ISSN 0008-3194 (p)/ISSN 1715-6181 (e)/2019/171–177/$2.00/©JCCA 2019

Clinical presentation of an adolescent female synchronized swimmer with a simple bone cyst in the proximal humerus: a case report

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Objective: *This case raises awareness for healthcare practitioners who may not suspect a simple bone cyst (SBC) in a pediatric athlete with a high risk of pathological fracture.*

Case summary: *SBC's are often considered as asymptomatic lesions which are commonly found incidentally on plain film radiographs. Presented here is a case of a 14-year-old competitive synchronized swimmer with a SBC in the proximal humerus with no MRI evidence of soft tissue pathology or pathological fracture, presenting clinically with refractory posterior shoulder pain.*

Summary: *Early detection of a SBC in a pediatric athlete is essential so an interdisciplinary care approach can be employed to ensure the appropriate management in those where there is high risk of pathological fracture. Clinicians should be aware of the risk factors for the*

Objectif : *Ce cas sensibilise les professionnels de la santé qui ne soupçonnent pas la présence d'un kyste osseux essentiel (KOE) chez un jeune athlète présentant un risque élevé de fracture pathologique.*

Résumé du cas : *Les KOE sont souvent considérés comme des lésions asymptomatiques que l'on trouve souvent par hasard sur les radiographies ordinaires. On présente ici le cas d'une adolescente âgée de 14 ans pratiquant la natation synchronisée, souffrant d'un KOE dans l'humérus proximal et présentant cliniquement une douleur réfractaire à l'épaule postérieure. L'IRM n'a pas mis en évidence de pathologie des tissus mous ni de fracture pathologique.*

Résumé : *La détection précoce d'un KOE chez un jeune athlète est essentielle pour qu'une approche de soins interdisciplinaires puisse être mise en œuvre afin d'assurer une prise en charge appropriée chez ceux qui présentent un risque élevé de fracture pathologique. Les cliniciens doivent être conscients des facteurs de risque*

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The authors have no disclaimers, competing interests, or sources of support or funding to report in the preparation of this manuscript.
The involved patient provided consent for the use of clinical information and imaging with respect to this case report.
The CMCC Research and Ethics Board have approved this case report.

development of a pathological fracture, and for other potential complications that may arise thereafter in those with a SBC.

(JCCA. 2019;63(3):171-177)

KEY WORDS: chiropractic, benign tumour, simple bone cyst, swimmer, synchronized

de fracture pathologique et des autres complications potentielles qui peuvent survenir par la suite chez les personnes atteintes d'un KOE.

(JCCA. 2019;63(3):171-177)

MOTS CLÉS : chiropratique, tumeur bénigne, kyste osseux essentiel, nageuse, synchronisée

Introduction

A simple bone cyst (SBC), also referred to as a unicameral bone cyst, is the most common benign tumour that is seen in children and adolescents (up to 85% of all cases).[1,2] These bone cysts represent approximately 3% of all tumours of the bone and have been reported to occur more frequently in boys than girls (2:1).[2] The average age of diagnosis is nine years, with a peak incidence between the ages of three and 14 years.[1,2] They are cystic, fluid filled cavities that can develop in tubular and flat bones, most typically found in the proximal humerus and femur, which account for greater than 90% of all cases.[1–4] These cysts do have a tendency to expand and weaken the bone they reside in, despite not being a true neoplasm.[1]

A definite pathogenesis of a SBC to this day remains controversial. The most widely accepted etiology is a venous outflow obstruction in the cancellous bone causing an increase in intraosseous pressure. This increased pressure results in elevation of inflammatory protein markers released by endothelial cells along the wall of the cyst, causing subsequent osteoclastic activation and bone resorption.[2–5] Other suggested mechanisms of pathogenesis include: a disturbance in local bone growth, intramedullary (post-traumatic) hemorrhage, and trapped intraosseous nests of synovial cells.[2]

Although SBC's are common and often considered asymptomatic[2,6], it has been reported that approximately two thirds of these lesions will at some point endure pathological fracture[7]. This should be of particular concern in the pediatric athletic population due to the increased load and torque forces that may be placed through the humerus.[8] This suggests that even in a non-contact scenario, the humerus may be at risk for fracture in the presence of a cyst.[8] In sports where body contact with others is more probable, this risk of fracture is only expected to increase. This raises further concern in such sports particularly if early detection has not been made.

