations, one subluxation and two fractures. Furthermore, three patients had revision surgery at 2 mo, 6 mo and 2.8 years.

Younger patients requiring shoulder arthroplasty present several challenges as they have higher functional demands and require longer implant survival when compared to elderly patients. In our series, FF-RSA was used in patients with a mean age of 47 years. At short-term follow up there was an improvement in functional outcome with no dislocations or revisions. This suggests that FF-RSA may be able to overcome some of the problems associated with more traditional reverse anatomy designs and provide a suitable alternative to invasive fusion surgery in a younger population. However, further long-term studies are required because clinical outcomes have been noted to deteriorate with time following RTSA[23].

We have reported the first series in the literature of patients with epilepsy-related recurrent shoulder instability to be treated with fixed-fulcrum constrained reverse anatomy arthroplasty. Our results suggest that it is successful in reducing pain and eliminating actual and perceived instability in this population. Contrary to previous reports there were no cases of glenoid loosening, implant failure or revision procedures. Post-operatively, there was a significant improvement in functional outcome as illustrated by an increase in the OSIS and SSV. This was accompanied by a mean improvement of 25° in active external rotation and 29° in active forward flexion. This is in contrast to other studies in which limitation of active external rotation characterises reversed-polarity semi-constrained prostheses[24-26]. We attribute our findings to the maintenance of teres minor and recruitment of the posterior deltoid, which can work more effectively against a fixed fulcrum with its less-medialised center of rotation[27].

Limitations of this small cohort study include those associated with its retrospective design and the mean follow-up of 4.7 years, which is relatively short in terms of prosthesis survivorship. However, Uri *et al*[27] have shown that if a FF-RSA is becoming loose, it is usually apparent within this time frame. We believe that these prostheses can obtain durable fixation through secondary osseointegration even in the challenging environment of a patient in whom epilepsy is a concomitant problem. This study of a unique group of patients previously considered poor candidates for shoulder arthroplasty provides some evidence of the value of FF-RSA in the treatment of a difficult and relatively uncommon problem. Further prospective studies examining the performance of this implant in this and similarly challenging patient populations in which the stability of conventional reverse-polarity arthroplasty remains uncertain are warranted.

In conclusion, epileptic patients with recurrent shoulder instability pose a significant challenge. Traditional operative measures may be unsuccessful and multiple revisions are common. In our series, FF-RSA eliminated recurrent instability and significantly improved functional outcome. This was accompanied by an improvement in pain and range of movement (external rotation and forward flexion). When managing this complex patient group, medical optimisation is essential and we therefore recommend that a neurologist be involved at the earliest possible juncture.

COMMENTS

Background

Epileptic patients with recurrent shoulder instability pose a significant challenge. Significant bone loss from the glenoid and humeral head is a common finding.

Patients are often young, in education or seeking work, and find the prospect of living with a painful unstable shoulder unbearable. In this context, arthrodesis has been reported to be a successful treatment strategy but the limitation in range of movement that inevitably results means that it is not suitable for all patients. The purpose of this study was to report the results of fixed-fulcrum fully constrained reverse shoulder arthroplasty for the treatment of recurrent shoulder instability in patients with epilepsy.

Research frontiers

The authors have reported the first series in the literature of patients with epilepsy-related recurrent shoulder instability to be treated with fixed-fulcrum constrained reverse anatomy arthroplasty. This study of a unique group of patients previously considered poor candidates for shoulder arthroplasty provides some evidence of the value of fixed-fulcrum constrained reverse anatomy arthroplasty in the treatment of a difficult and relatively uncommon problem.

Innovations and breakthroughs

At a mean of 4.7 years follow-up, there were no further episodes of instability, and no further stabilisation or revision procedures were performed in the cohort. A significant improvement was noted in functional outcome, as assessed by the Oxford Shoulder Instability Score (P = 0.015) and Subjective Shoulder Value (P = 0.016). This was accompanied by a mean improvement of 25° in active external rotation and 29° in active forward flexion. No cases of scapular notching or loosening were noted.

Applications

Fixed-fulcrum fully constrained reverse shoulder arthroplasty should be considered for the treatment of recurrent shoulder instability in patients with epilepsy.

Peer-review

This is a well written paper regarding a very difficult clinical problem to manage.

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P- Reviewer: Anand A, Fanter N, Garg B, Yamakado K
S- Editor: Kong JX **L- Editor**: A **E- Editor**: Wu HL

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DOI: 10.5312/wjo.v7.i7.442

World J Orthop 2016 July 18; 7(7): 442-447
ISSN 2218-5836 (online)

ORIGINAL ARTICLE

Retrospective Study

Trochanter/calcar preserving reconstruction in tumors involving the femoral head and neck

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Author contributions: Lee YK designed and performed the research and wrote the paper; Koo KH designed the research and supervised the report; Cho HS designed the research and contributed to the analysis; Cho HS, Lee YK, Ha YC and Koo KH provided clinical advice.

Institutional review board statement: The design and protocol of this retrospective study were approved by the institutional review board in Seoul National University Bundang Hospital.

Informed consent statement: Patients were not required to give informed consent to the study because the analysis used anonymous clinical data that were obtained after each patient agreed to treatment by written consent.

Conflict-of-interest statement: We have no financial relationships to disclose.

Data sharing statement: No additional data are available.

Manuscript source: Invited manuscript

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Received: March 3, 2016
Peer-review started: March 4, 2016
First decision: April 15, 2016
Revised: April 22, 2016
Accepted: May 10, 2016
Article in press: May 11, 2016
Published online: July 18, 2016

Abstract

AIM: To evaluate the results of hip reconstruction with extensive excision for tumor confined to the femoral head and neck.

METHODS: We designed a resection preserving the greater trochanter and lower portion of calcar femorale, and utilized conventional total hip prosthesis. We retrospectively reviewed 7 patients, who underwent a wide resection and reconstruction using conventional hip prosthesis. There were 3 men and 4 women and their mean age was 42.5 years (22 to 65 years). The histologic diagnosis of each patient was low-grade osteosarcoma, diffuse large B-cell lymphoma, liposclerosing myxofibroma, intraosseous lipoma, chondroblastoma, giant cell tumor and focal intramedullary fibrosis.

RESULTS: One patient with lymphoma died due to disease dissemination at 10 mo postoperatively and the remaining 6 patients were followed for a mean of 4.7 years (3 to 6 years). All patients were able to return to their daily activities and no patient had local recurrence. No radiographic signs of loosening, wear, and osteolysis were found at the last follow-up.

CONCLUSION: Trochanter/calcar-preserving resection

of the proximal femur and reconstruction using conventional total hip prosthesis, is a satisfactory treatment for tumors confined to the femoral head and neck.

Key words: Total hip arthroplasty; Reconstruction; Tumor; Femoral head

Core tip: This is a retrospective study to evaluate the results of trochanter and calcar preserving reconstruction in tumors involving the femoral head and neck. While usual osteotomy for primary total hip arthroplasty is made straightly at 0.5 inch above the lesser trochanter, we made a curved osteotomy in coronal plane from the tip of greater trochanter to lower level or below the lesser trochanter to remove the tumor lesion confined to femoral head and neck. This technique can preserve the greater trochanter and lower portion of the calcar femorale. This surgical technique is a satisfactory treatment for tumors confined to the femoral head and neck.

Cho HS, Lee YK, Ha YC, Koo KH. Trochanter/calcar preserving reconstruction in tumors involving the femoral head and neck. *World J Orthop* 2016; 7(7): 442-447 Available from: URL: http://www.wjgnet.com/2218-5836/full/v7/i7/442.htm DOI: http://dx.doi.org/10.5312/wjo.v7.i7.442

INTRODUCTION

The proximal femur is a common site of primary malignant bone tumors, including chondrosarcoma, Ewing's sarcoma and osteosarcoma[1]. An array of benign bone tumors; giant cell tumor, chondroblastoma, and clear cell chondrosarcoma may develop at the proximal femoral epiphysis and extend to the metaphysis[2-4]. Intra-articular involvement is rare in these tumors, although they might occur following a pathologic fracture.

Patients with tumors confined to the femoral head have been candidates for curettage-bone graft[5] or an extensive resection of the proximal femur and reconstruction, usually hemiarthroplasty, using tumor prosthesis depending on the biologic aggressiveness of the tumor[6-8].

In the process of resection and reconstruction, the greater trochanter is osteomized or excised and after then, the greater trochanter or the insertion of the abductor muscle is attached to the tumor prosthesis with wires or cable grip, which can induce complications including wire or cable breakage, trochanteric fragment migration, nonunion, bursitis, and metallosis[9-12].

Tumor prosthesis is highly costive. One more drawback of the tumor prosthesis is the difficulty on determining the actual length and width of the resected bone even with a use of modular endoprosthesis[13]. Intermediate to long term survivorship of bipolar tumor prosthesis is not satisfactory compared to conventional total hip arthroplasty[7,8,14].

Since 2007, we have treated tumors involving femoral head and neck using a trochanter/calcar-preserving resection and conventional total hip prosthesis. In this study, we present the operative technique and evaluate the results after trochanter-preserving resection with use of conventional prosthesis.

MATERIALS AND METHODS

The surgical treatment algorithm of tumors of the femoral head and neck at our department is as follows. The primary treatment for histologically-proven benign bone tumors confined to the femoral head and neck is curettage and bone graft[5]. However: (1) when there is a suspicion of malignancy or solitary bone metastasis; (2) when there is a risk of superior retinacular or lateral epiphyseal arterial damage during the curettage and consequent develop of osteonecrosis; (3) when the lesion is large and located at the subchondral portion of the femoral head apex and consequent collapse is expected after the curettage; and (4) when there is a local recurrence after the curettage, we perform a trochanter/calcar-preserving resection of the proximal femur and reconstruction using conventional hip prostheses.

Tumors with a pathologic fracture, an involvement of the greater trochanter, involvement of the lesser trochanter, cortical penetration, or intra-articular involvement are treated with more extensive resection and reconstruction using revision prosthesis or tumor-prosthesis.

Between June 2007 and December 2011, 20 patients were operated due to tumors of the femoral head and neck at the authors' hospital. Among them, 13 patients were treated with curettage with bone graft (11 patients) or cement filling (2 patients). The remaining seven patients, who were operated with a trochanter-preserving resection of the proximal femur and total hip arthroplasty using conventional prosthesis, were subjects of this study.

There were 3 men (3 hips) and 4 women (4 hips), and the mean age at the time of operation was 42.5 years (range, 22 to 65 years). All patients presented with a pain of affected hip. The mean time interval between the onset of hip pain and the index operation was 13.6 mo (range, 1.3 to 48.1 mo).

We made preoperative diagnoses and evaluated the tumor extent on plain radiographs, computed tomography and/or magnetic resonance image. We planned a wide resection ($\geqslant$ 1 cm from the tumor margin) bearing in mind the possibility of the malignant tumor (Figure 1).

All THAs were carried out by one surgeon. The patient was placed in a lateral position on the operating room table. A longitudinal posterolateral incision was made. Trochanteric bursa and underlying fat tissues

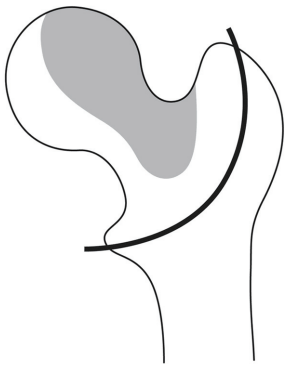

Figure 1 Preoperatively a wide resection margin was planned ⩾ 1 cm from the tumor margin.

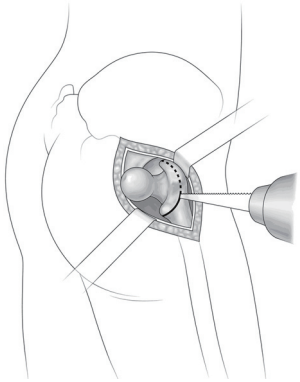

Figure 2 To obtain adequate resection margin from tumor, the curved osteotomy is performed from middle to lower portion of the lesser trochanter to the tip of greater trochanter.

were removed to expose short external rotators and sciatic nerve. External rotators were cut at their tendinous attachments to the trochanteric crest and the posterior capsule of the hip joint was incised. The femoral head was dislocated posteriorly.

To remove the diseased femoral head and neck, preserving the greater trochanter and lower portion of the calcar femorale, we made a curved osteotomy in coronal plane from the tip of greater trochanter to lower level or below the lesser trochanter according to the tumor margin while usual osteotomy for primary total hip arthroplasty is made straightly at 0.5 inch above lesser trochanter. The posterior cortex and endosteal cancellous bone was cut using a 7 mm width osteotome and the anterior cortex was cut with use of a reciprocating saw (Figure 2). This osteotomy is similar to curved varus osteotomy, which has been use for the surgical treatment of femoral head osteonecrosis[15].

After the planned intertrochanteric osteotomy, attachments of vastus muscles and the anterior capsule were detached from the intertrchanteric line of proximal segment to remove the proximal segment. And attachment of psoas was partially detached from the lesser trochanter.

On the inspection of the resected segment, no tumor showed a penetration into the joint or cortical invasion. Resected specimens were submitted for pathological evaluation.

The rest of the procedure was performed in the ordinary manner of cementless THA.

Three designs of implants were used; PINNACLE cup with Corail stem (DePuy, Saint-Priest, France) in 4 hips, PLASMACUP® SC acetabular component with BiCONTACT® stem (Aesculap, Tuttlingen, Germany) in 2 hips, and Bencox cup with Bencox stem (Corentec, Seoul, South Korea) in 1 hip. Third-generation ceramic articulation (BIOLOX Forte alumina head and liner; CeramTec, Plochingen, Germany) was used in 2 hips, and fourth-generation (BIOLOX Delta alumina head and liner; CeramTec) in 5 hips. The diameter of the femoral head was 28 mm in 1 hip, 32 mm in 5 hips, and 36 mm in 1 hip.

The final diagnoses by histological examination were low-grade osteosarcoma, diffuse large B-cell lymphoma, liposclerosing myxofibroma, intraosseous lipoma, chondroblastoma, giant cell tumor, and focal intramedullary fibrosis (Table 1). Surgical margins were negative for the tumor in all patients.

Two patients with malignant tumors underwent a computed tomographic scan of the chest and whole body bone scan, which revealed no evidences of distant metastasis. The patient with lymphoma was treated with adjuvant chemotherapy. Patients were instructed to walk with partial weight bearing with the aid of two crutches for four weeks after surgery.

Routine follow-up visits were scheduled for six weeks, three, six, nine, twelve months, and six months thereafter. Patients who had not returned for regularly scheduled visits were contacted by telephone.

Clinical evaluation was performed with use of the Harris hip score (HHS)[16], and the functional classification system of the International Society of Limb Salvage (ISOLS), which includes six functional parameters; pain, function, emotional acceptance, use of walking supports, walking ability, and gait. Each parameter is scored from 0 to 5 (a maximum score of 30)[17].

The radiographic evaluation was done to confirm if there was the evidence of recurrence; a newly discovered osteolytic or osteosclerotic lesion on follow-up radiographs or MRI.

Fixations of the acetabular and femoral components[18,19], ceramic wear[20] and osteolysis[21-23] were assessed on serial radiographs.

The design and protocol of this retrospective study were approved by the institutional review board in our hospital, which waived the informed consents.

RESULTS

Fracture of the greater trochanter occurred in one patient who was operated due to intraosseous lipoma. The fracture was detected on postoperative 6 wk radiographs. In this patient, the inner portion of the greater trochanter had been removed during the operation to achieve a wide resection and only a thin

Table 1 Patients demographics

Patient	Sex/age	Initial diagnosis on MRI	Final histologic diagnosis	Follow-up (yr)	ISOLS score
1	F/36	Giant cell tumor	Liposclerosing myxofibroma	6	30
2	F/38	Low-grade osteosarcoma	Low-grade osteosarcoma	6	29
3	F/64	Clear cell chondrosarcoma	Intraosseous lipoma	5	28
4	M/22	Chondroblastoma	Chondroblastoma	5	30
5	M/33	Giant cell tumor	Giant cell tumor	3	30
6	F/60	Breast cancer metastasis	Focal intramedullary fibrosis	3	28
7	M/41	Aneurysmal bone cyst	Lymphoma	Died at 10 mo	

F: Female; M: Male; MRI: Magnetic resonance imaging.

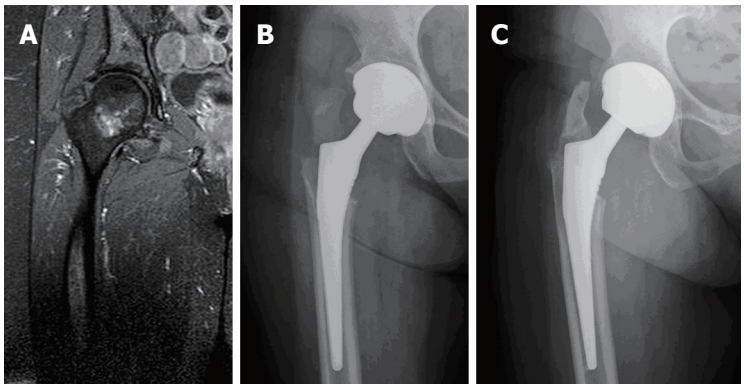

Figure 3 Radiographs of patient 3. A: Preoperative magnetic resonance imaging of a 64-year-old woman shows a high signal lesion confined to femoral head and neck; B: Postoperative radiograph at 6 wk after total hip arthroplasty with extensive excision at the intertrochanteric level shows an avulsion fracture of greater trochanter; C: Postoperative radiograph at 5 years after total hip arthroplasty with extensive excision at the intertrochanteric level shows no evidence of local recurrence and implant loosening or osteolysis.

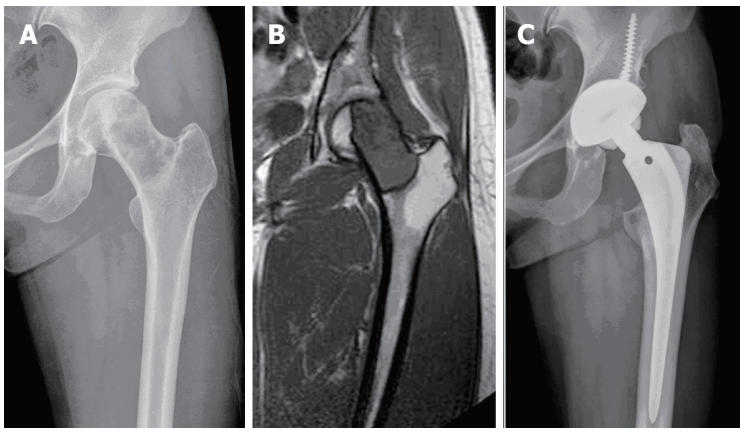

Figure 4 Radiographs of patient 1. A: Radiograph of a 36-year-old woman shows an osteolytic lesion confined to femoral head and neck; B: Preoperative magnetic resonance imaging shows a low signal intensity lesion with well-defined surrounding rim; C: Postoperative radiograph at 6 years after total hip arthroplasty with extensive excision at the intertrochanteric level shows no evidence of local recurrence and implant loosening or osteolysis.

cortical portion had been left. This patient had no history of trauma, and had little pain postoperatively. Thus, the fracture seemed to be an avulsion fracture. It healed completely with protected weight-bearing for 3 mo and an osseointegration of the prosthesis was achieved (Figure 3).