We present an atypical clinical presentation of an adolescent competitive synchronized swimmer who experienced refractory shoulder pain despite a lack of MRI evidence of soft tissue pathology or pathological fracture of a SBC in the proximal humerus. We will also highlight possible complications and risk factors associated with SBC to raise awareness of a condition that may possibly go undiagnosed in young athletes.

Case presentation

A 14-year-old female, competitive synchronized swimmer presented with a four day history of localized, right-sided, posterior shoulder pain sustained while performing a manoeuvre referred to as sculling. Sculling is an action in synchronized swimming, whereby the swimmer treads water solely with their arms, while inverted as they are performing out of water movements with their legs. This action involves heavily resisted and repetitive internal and external rotation of the shoulders (Figure 1).

Upon presentation, the patient complained of a dull ache in the posterior shoulder at rest, which became sharp when she performed sculling. The patient did not report any previous shoulder injuries in her past. On examination, there was full, pain-free right shoulder range of motion (ROM) with no signs of swelling, erythema, or bony malalignment. Empty can, Hornblower's test and manual muscle testing of external rotation strength at zero degrees shoulder abduction were positive for pain and were rated 4/5 on the right for muscle strength. Palpation of the infraspinatus and teres minor insertions at the posterior aspect of the humeral head reproduced the patient's chief complaint. An evaluation of generalized joint hypermobility (GJH) using the Beighton Test[9] revealed a

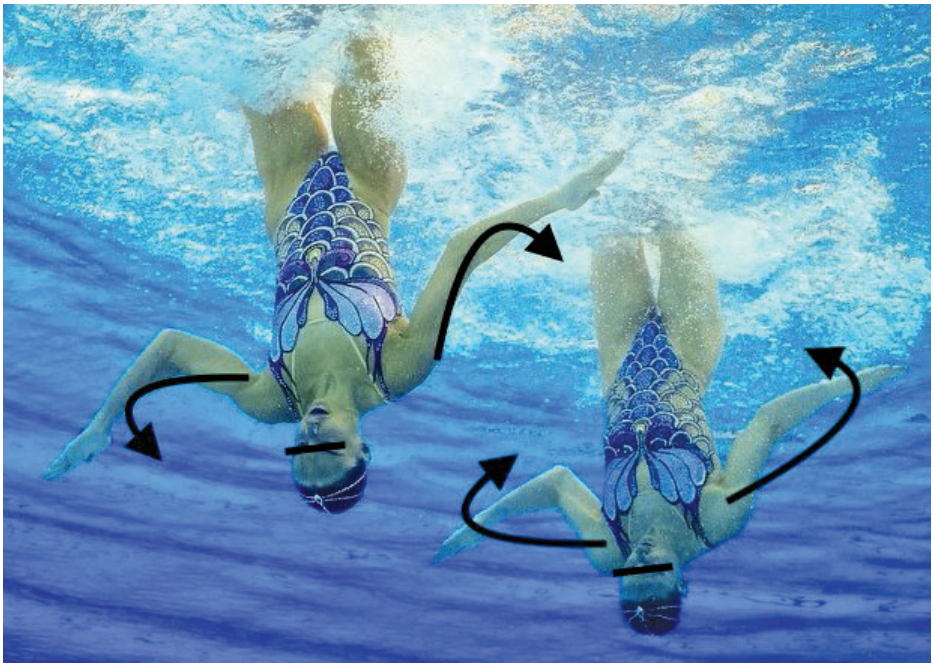

Figure 1.
Sculling is the underwater action used by synchronized swimmers to hold position and propel their bodies in different directions while inverted by repetitively and forcefully internally and externally rotating their shoulders. This is similar to treading water with your legs, known as "egg-beating".

score of 3/9 (+ bilateral hyperextension of 5th metacarpophalangeal joint >90° & + palm of hands touching flat on the ground when knees extended).