One patient with lymphoma recovered well and returned to his normal activity after the operation. The postoperative radiographs at 6 mo were uneventful. However, this patient died because of disseminated disease at the 10 mo postoperative.

The remaining 6 patients were followed-up for an average of 4.7 years (3 to 6 years). All patients returned to their daily activities and were walking with full weight bearing. At the last follow-up, the mean HHS was 98.0 points (range 96-100 points), and the mean ISOLS functional score was 29.2 points (range 28-30 points) (Table 1). During the follow-up there was no evidence of local recurrence. At the latest follow-up, there were no radiographic signs of aseptic loosening, wear, or osteolysis (Figure 4).

DISCUSSION

The proximal femur is a common site for primary bone tumors and the most common site of metastatic tumors. Since the introduction of tumor prosthesis in 1980s, tumors in the femoral head and neck have been treated with extensive resection and reconstruction of the proximal femur. The femur is resected below the lesser trochanter and the greater trochanter is resected or osteomized. The reconstruction is consisted of a hemiarthroplasty with use of an endoprosthesis. Although modular system improved the endoprostheic reconstruction[14], there are several drawbacks in this procedure. The greater trochanter is osteotomized or

the abductors are transected through their tendinous attachments. If the greater trochanter is resected *en-bloc*, the remaining abductors are attached to the prosthesis. If a fragment of the greater trochanter is remains, it is fixed to the prosthesis with a cable grip system.

However, problems have appeared after the use of cable grip. Silverton *et al*[24] reviewed 68 trochanteric osteotomies, which were repaired with Dall-Miles cable grip system (Howmedica, Rutherford, NJ). Trochanteric nonunion occurred in 25%, with fraying and fragmentation of the cable. Among the 51 patients with trochanteric union, 35% also had signs of fraying and fragmentation. Osteolysis around the cable was seen in 10%. Metallosis at the inferior border of the acetabulum were seen in 12%[24].

As the life expectancy of patients with bone tumors improves, endoprosthetic replacement of the proximal femur is not durable in young patients with low-grade tumor (IA/IB or benign)[7]. Bernthal *et al*[7] reviewed 86 proximal femoral replacements used for tumor reconstruction. Their study included 43 high-grade tumors (IIA/IIB), 20 low-grade tumors (IA/IB or benign), and 23 with metastatic disease. The 5-, 10-, and 20-year survival for IIA/IIB patients was 54%, 50%, and 44%, respectively; all patients with low-grade disease survived; the 5-year survival rate for patients with metastatic disease was 16%. The 5-, 10- and 20-year implant survivorships were 93%, 84%, and 56%, respectively. Although bipolar proximal femoral reconstruction proved a durable technique in patients with metastatic disease and high-grade disease, patients with low-grade disease outlived their implants[7]. In this study, there was a suspicious solitary bone metastasis (patient 6), which was confirmed intramedullary fibrosis. Solitary bone metastasis is defined as a single skeletal metastasis with no tumor in any other part of the body including the primary cancer site or with a primary lesion in resectable status. Although there has been some debate on whether curative resection for a solitary bone metastasis leads to survival gain, most authors believed that patients with a solitary bone metastasis from several cancers live longer than those with multiple metastasis regardless of treatment modalities. Jung *et al*[25] reported that patients who had wide resection for a solitary bone metastasis had a disease-specific survival rate of 100% at mean follow-up of 69 mo. Therefore, they suggested that patients with solitary bone lesion are candidates for aggressive surgical treatment with curative intent. In addition, durable reconstruction is needed to avoid revision surgery which may complicate future management for cancer.

Our study showed that trochanter/calcar preserving resection allows adequate surgical margins for tumors in femoral head and neck and reconstruction using conventional total hip prosthesis affords a satisfactory functional outcome without local recurrence or prosthetic loosening.

There were several limitations in our study. Our study was a retrospective review performed in a small number of cases. There was no control group of wide resection of the proximal femur and reconstruction using endo-prosthesis. However, the mean HHS (98 points) was satisfactory at last follow-up, which was comparable with that of primary THA[26]. Our procedure is applicable on the condition that the lesion was confined to the femoral head and neck. Therefore, a careful evaluation of tumor extent using MRI is mandatory preoperatively.

Our results of trochanter/calcar preserving resection and reconstruction using conventional total hip prosthesis were satisfactory. We would recommend this procedure as a primary surgical treatment along with curettage and bone graft for tumors confined to the femoral head and neck, especially in young patients.

COMMENTS

Background

Tumors confined to the femoral head have been treated by curettage-bone graft or tumor prosthesis following an extensive resection of the proximal femur according to the suspected pathology. When using tumor prosthesis with high cost, an extensive resection of the proximal femur leads to sacrifice of the greater trochanter or the insertion of the abductor muscle. In this study, the authors presented a trochanter/calcar-preserving resection and conventional total hip prosthesis in tumors involving femoral head and neck, and evaluated the outcomes.

Research frontiers

This study contributes to presenting the surgical technique of total hip arthroplasty following trochanter/calcar-preserving resection in the selected patients.

Innovations and breakthroughs

In this study, trochanter/calcar-preserving resection was presented for patients with tumor confined to femoral head and neck. This technique does not require a sacrifice of greater trochanter which abductor muscles are inserted. This means that patients can preserve their abductor mechanism, even though surgery of proximal femur.

Applications

This study suggests that trochanter/calcar-preserving resection is useful for treatment in patients with tumor confined to femoral head and neck.

Terminology

Calcar: The dense, vertically oriented bone present in the posteroemedial region of the femoral shaft inferior to the lesser trochanter of the femur.

Peer-review

The effort to reduce postoperative morbidity by preserving trochanter and calcar seemed to have yielded a good result.

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P- Reviewer: Bicanic G, Jun Y **S- Editor**: Ji FF **L- Editor**: A
E- Editor: Wu HL

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DOI: 10.5312/wjo.v7.i7.448

World J Orthop 2016 July 18; 7(7): 448-451
ISSN 2218-5836 (online)

CASE REPORT

Extensor pollicis brevis tendon can hyperextend thumb interphalangeal joint in absence of extensor pollicis longus: Case report and review of the literature

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Author contributions: All authors contributed to the acquisition of data, writing, and revision of this manuscript.

Institutional review board statement: Attached-no formal IRB approval needed for case report.

Informed consent statement: The patient involved in this study gave his verbal consent to inclusion in this case report.

Conflict-of-interest statement: The authors have no conflict of interests.

Manuscript source: Invited manuscript

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Received: January 30, 2016
Peer-review started: February 1, 2016
First decision: April 15, 2016
Revised: April 17, 2016
Accepted: May 7, 2016
Article in press: May 9, 2016

Published online: July 18, 2016

Abstract

We are reporting a case of extensor pollicis longus tendon rupture which did not require tendon transfer owing to the ability of the intact extensor pollicis brevis (EPB) to fully hyperextend the thumb interphalangeal joint. The thumb metacarpophalangeal joint was also able to be fully actively extended by the EPB. Previous anatomical studies have demonstrated that the insertional anatomy of the EPB tendon is highly variable and sometimes inserts onto the extensor hood and distal phalanx, which is likely the mechanism by which our patient was able to fully extend the thumb interphalangeal joint. Despite the potential for the EPB to extend the IP joint of the thumb, virtually all previously reported cases of extensor pollicis longus (EPL) tendon rupture had deficits of thumb IP extension requiring tendon transfer. This case highlights the potential ability of the EPB tendon to completely substitute for the function of the EPL tendon in providing thumb IP joint extension.

Key words: Extensor pollicis brevis; Extensor pollicis longus; Tendon rupture; Extensor pollicis longus tendon rupture

Core tip: The extensor pollicis brevis may be able to substitute for extensor pollicis longus (EPL) function in some patients when EPL has ruptured.

Strauch RJ, Strauch CB. Extensor pollicis brevis tendon can hyperextend thumb interphalangeal joint in absence of extensor

pollicis longus: Case report and review of the literature. *World J Orthop* 2016; 7(7): 448-451 Available from: URL: http://www.wjgnet.com/2218-5836/full/v7/i7/448.htm DOI: http://dx.doi.org/10.5312/wjo.v7.i7.448

INTRODUCTION

Extensor pollicis longus (EPL) tendon rupture may occur spontaneously or following distal radius fracture, surgical fixation, or repetitive use[1,2]. Virtually all reported cases of EPL tendon rupture have been treated by tendon transfer or tendon grafting in order to restore thumb interphalangeal joint extension. The extensor pollicis brevis (EPB) tendon is classically described as inserting upon the base of the thumb proximal phalanx providing thumb metacarpophalangeal joint extension. Several anatomic studies, however, have demonstrated the EPB insertional anatomy to be highly variable with potential insertions on the extensor hood as well as the distal phalanx (Table 1)[3-8]. One study demonstrated that 21% of EPB tendons, when pulled on at the wrist level, would cause thumb IP joint extension in cadavers[7]. It is therefore interesting that, to the best of our knowledge, there have been no reports of EPL rupture in which thumb MP and IP joint extension remains normal. We report a case of EPL tendon rupture in which normal thumb MP and IP motion was preserved most likely owing to the EPB inserting on the extensor hood and/or the distal phalanx.