An assessment of shoulder instability using the Feagin Test, Load and Shift Test or sulcus sign was not made. Cervical spine range of motion was full and pain free, and foraminal encroachment tests of the cervical spine were negative. Sensation and motor testing of the C5-T1 levels were within normal limits bilaterally and deep tendon reflexes of C5-7 were 2+ bilaterally.

The patient was given a clinical diagnosis of an acute, right-sided rotator cuff tendinopathy and a six-week conservative plan of management consisting of Active Release Therapy (ART®) and a progressive rotator cuff and scapular stabilizer strengthening rehab protocol was implemented. The patient was treated with this approach for 9 visits over 6 weeks. Following this treatment plan, the patient showed minimal clinical change in her symptomatology and was unable to perform sculling due to pain.

A radiograph of the right shoulder was ordered (Figure 2) which revealed a well-defined, osteolytic lesion. Differential diagnoses based off the radiographic findings included: a SBC, aneurysmal bone cyst (ABC), and a chondroid lesion, such as an enchondroma. The patient was

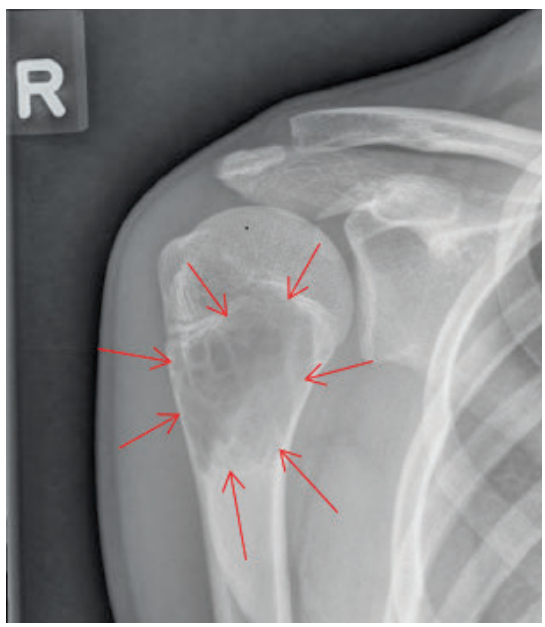

Figure 2.
AP plain film radiograph of the right shoulder revealing a well-defined, slightly expansile, osteolytic lesion (outlined by arrows), located centrally within the metaphysis abutting the epiphyseal growth plate with no signs of periosteal reaction, fracture, or soft tissue mass.

immediately referred to an orthopedic surgeon with a specialty in bone tumours of the upper extremity, who sent the patient for a magnetic resonance imaging (MRI) of the right shoulder (Figure 3) which demonstrated a slightly expansile, T2 hyper-intense and T1 hypo-intense intramedullary unilocular cystic lesion in the proximal humeral metaphysis causing endosteal scalloping and cortical thinning with no evidence of fracture. All rotator-cuff and long head of biceps tendons appeared to be intact with no sign of pathology. An orthopedic surgeon confirmed the diagnosis of a SBC based off the aforementioned MRI features which are pathognomonic for this lesion, along with the consideration of the patient's age which is also consistent with its clinical presentation. One possible inconsistency with the clinical presentation of this suspected SBC is that they are not typically painful.

At this point in time (six to eight weeks post initial presentation), the patient was still experiencing intermittent, refractory dull, achy pain along the posterior shoulder, despite her diligence with pain-free rehabilitation exercises, and avoiding all aggravating factors including synchronized swimming. The orthopedic surgeon handling the patient's case prescribed a conservative "wait and see" approach which simply involved a follow-up up MRI, activity restriction, and to continue rehab for the rotator cuff and scapular stabilizers at a recommended physiotherapy clinic.