CASE REPORT

A 64-year-old right handed man had sustained a right wrist scaphoid fracture in 1969 which went on to develop a scaphoid nonunion advanced collapsed deformity (SNAC wrist) with arthritic changes and a large bone spur from the dorsal scaphoid. Nevertheless, he had no wrist pain and the right wrist never bothere him enough to warrant treatment. Six weeks prior to presentation, he was reaching for something and felt a pop in his right wrist. His physician referred him for hand surgery evaluation. On physical examination his right wrist extended to 30 degrees and flexed to 30 degrees compared to 45 degrees and 70 degrees respectively on the left wrist. His right wrist had bony enlargement dorsally on the radial side, but was non-tender and there was no pain with motion. His EPL tendon was noted to be completely ruptured. However, his EPB tendon could visibly fully extend both the thumb MP and IP joints (Figures 1-3). His active thumb motion was 0-60 degrees at the MP joint and 15 degrees of hyperextension to 50 degrees of flexion at the IP joint. The only difference between the right and left thumbs was that when his hands were put flat on the table, he was unable to fully lift the right thumb up off the table compared to the left side (Figure 4 and Video core tip 1). X-rays of his right wrist revealed a SNAC wrist with a large bony osteophyte protruding dorsally from the scaphoid. The impression was that the EPL tendon had ruptured over the dorsal spike from the scaphoid as this was directly underlying the normal course of the EPL tendon and there was concern that his wrist extensors might also rupture over the bony spike. Therefore, a magnetic resonance imaging of the right wrist was obtained which revealed that the extensor carpi radialis longus (ECRL) tendon was 50% eroded over the dorsal spike of the scaphoid nonunion and that the EPL tendon had completely ruptured. After discussion with the patient, it was decided to perform a surgical procedure to debride the bone spike from the scaphoid nonunion that could potentially lead to rupture of his wrist extensor tendons, having already ruptured the EPL tendon. Since he had no pain in the wrist nor deficits of thumb IP joint extension we did not plan to perform any procedure to address the arthritis or the EPL rupture. At the time of surgery, which was performed under a regional anesthetic, the EPL tendon was seen to be completely ruptured with the proximal end retracted and the ECRL tendon was 50% eroded. The sharp spikes from the scaphoid nonunion were debrided and the capsule was closed over the scaphoid as a soft tissue interposition between the bone and the wrist

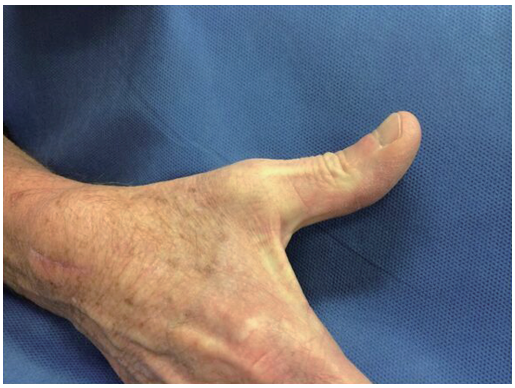

Figure 1 Patient demonstrating full thumb MP and IP extension using only extensor pollicis brevis tendon.

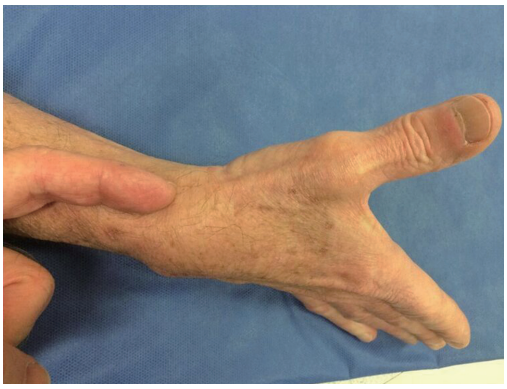

Figure 2 Examiner's finger pointing to extensor pollicis brevis tendon which is extending thumb at MP and IP joints.

Table 1 Previous anatomic studies of extensor pollicis brevis insertional anatomy

Ref.	Type of study	# of hands	EPB insertion site
Stein[3]	Cadaver study	42 cadavers, 84 wrists dissected	No comment on insertional anatomy
Dawson *et al*[4]	Cadaver Study	16 hands of eight cadavers	56% inserted partly to the base of the first phalanx and partly to the extensor hood 25% inserted entirely on the base of the thumb proximal phalanx 19% inserted entirely onto the extensor hood 5% were absent - the Abductor pollicis longus tendon instead inserted partly to the extensor hood and partly to proximal phalanx Four out of the eight cadavers showed asymmetry of the EPB between right and left hands
Brunelli*et al*[5]	Cadaver study	52 hands	19% inserted onto proximal phalanx with most also having attachments to extensor hood 69% inserted into the extensor hood 8% inserted into the base of the distal phalanx 4% were absent
Kulshreshtha *et al*[6]	Cadaver study	44 hands 23 cadavers	25% inserted onto proximal phalanx 25% of tendons insert partly to the base of the proximal phalanx and partly to the extensor hood 2% of tendons inserted entirely into extensor hood 27% of tendons inserted partly to the base of the proximal phalanx and partly to the extensor hood, and from there, continuing further to the base of the distal phalanx with EPL 20% of tendons inserted into the extensor hood and, from there, continued further to the base of the distal phalanx with EPL The EPB was present in all hands, but anatomy of the EPB is variable on the left and right sides of 14 of the 21 paired hands
Alemohammad *et al*[7]	Clinical study and Cadaver study	90 cadaver wrists, and 143 patients undergoing Dequervain's release surgery	In the cadaver group - in 21% pulling on the EPB tendon produced thumb IP joint extension 79% inserted onto proximal phalanx 17% inserted onto distal phalanx 4% inserted onto extensor hood
Shigematsu *et al*[8]	Cadaver study	72 cadaver specimens, 144 hands	29% inserted entirely onto the extensor hood 22% inserted onto the base of the proximal phalanx 19% inserted partly onto the base of the proximal phalanx and partly into the extensor hood 9.0% inserted onto the base of the proximal phalanx and into the extensor hood, and then on the base of the distal phalanx, along with EPL 9.0% inserted onto the extensor hood, and then onto the base of the distal phalanx, along with the EPL 2% were completely absent with no accessory tendon 6% were absent but an accessory tendon inserted at the MP joint 4% had 2 EPB tendons - with variable insertions

EPB: Extensor pollicis brevis; EPL: Extensor pollicis longus.

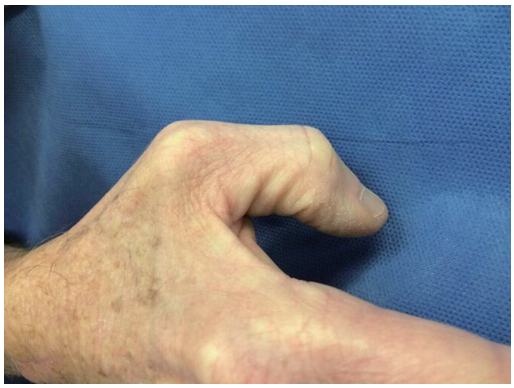

Figure 3 Patient demonstrating thumb MP and IP flexion.

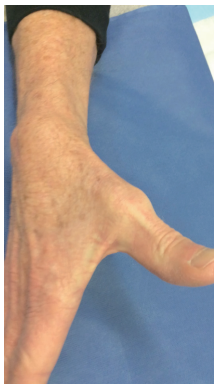

Figure 4 Extensor pollicis brevis tendon extending thumb (Video core tip 1).

extensor tendons. Post-operatively, he was immobilized for 10 d at which time the sutures were removed and he resumed normal activities without pain. At 3 mo follow up he had no complaints of pain and was using

his hand and wrist normally.

DISCUSSION

To the best of our knowledge, virtually all previously reported cases of EPL tendon rupture have been associated with deficits of thumb interphalangeal joint extension. Anatomical dissections have shown the EPB tendon insertion to be highly variable with insertions on the thumb proximal phalanx as well as the extensor hood and the distal phalanx (Table 1). It is, therefore, reasonable to assume that a minority of EPL tendon ruptures may be clinically overlooked owing to the ability of the EPB tendon to fully extend the thumb interphalangeal joint. These cases may never come to the attention of physicians since, while the patients may experience a pop and temporary discomfort, their thumbs may continue to work fairly normally. The cases presenting to orthopedic and hand surgeons, therefore, may represent patients in whom the EPB tendon is unable to fully compensate for the function of the EPL tendon. This case report highlights the anatomical ability of the EPB tendon to fully substitute for the EPL tendon with respect to thumb interphalangeal joint extension. EPL tendon rupture may therefore be more common than is currently appreciated since cases such as these may never present for treatment.

COMMENTS

Case characteristics

A 64-year-old man with rupture of the extensor pollicis longus (EPL) tendon presenting with preserved ability to full extend his thumb at the interphalangeal and metacarpophalangeal joints.