The patient was contacted a year later, reporting that she had stopped participating in synchronized swimming. She has no pain at rest or during any of her activities of daily living. By this point, the patient decided to not follow-up with any radiographs and/or MRI, and as such, the status of the SBC is currently unknown. Any further follow-up with the patient was lost as she no longer attends the clinic where the assessment and treatments were conducted.

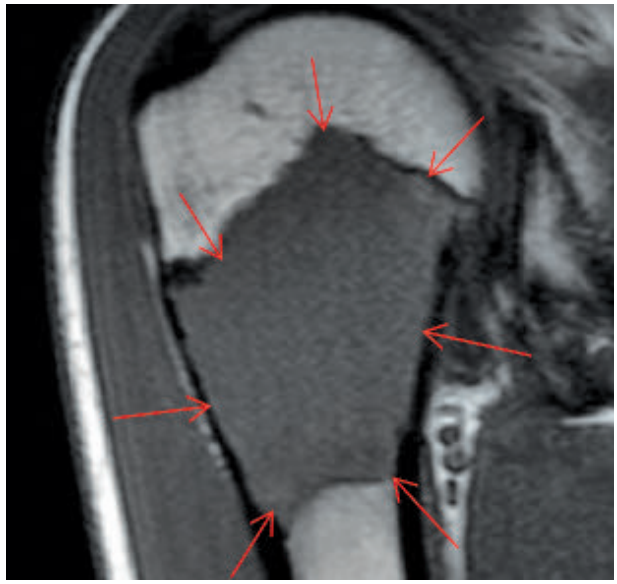

Figure 3.
T1 weighted, coronal slice MRI of the right shoulder revealing a 4.5 x 3.6 x 3.3 cm slightly expansile, hypointense intramedullary unilocular cystic lesion (outlined by arrows), in the proximal humeral metaphysis causing endosteal scalloping and cortical thinning with no evidence of fracture.

Discussion

The diagnosis of a SBC can normally be made with the use of plain film radiographs, however, confirmation can be made with advanced imaging (such as MRI) in cases where there is a strong resemblance to other benign bony lesions. An example of such a lesion, and a strong differential diagnosis is an ABC, which is at higher risk of gross expansion and pathological fracture.[1] Although generally not as expansile as an ABC, SBC's have the capability to expand and thin the cortex that it surrounds. This may compromise the cortex possibly resulting in a pathological fracture, which is not uncommon in the athletic population.[10] SBC's also reside centrally within the medullary cavity, unlike ABC's which have a more eccentric presentation. The more central the location, the smaller the likelihood of fracture risk and soft tissue involvement.[2] Ten percent of those that fracture will present with a fallen fragment sign on plain film radiographs, which represents a small portion of the fractured bone which can separate and sink to the bottom of the cyst.[2,11]

With that said, it is important to understand risk factors which may further increase fracture risk. A retrospective study by Urakawa *et al*.[12] identified 155 subjects with a SBC, 141 of which exceeded six-month follow-up. The majority of subjects (35%) had a SBC which resided in the proximal humerus, followed by five other locations in the lower extremities. In this study, a multivariate analysis revealed that ballooning of the bone, a cyst located in a long bone, male sex, and multilocular cysts were signifi-

cant risk factors in increasing the incidence of pathological fractures.[12] Kaelin and MacEwan[13] proposed a cyst index formula (cyst area divided by the diaphyseal diameter^2) and cut-off score to help determine fracture risk. A cyst index greater than 3.38 was predictive of pathological fracture in this particular study. Similar studies have reported other risk factors associated with increased incidence of pathological fracture, which include: a high cyst index, high percentage of bone occupied by the cyst in the transverse plane, thin cortical thickness, and those active phase cysts residing in the upper limb.[13–16]