Clinical diagnosis

The EPL tendon was noted to be ruptured both visibly, on examination, on magnetic resonance imaging (MRI) and at the time of surgical exploration, and yet the extensor pollicis brevis (EPB) tendon was able to fully extend the thumb at the IP and MP joints.

Differential diagnosis

The EPB tendon was clearly seen extending the thumb, likely due to its insertion on the extensor hood and/or distal phalanx.

Imaging diagnosis

MRI showed EPL tendon rupture.

Treatment

The prominent bony outgrowth that had caused the EPL rupture was surgically excised so it did not further erode the wrist extensor tendons.

Related reports

Virtually all previously reported series of EPL tendon rupture had deficits of IP thumb joint extension requiring surgery with tendon transfer or grafting. This case report highlights that tendon transfer may not always be required for EPL tendon rupture.

Term explanation

The EPB tendon and the EPL tendon both extend the thumb.

Experiences and lessons

There may be many patients with EPL tendon rupture who do not require tendon transfer due to the ability of the EPB to extend the thumb IP joint.

Peer-review

This is a concise and well written report which is of surgical interest. The video is a useful adjunct.

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DOI: 10.5312/wjo.v7.i7.452

World J Orthop 2016 July 18; 7(7): 452-457
ISSN 2218-5836 (online)

CASE REPORT

Hemorrhagic lumbar synovial facet cyst secondary to transforaminal epidural injection: A case report and review of the literature

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Author contributions: All the authors contributed in outlining the manuscript, gathering the data, and writing the manuscript.

Institutional review board statement: This case report was exempt from the Institutional Review Board.

Informed consent statement: The patient involved in this study gave her written informed consent authorizing use and disclosure of her protected health information.

Conflict-of-interest statement: None of the authors have any financial or any other conflicts of interest that may bias the current study.

Manuscript source: Invited manuscript

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Received: January 25, 2016
Peer-review started: January 26, 2016
First decision: March 1, 2016
Revised: March 12, 2016
Accepted: May 10, 2016
Article in press: May 11, 2016

Published online: July 18, 2016

Abstract

A 64-year-old-female presented with progressive left foot weakness, low back and radicular pain after a left sided S1 transforaminal epidural steroid injection (ESI). Magnetic resonance imaging revealed left side L5-S1 large extradural heterogeneous mass with layering areas suggesting different stages of hematoma formation. Past medical history was significant for peripheral vascular disease and transient ischemic attacks, for which she took aspirin and clopidogrel (antiplatelet agent). These medications were discontinued one week prior to ESI. Although synovial cysts associated with facet arthropathy are common, hemorrhagic cyst is not. To the best of the authors' knowledge, this is the first reported case of symptomatic hemorrhagic lumbar facet synovial cyst following ESI on a patient taking anti-platelet medications.

Key words: Lumbar synovial cyst; Hemorrhage; Anti-platelet therapy; Epidural injection

Core tip: With modern advances in magnetic resonance imaging studies, synovial cysts are seen with degenerative lumbar facet disease. A symptomatic synovial cyst usually presents with a gradual onset low back pain originating from facet arthropathy and radicular pain or neurogenic claudication due to nerve roots compression. Previous reports showed synovial cyst can present with a progressive radicular pain and weakness secondary to spontaneous hemorrhage into the cyst. To the authors' best knowledge, this is the first case report of iatrogenic hemorrhagic lumbar synovial cyst.

Elgafy H, Peters N, Lea JE, Wetzel RM. Hemorrhagic lumbar synovial facet cyst secondary to transforaminal epidural injection: A case report and review of the literature. *World J Orthop* 2016; 7(7): 452-457 Available from: URL: http://www.wjgnet.com/2218-5836/full/v7/i7/452.htm DOI: http://dx.doi.org/10.5312/wjo.v7.i7.452

INTRODUCTION

Synovial cysts are relatively uncommon in the spine. Most cases are associated with facet joint arthritis and degenerative spondylolisthesis and occur most frequently at the L4-5 level. With modern advances in imaging studies, synovial cysts are becoming more evident as a common component of degenerative lumbar facet disease. Normally synovial cysts cause a gradual onset of symptoms that are typically indistinguishable from disc herniation or spinal stenosis[1,2]. The purpose of the current study was to present a case of progressive radicular pain and weakness secondary to hemorrhage into a lumbar synovial cyst after transforaminal epidural injection. To the authors' best knowledge, this is the first case report of iatrogenic hemorrhagic lumbar synovial cyst.

CASE REPORT

A 64-year-old female patient was referred to the spine clinic with three weeks history of increasing low back pain radiating to the posterior aspect of the left thigh and leg. She also complained of a new-onset weakness in her left foot and difficulty with ambulation, which started after a left sided S1 transforaminal epidural steroid injection (ESI) that was performed by pain management physician. Past medical history was significant for hypertension, peripheral vascular disease and transient ischemic attacks, for which she took aspirin and clopidogrel (antiplatelet agent used in coronary artery disease, peripheral vascular disease, and cerebrovascular disease). These medications were discontinued one week prior to ESI. She had a several year history of L4-5 degenerative spondylolisthesis, multilevel disc degeneration and spinal stenosis and radiculopathy that had previously been effectively treated by ESI. Neurological examination revealed positive straight leg raise on the left side, diminished sensation over left L5, S1 nerve root dermatomes, and decreased motor strength (2/5) in extensor hallucis longus (EHL), plantar and dorsiflexion of the ankle on the left.

Plain radiographs showed severe multilevel disc degeneration and a grade I spondylolisthesis at L4-L5. Magnetic resonance imaging (MRI) showed an intraspinal extradural heterogeneous mass with decreased signal centrally within the T2 weighted sequences at L5-S1 on to the left side with displacement of the thecal sac to the right (Figure 1). In STIR images, the lesion appeared heterogeneous but predominantly low in signal intensity and in T1 weighted images, it showed heterogeneous increased signal and layering areas suggesting different stages of hematoma formation (Figure 2). Blood work including erythrocyte sedimentation rate, C-reactive protein, uric acid level, coagulation and platelet studies were within normal limits.

The working diagnosis was a hemorrhagic synovial cyst with a differential diagnosis of sequestrated disc, epidural metastasis and infection. Surgical treatment was indicated due to the increased back, leg pain, progressive left foot weakness and the severity of the spinal canal stenosis. The goal of the surgical treatment was decompression, excision of the lesion and L3-S1 instrumented fusion for the degenerative changes. The patient underwent L3-S1 midline sparing left side laminotomy, facetectomy and foraminotomy. The surgical microscope was used to identify the thecal sac above and below the lesion (Figure 3). Grossly, the lesion appeared to be a hematoma within a synovial cyst (Figure 4). This was impinging upon the transversing L5 and S1 nerve roots on the left side. The intraspinal cyst was then excised without sustaining any dural tear and L3-S1 instrumentation and posterolateral fusion performed for the degenerative lumbar spine (Figure 5). Postoperative histology assessment of the excised lesion confirmed the diagnosis of hematoma within a synovial cyst. At two weeks follow up, the patient's radicular pain had resolved. Additionally, she had improved strength in EHL, plantar and dorsiflexion of the left ankle. Two months post-operatively, the patient regained full strength, denied any radicular or back pain and remained pain free at the last follow up at two years.

DISCUSSION

The most common location of intraspinal synovial cyst is L4-5 motion segment as it is the most mobile level allowing for more degeneration of the facet joint. Symptomatic synovial cysts present with low back pain originating from facet arthropathy and radicular pain or neurogenic claudication due to nerve roots compression[1-3]. The synovial cyst in the current case was located at L5-S1 rather than the most come L4-5 level with presenting symptoms of diminished sensation over left L5, S1 nerve root dermatomes and weakness of EHL, plantar and dorsiflexion of the left ankle. Acute exacerbation of the patient's symptoms was related to hemorrhage inside the cyst. There are previous reports of spontaneous hemorrhage without epidural injection. However, in the current case report, the timing of the presenting symptoms supports the thought that introduction of a needle into a pre-existing synovial cyst had led to subsequent intra-cystic hemorrhage and onset of radicular symptoms and weakness.

To the best of the authors' knowledge, there have been no published reports of hemorrhagic synovial

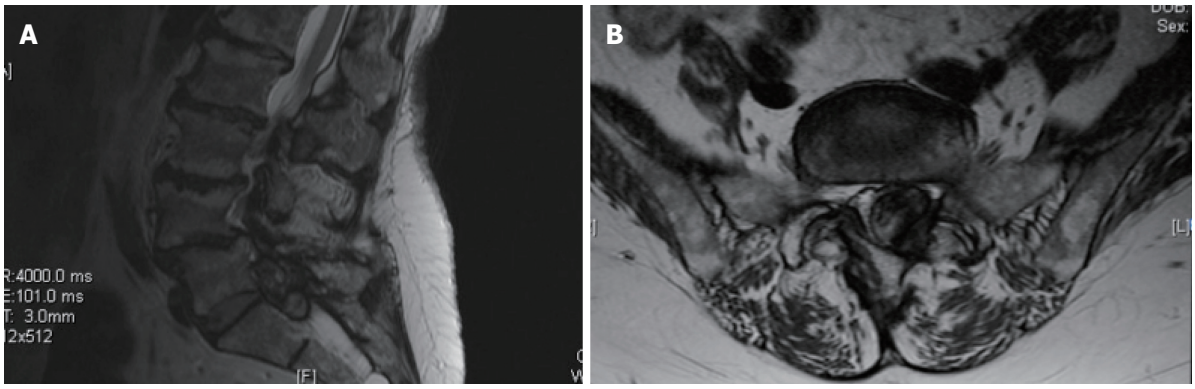

Figure 1 T2 weighted midsagittal (A) and axial (B) magnetic resonance imaging scan showed intraspinal extradural well delineated capsule containing a mixed signal intensity lesion on the left side at L5-S1 causing server canal stenosis with displacement of the thecal sac to the right side.

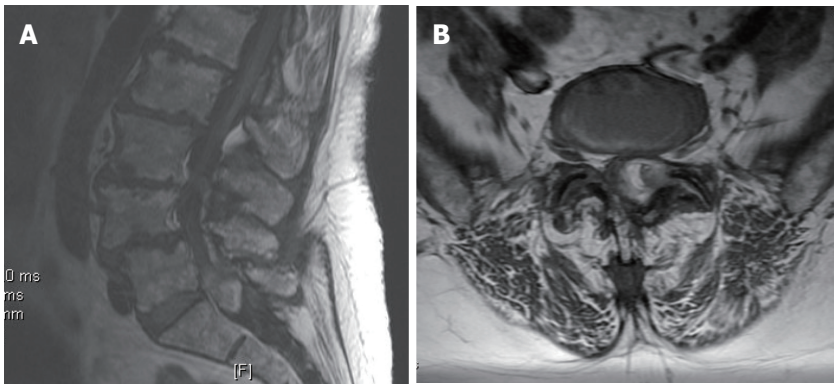

Figure 2 T1 weighted midsagittal (A) and axial (B) magnetic resonance imaging scan showed increased signal intensity with layering areas suggesting different stages of hematoma formation.

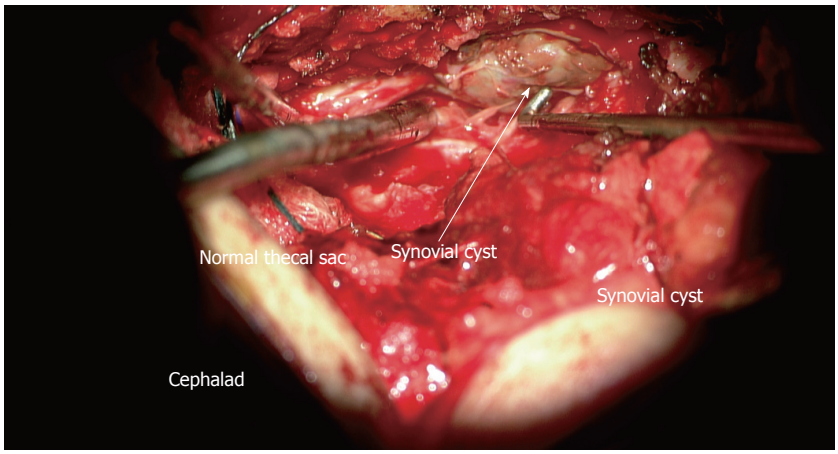

Figure 3 Intraoperative photograph taken by the surgical microscope showed a well-demarked brown color cyst that was adherent to the thecal sac (arrow), a hematoma was found inside the cyst.

cyst following ESI on a patient taking anti-platelet medications. There have been several reported cases of symptomatic epidural hematoma following ESI. The literature reports an incidence of 1:150000 to 1:220000 for epidural hematoma following epidural anesthesia, however the incidence following transforaminal ESI is not quantified[4-6].

MRI is the study of choice for diagnosing, however their radiographic characteristics vary in the literature. Acutely, the cysts have been described as appearing hypo-intense on T1 weighted images while appearing hyper-intense on T2 weighted images. However, in the more commonly imaged subacute period, both the T1 and T2 signals may appear as hyper- or hypo-intense depending on the intracystic methemoglobin and hemosiderin content and the amount of calcification in the

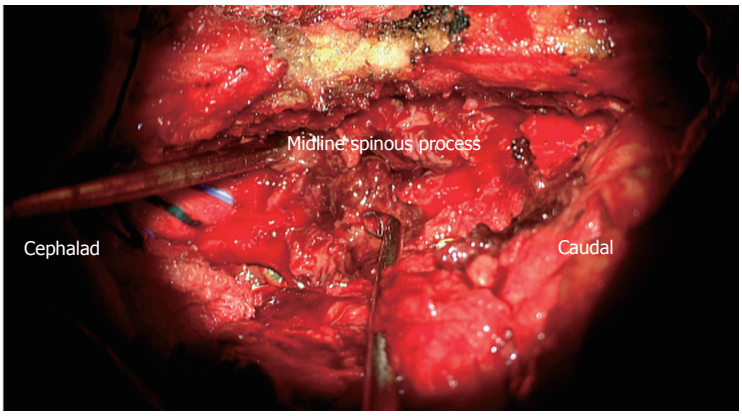

Figure 4 Intraoperative photograph taken by the surgical microscope showed hematoma inside the cyst (arrow).

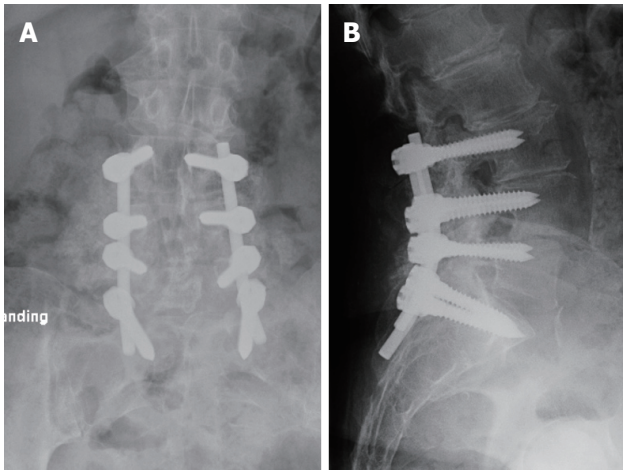

Figure 5 Postoperative anteroposterior and lateral roentgenograms showed L3-S1 instrumentation and posterolateral fusion.

cyst wall[7,8]. The current case showed heterogeneous signals in the T2 and T1 weighted images with the signal predominantly decreased in T2 and increased in T1 with layering areas, which is consistent with the subacute interval between the onset of symptoms and the timing of the MRI.

The mainstay of surgical treatment is excision of the cyst and decompression of the involved neural structures. The role of fusion at the time of cyst excision and decompression remains controversial in patients displaying preoperative spondylolisthesis. The need for fusion is influenced by symptomatic preoperative instability as evidenced by dynamic radiographs or intraoperative instability that may be created by iatrogenic resection of spinal structures such as the pars interarticularis or the facet joints[3]. Epstein *et al*[9] treated 80 patients with cyst excision and decompression without fusion. Thirty-five patients demonstrated preoperative Meyerding grade 1 spondylolisthesis. At two years follow up, 58% of patients who had no evidence of preoperative spondylolisthesis and 63% of patients with preoperative spondylolisthesis demonstrated good or excellent surgeon based outcomes. However, a moderate improvement was noted in only 2 of 8 SF-36 measures. Eleven patients had progression of listhesis and two patients required surgical stabilization. Bydon *et al*[10] in a systemic review noted that 811 patients (84%) underwent decompressive excision alone while 155 patients (16%) had concomitant fusion. They reported that 6.2% of the patients required further operative intervention, the majority of which were for instability and mechanical back pain. Additionally, they found that cyst recurrence occurred in 1.8% of patients who underwent decompression alone while no cases of recurrent cysts were found in patients who underwent fusion.

Surgical excision and decompression of lumbar cysts are not without complication. Banning *et al*[11] reported dural tear in 3 of 29 (9%) patients who underwent laminotomy and cyst excision. Larger cysts or cysts that are calcified due to chronicity can become adherent to the dura making their dissection more complex. Other reported complications include spinal nerve injury, epidural hematoma, seroma, and cyst recurrence[12].

Definitive diagnosis is typically reached by histologic examination. Hemorrhagic synovial cysts are characterized by hematologic, fibrous and endothelial elements. Hemosiderin or blood may be seen depending on the acuity and size of the hemorrhage[7,8,13]. The patient's pathology in the current study demonstrated blood elements, fibrin and synovium consistent with the diagnosis of hemorrhagic synovial cyst.

Anticoagulation has been implicated in the pathogenesis of hemorrhagic cysts. Nourbakhsh *et al*[8] as well as Eck and Triantafyllou, reported cases of radiculopathy secondary to hemorrhagic cyst formation in patients taking warfarin. Both patients required surgical decompression and experienced postoperative resolution of their symptoms[8,13]. Spontaneous spinal epidural hematoma in patients taking anti-platelet medication has been reported in the literature[14-16]. Much research has been directed at spinal anesthesia in patients taking anti-thrombotic or anti-platelet medications. There is no current data that suggest low dose aspirin is associated with increased risk of epidural hematoma in the setting of a normal platelet count. Current guidelines set forth by the American Society of Regional Anesthesia and Pan Medicine call for cessation of clopidogrel 7 d prior to a neuraxial procedure such as

ESI[17,18]. The patient presented in the current report had discontinued the anti-platelet medication in a manner consistent with published guidelines, and coagulation and platelet studies were within normal limits before the transforaminal ESI. However, the timing of the presenting symptoms as well as the radiographic and histologic findings support the thought that introduction of a needle into a pre-existing synovial cyst had led to subsequent intra-cystic hemorrhage and onset of radicular symptoms and weakness.