Furthermore, active phase cysts are a potential risk factor for fracture. It has generally been accepted throughout the literature that cysts residing within 1cm of the growth plate are considered active[2], minimizing the amount of normal bone between them. Haidar *et al*.[17] underwent a retrospective analysis whereby a measurement of two centimeters from the growth plate was determined as the cut-off point between active versus latent cysts. The destined progression in the natural history of the cyst is to migrate down into the diaphysis through endochondral ossification where it eventually becomes latent (non-active). These cysts are not commonly seen in adults, owing to the fact that they spontaneously fill and ossify on their own sometime after skeletal maturity is reached. Since SBC's are typically asymptomatic in the absence of pathological fracture, there is no inquiry for imaging and thus go undetected during pre-adulthood prior to ossification, making natural history difficult to document.[2,3,18]

Analysis of the present patient's SBC on MRI (Figure 2) reveals many of the risk factors mentioned above. The cyst's location is in a long bone, it is showing properties of expansion (ballooning), a high percentage of the bone is occupied by the cyst in the transverse plane, there is presence of thinning cortex, and it clearly appears to be in an active phase, as it's shown abutting the epiphyseal growth plate. Furthermore, using the equation proposed by the Kaelin and MacEwan[13], the patient had a cyst index well above the cut-off score (4.5cm x 3.6cm = 16.2/1.32cm^2 = 9.30). The only fracture risk factors that do not appear to exist in the present patient's case are the fact that the cyst is not multilocular in nature, and she is not of male sex. It was also described in the study by Urakawa *et al*.[12] that males in the study were more likely to be physically active, which may be the main contributor to the increased incidence of fracture within this cohort. The presence of multiple risk factors suggest that the present patient is of high risk of developing a pathological fracture.

As for management, Urakawa *et al*. suggested that high risk cysts should either be considered for surgery, or at the very least, careful observation.[12] Due to the fact that SBC's are self-limiting and have a tendency to heal without any intervention, it is common to adopt a more conservative plan of management, especially in children who have asymptomatic, latent cysts.[5] Educating and restricting activity in those athletes with a SBC who are at high risk of fracture should be of utmost importance in those who are following this conservative approach. In a recent survey of members of the European Pediatric Orthopedic Society (EPOS) and Pediatric Orthopedic Society of North America (POSNA)[19] surgery was highly recommended only in those children with symptomatic SBC's under 10 to 12 years of age (by 71% of members) and in those children with asymptomatic, high risk of fracture SBC's (by 53% of members). The most commonly reported surgical options used include: curettage with or without bone grafting, percutaneous decompression, corticosteroid injections, or autologous bone marrow injections. It is important to note that many of these interventions have high rates of recurrence, with improved outcomes seen with cysts measuring >2cm from the growth plate.[19]

As a clinician working with pediatric athletes, not only is it important to detect the presence of a bone cyst, or any other bony lesions, it is also important to understand the possibility or risk of other complications that may arise. In the presence of a pathological fracture, one such potential complication is the development of avascular necrosis (AVN) of the humeral head. Urakawa *et al*. reported one subject with a femoral neck SBC who developed AVN after a displaced fracture.[12] There are other reports in the literature that have described similar instances of AVN of the femoral head as a complication after pathological fracture.[12] Brooks *et al*. specify that AVN of the humeral head may result after either displaced fractures, or fracture dislocations.[20] Humeral head AVN has not been reported in SBC cases, likely due to the fact that, to the authors' knowledge, there presently is no report of displaced humeral SBC pathological fractures in the literature.