COMMENTS

Case characteristics

A 64-year-old female presented with progressive left foot weakness, low back and radicular pain after a left sided S1 transforaminal epidural steroid injection (ESI). Past medical history was significant for peripheral vascular disease and transient ischemic attacks, for which she took aspirin and clopidogrel (antiplatelet agent). These medications were discontinued one week prior to ESI.

Clinical diagnosis

Neurological examination revealed positive straight leg raise on the left side, diminished sensation over left L5, S1 nerve root dermatomes, and decreased motor strength (2/5) in extensor hallucis longus, plantar and dorsiflexion of the ankle on the left.

Differential diagnosis

Sequestrated disc, epidural metastasis and infection.

Laboratory diagnosis

Blood work including erythrocyte sedimentation rate, C-reactive protein, uric acid level, coagulation and platelet studies were within normal limits.

Imaging diagnosis

Plain radiographs showed severe multilevel disc degeneration and a grade I spondylolisthesis at L4-L5. Magnetic resonance imaging showed an intraspinal extradural heterogeneous mass with decreased signal centrally within the T2 weighted sequences at L5-S1 on to the left side with displacement of the thecal sac to the right.

Treatment

The patient underwent midline sparing left side laminotomy, facetectomy and foraminotomy. The intraspinal cyst was then excised under the surgical microscope.

Related reports

To the best of the authors' knowledge, this is the first reported case of symptomatic hemorrhagic lumbar facet synovial cyst following ESI on a patient taking anti-platelet medications.

Term explanation

Intraspinal extradural synovial cysts are associated with facet joint arthritis and degenerative spondylolisthesis and occur most frequently at the L4-5 level.

Experiences and lessons

Patients taking anticoagulation or anti-platelet medication should be informed that even if they stop their medication at the time recommended, there is still a possibility of intraspinal bleeding that may result in progressive neurological deficit after transforaminal ESI. Furthermore, transforaminal ESI may be avoided at the level, where a synovial cyst presents, to mitigate such a complication.

Peer-review

The authors present an interesting case of a hemorrhagic synovial cyst developing after a transforaminal epidural steroid injection.

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World J Orthop 2016 July 18; 7(7): 458-462
ISSN 2218-5836 (online)

CASE REPORT

Operative stabilization of the remaining mobile segment in ankylosed cervical spine in systemic onset - juvenile idiopathic arthritis: A case report

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Author contributions: Suhodolčan L gathered data of presented manuscript, made interpretation of data, wrote the majority of manuscript, processed figures, predominantly involved in designing and drafting manuscript; Mihelak M gathered literature regarding manuscript, participated in presenting data and designing manuscript body; Brecelj J made clinical examination and follow-up visits, made all surgical interventions on patient's lower limbs previous to surgical intervention presented in this manuscript, made substantial contribution in designing manuscript, participated coordination and helped to draft the manuscript; Vengust R made the surgical intervention presented in this manuscript, has been involved in drafting the manuscript and revising it critically for important intellectual content.

Institutional review board statement: This case report was exempt from the Institutional Review Board standards at University of Ljubljana, Slovenia.

Informed consent statement: Patient gave informed consent.

Conflict-of-interest statement: No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

Manuscript source: Invited manuscript

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Received: January 21, 2016
Peer-review started: January 22, 2016
First decision: March 24, 2016
Revised: April 4, 2016
Accepted: May 31, 2016
Article in press: June 2, 2016
Published online: July 18, 2016

Abstract

We describe a case of a 19-year-old young man with oligoarthritis type of juvenile idiopathic arthritis, who presented with several month duration of lower neck pain and progressive muscular weakness of all four limbs. X-rays of the cervical spine demonstrated spontaneous apophyseal joint fusion from the occipital condyle to C6 and from C7 to Th2 with marked instability between C6 and C7. Surgical intervention began with anterolateral approach to the cervical spine performing decompression, insertion of cage and anterior vertebral plate and screws, followed by posterior approach and fixation. Care was taken to restore sagittal balance. The condition was successfully operatively managed with multisegmental, both column fixation and fusion, resulting in pain cessation and resolution of myelopathy. Postoperatively, minor swallowing difficulties were noted, which ceased after three days. Patient was able to move around in a wheelchair on the sixth postoperative day. Stiff neck collar was advised for three months postoperatively with neck pain slowly decreasing in the course of first postoperative month.

On the follow-up visit six months after the surgery patient exhibited no signs of spastic tetraparesis, X-rays of the cervical spine revealed solid bony fusion at single mobile segment C6-C7. He was able to gaze horizontally while sitting in a wheelchair. Signs of myelopathy with stiff neck and single movable segment raised concerns about intubation, but were successfully managed using awake fiber-optic intubation. Avoidance of tracheostomy enabled us to perform an anterolateral approach without increasing the risk of wound infection. Regarding surgical procedure, the same principles are obeyed as in management of fracture in ankylosing spondylitis or Mb. Forestrier.

Key words: Juvenile idiopathic arthritis; Cervical ankylosis; Spastic tetraparesis; Multilevel both column fixation; Unstable cervical segment

Core tip: The spontaneous cervical apophyseal joint fusion is rare and only seen in juvenile type rheumatoid arthritis, where spontaneous fusions of more than three cervical segments are extremely uncommon. We present a patient with fusion of all but C6-C7 level. The single mobile segment became highly unstable, producing mechanical pain with symptoms of myelopathy. Prior to the surgery, awake fiber-optic intubation was used. Surgical intervention began with the anterolateral approach to the cervical spine performing decompression, insertion of cage and anterior vertebral plate and screws, followed by posterior approach and fixation. Care was taken to restore sagittal balance.

Suhodolčan L, Mihelak M, Brecelj J, Vengust R. Operative stabilization of the remaining mobile segment in ankylosed cervical spine in systemic onset - juvenile idiopathic arthritis: A case report. *World J Orthop* 2016; 7(7): 458-462 Available from: URL: http://www.wjgnet.com/2218-5836/full/v7/i7/458.htm DOI: http://dx.doi.org/10.5312/wjo.v7.i7.458

INTRODUCTION

Juvenile idiopathic arthritis (JIA) is an exclusion diagnosis that applies to any arthritis of unknown origin, persisting for more than 6 wk and with the onset before the age of 16 years[1]. Patients who belong to oligoarthritis type of JIA are characterized by an asymmetric arthritis affecting mainly large joints of an early onset (before 6 years of age), a female predilection, a high frequency of positive antinuclear antibodies, a high risk for developing chronic iridocyclitis and consistent HLA associations, which is typical of children and is not seen in adults[2,3].

Recent advances in medical treatment enable more favorable prognosis, but rapid progressing cases may result in severe complications such as joint deformities, joint ankylosis, osteoporosis and decreased bone mass. The common clinical manifestation of spinal involvement is the instability of upper cervical spine. The spontaneous cervical apophyseal joint fusion is rare and only seen in juvenile type rheumatoid arthritis, where one-or two level fusions occur in up to 41 percents of cases[4]. On the other hand, spontaneous fusions of more than three cervical segments are reported to be extremely rare (Table 1)[5].

We report a case of spontaneous cervical ankylosis from the occiput to C6 and from C7 to Th2 with subjacent instability at C6-C7 level and neurological deficit in patient with JIA, which has been operatively stabilized.

Table 1 Characteristics of first-reported case of long-segment cervical fusion[5]

Patient characteristics
27-yr-old female
JIA for 10 yr
Symptoms
Posterior neck pain
Progressive weakness
Paresthesia and dysesthesia below the C7 dermatome
Imaging
Spontaneus apophyseal joint fusion from the occipital condyle to C6
Instability at C6/C7 level - only mobile segment
Problems during procedure
Contracture of temporo-mandibular joint
Endotracheal intubation not feasible due to stiff neck
Tracheostomy needed
Severe osteoporosis noted
Approach/type of fixation
Posterior approach
Decompressive subtotal laminectomy
Interspinous process wiring with titanium-braded cable and addition of integrated bone graft at C6/7 level
Results
Pathologic reflexes disappeared
Well established bone graft incorporation 6 mo post-op
Conclusion
First-reported case of long-segment cervical fusion, may be related with rapid progression of the disease

JIA: Juvenile idiopathic arthritis.

CASE REPORT

A 19-year-old young man with oligoarthritis type of JIA presented with several month duration of lower neck pain and progressive muscular weakness of all four limbs. He had been treated previously for major contractures of the hip, knee and ankles. The valgus of the both knees was corrected with a temporary hemiepiphysiodesis at the age of 12. At the age of 15 an elongation of Achilles tendon and the knee flexors were performed. Total hip replacement was performed at the age of 15 on the left side and a year later on the right side. He has been partly wheelchair-bound for 3 years before admission due to extensive contractures of hips, knees and ankles. But he was able to walk short distances with the help of crutches. At the age of 12,

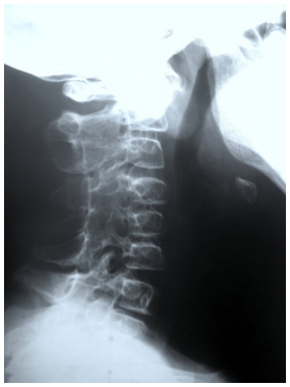

Figure 1 Plain X-rays at the age of 12 of cervical spine showed loss of lordosis and posterior fusion from C2 to C6 joints.