Another possible complication is growth arrest, especially in those active cysts which are likely communicating with the epiphyseal growth plate. Although extremely

rare, growth arrest is a possibility, especially when a displaced femoral neck fracture is involved. Other possible mechanisms for growth arrest may include: the hypothetical instance where a pathological fracture involves the disruption of the growth plate, or through accidental damage of the growth plate through invasive surgical techniques.[5] Fortunately, there are no reported cases of malignant transformation of SBCs.[3]

It is not entirely understood as to the source of the present patient's pain. SBC's that have not fractured are typically asymptomatic, however, there is suggestion in the literature that pain could arise from fissuring of the cyst throughout its growth and migration.[1,3] It is not known if this bears any relationship to endosteal scalloping which was reported in the patient's imaging. Given that there was no evidence of other shoulder pathology in the patient's MRI, it is plausible that this fissuring mechanism may have played a factor in the patient's experience of refractory shoulder pain. It is also important to note that the patient's pain was aggravated by certain movements and can be elicited with palpation of muscular structures, which would suggest either pain of musculo-tendinous origin or local hyperalgesia as a compensatory response from the cyst. Entertaining the idea of pathology present within the surrounding musculo-tendinous structures (i.e rotator cuff) as a source of the patient's pain may still be warranted considering the biomechanical joint stresses involved with certain sport-specific movements, such as sculling. Although rotator cuff tears in the general pediatric population account to less than 1% of all total rotator cuff tears[21], those who participate in repetitive and large rotational torqueing movements of the glenohumeral joint increase the risk of rotator cuff tears through tensile shearing forces[21]. Furthermore, there is always the possibility for false-negative MRI results considering the sensitivity of detecting tendinopathy and partial tears. A meta-analysis identified the pooled sensitivity for detecting partial rotator-cuff tears and tendinopathy (0.67-0.83) is much lower than for full tears (>0.90) using either MRI or ultrasound (US).[22] Having said that, it is plausible to assume that the patient may have suffered from rotator cuff pathology despite a normal MRI. Another explanation to her musculoskeletal pain should include the possibility of GJH resulting in instability of the humeral head within the glenoid. Competitive swimmers may have this propensity to hypermobility[23] and resultant glenohumeral subluxation which may negatively influence adjacent soft tissue structures (i.e. rotator cuff) through repetitive abutment. There seems to be a positive correlation between GJH, measured using the Beighton Test[9], and an increased active horizontal shoulder abduction (AHSA) score in 10 to 15 year old competitive swimmers[23]. The patient in this case scored a 3/9 on the Beighton Test, suggesting an absence of GJH (+ score is 4/9 or >).[9] MRI of the patient's shoulder also revealed insignificant soft tissue injury findings (no presence of edema, tendinopathy or tear). Despite these results, assessing for GJH is an important aspect of the physical exam in athletes who participate in activities which require excessive and repetitive joint ROM. Perhaps one factor as to why there appears to be a strong correlation between GJH and an increased AHSA in pediatric competitive swimmers is genetic selection. Genetic variability may make it easier for some individuals to participate in sports which require certain sport-specific movements, particularly at a high level which may pose as an injury risk. Instead of focusing on soft tissue related diagnoses (i.e rotator cuff tendinopathy), it may be beneficial to shift the clinical focus to movement-related deficiencies in those who are hypermobile. From a rehabilitation perspective, a rationale to compensate for deficits in the passive structures surrounding the glenohumeral joint is to retrain the dynamic shoulder stabilizers in order to maintain glenohumeral joint centration.[24]

As a treating practitioner, early detection of SBCs is central to their management. When identified early, decisions relating to activity restriction (or elimination), and early referral to an orthopedic surgeon can be undertaken. This will help to not only decrease the likelihood of developing a pathological fracture, but will also allow for the development of a plan of management that will be more conducive to treating structures around a weakened bone. That being said, practitioners must be cognizant when treating soft tissue structures surrounding the cyst, in order to help decrease the risk of iatrogenic fracture which could plausibly be caused by manual therapy.

Summary

This case highlights the importance of early detection of a SBC in a pediatric synchronized swimmer. She presented with symptoms consistent with rotator cuff pathology, but was determined to have a SBC that was classified as a potential high risk for pathological fracture. As illustrated

by this case, it is important to determine the risk factors for the development of a pathological fracture, and for other potential complications that may arise thereafter in those with a SBC.

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