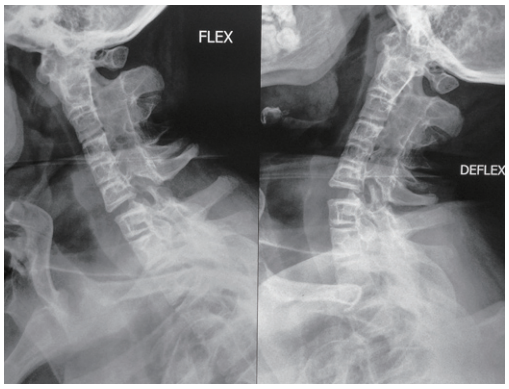

Figure 2 Bending films on admission showed spontaneous apophyseal joint fusion from the occipital condyle to C6 and from C7 to Th2 with marked instability between C6 and C7.

pain in the cervical spine occurred for the first time, followed by occasional mild pain episodes. The patient had been treated with methotrexate, steroids and antihypertensive therapy.

Clinical examination revealed markedly limited mobility of cervical spine with rotations being completely blocked. Flexion and extension resulted in significant pain in lower cervical spine. Patient was unable to walk independently, with signs of spastic tetraparesis.

Loss of lordosis and sclerotic changes of the facet joints in cervical plain X-rays had been present 7 years before admission (Figure 1). X-rays of the cervical spine on the day of the admission demonstrated spontaneous apophyseal joint fusion from the occipital condyle to C6 and from C7 to Th2. Bending films showed marked instability between C6 and C7 (Figure 2). There were no rentgenographic abnormalities of thoracolumbar spine. Computerized tomography/angiography confirmed apophyseal fusion of the entire cervical spine except for the C6-C7 level, where the left vertebral artery was dominant. Spinal canal was narrowed at C6-C7 due to posteriorly protruding spondylophites. MRI revealed C6-C7 disc degeneration with spinal canal stenosis but no signal changes in spinal cord.

The patient underwent surgical intervention because of progredient spastic tetraparesis. Surgery was started after awake fiberoptic intubation. We began the procedure using anterolateral approach to the spine, where discectomy and osteophyte removal was performed, followed by the insertion of cage filled with bone substitute at the C6-C7 level. Vertebral bodies C5, C6 were fixed to C7, Th1 using anterior vertebral plate and screws (Figure 3).

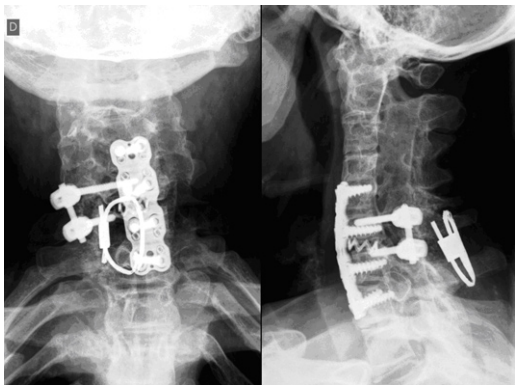

Figure 3 Post-operative plain X-rays.

Later on, the patient was turned prone, and cervical spine was accessed using the posterior approach. Posterior fixation was obtained using unilateral pedicular screws combined with an interspinous wire. Pedicular screws were inserted on the right side only, due to very small caliber vertebral artery ipsilaterally. Bone substitute was placed to the decorticated C6-C7 apophyseal joints. Care was taken to fix the spine in a position, which would enable the patient to gaze horizontally in a sitting position (Figure 4).

After the surgery patient had mild swallowing difficulties, which ceased after three days. He was able to move around in a wheelchair on the sixth postoperative day. Stiff neck collar was advised for three months postoperatively with neck pain slowly decreasing in the course of the first postoperative month. On the follow-up visit six months after the surgery the patient exhibited no signs of myelopathy, X-rays of the cervical spine revealed solid bony fusion at C6-C7 level. He was able to gaze horizontally while sitting in a wheelchair.

DISCUSSION

JIA of the spine was once believed to be a benign disease process. Many studies have shown, however, that the spectrum of joint erosion, deformity, ensuing neurological deficit, and underlying pathologic mechanisms warrant concern, especially in cervical spine[6-8]. The most common pathologic condition is a spontaneous posterior cervical fusion seen in the late stage of the disease. Fusion usually occurs in one or two segments without clinical consequences[5]. Apart from this, our patient presented with fusion of all but one level of cervical spine. The single mobile segment became highly

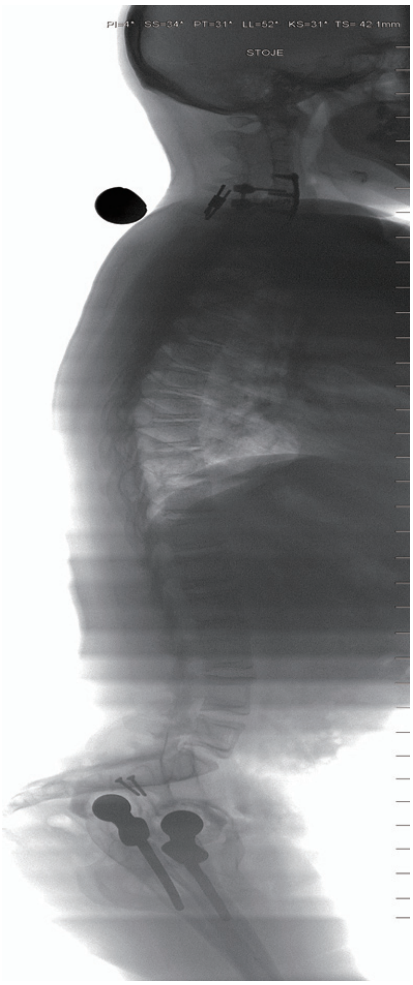

Figure 4 Post-operative scan; showing restored sagittal alignment.

unstable over time and produced mechanical pain, which along with symptoms of myelopathy forced the patient to consult a physician. The clinical course of progressive stiffness with escalating localized pain is unique in childhood and only resembles the pseudoarthrosis in ankylosing spondylitis in adults[9,10].

Regarding surgical procedure the same principles were obeyed as in management of fracture in ankylosing spondylitis or Mb. Forestier[11,12]. Multilevel both column fixation was performed employing combined anterolateral and posterior approach. Signs of myelopathy with stiff neck and single movable segment raised concerns about intubation, but were successfully managed using awake fiber-optic intubation. Avoidance of tracheostomy enabled us to perform the anterolateral approach without increasing the risk of wound infection. To the best of our knowledge, this is the first-reported case of multisegmental cervical ankylosis in JIA, operatively managed with combined anterior and posterior spinal interbody fusion.

As with other cases of stiff cervical spine, special care has to be taken to achieve proper sagittal alignment[13]. With careful preoperative planning fixation should be achieved, which suits patient best in terms of optimal gaze.

We present a JIA patient with a multisegmental spontaneous fusion of cervical spine. The one segment remaining mobile became overtly instable causing mechanical neck pain and progressive spastic tetraparesis. The condition was operatively managed with multisegmental, both column fixation and fusion, resulting in pain cessation and resolution of myelopathy symptoms six months postoperatively.

COMMENTS

Case characteristics

A 19-year-old young man with oligoarthritis type of juvenile idiopathic arthritis presented with several month duration of lower neck pain and progressive muscular weakness of all four limbs.

Clinical diagnosis

Clinical examination revealed markedly limited mobility of cervical spine with rotations being completely blocked. Flexion and extension resulted in significant pain in lower cervical spine. Patient was unable to walk independently, with signs of spastic tetraparesis.

Differential diagnosis

Ankylosing spondylitis, Mb. Forestier, Diffuse idiopathic skeletal hyperostosis.

Laboratory diagnosis

All labs were within normal limits.

Imaging diagnosis

X-rays of the cervical spine on the day of the admission demonstrated spontaneous apophyseal joint fusion from the occipital condyle to C6 and from C7 to Th2. Flexion and extension X-rays showed wide apophyseal joint space between C6 and C7, suggesting instability and excessive mobility. Computerized tomography confirmed ankylosis at the C0-C1 level.

Pathological diagnosis

Juvenile idiopathic arthritis.

Treatment

Surgical intervention with the anterolateral approach to the cervical spine, performing decompression, insertion of cage and anterior vertebral plate and screws, followed by posterior approach and fixation.

Related reports

Rapid progressing cases of juvenile idiopathic arthritis may result in severe complications such as joint deformities, joint ankylosis, osteoporosis and decreased bone mass. The common clinical manifestation of spinal involvement is the instability of upper cervical spine. The spontaneous cervical apophyseal joint fusion is rare and only seen in juvenile type rheumatoid arthritis, where one- or two level fusions occur in up to 41 percents of cases. Spontaneous fusions of more than three cervical segments are reported to be extremely rare.

Term explanation

Juvenile idiopathic arthritis (JIA) is an exclusion diagnosis that applies to any arthritis of unknown origin, persisting for more than 6 wk and with the onset before the age of 16 years.

Experiences and lessons

The single mobile segment of multisegmental cervical ankylosis in JIA that produces mechanical pain and neurologic deficit should be operatively managed. Awake fiber-optic intubation can be used prior to the surgery to overcome concerns of stiff neck. Avoidance of tracheostomy enables combined anterior

and posterior spinal interbody fusion without increasing the risk of wound infection. Care should be taken to restore sagittal balance.

Peer-review

It is a very good paper.

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P- Reviewer: Firstenberg MS, Jiang B, Kettering K, Najafi M, Peteiro J, Ueda H **S- Editor**: Qiu S **L- Editor**: A **E- Editor**: Wu HL

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