SYMPTOMATIC JUXTAPHYSEAL ANEURYSMAL BONE CYST OF PROXIMAL HUMERUS IN A 4 YEAR OLD: CASE REPORT

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ABSTRACT

Considered by many surgeons and radiologists as a "don't touch lesion" meaning lesions that need no further attention as described by CA Helms in 2013, Aneurysmal Bone Cyst (ABC) is a misnomer. This is so because it's neither an aneurysm nor a cyst. ABCs are benign expansile bone tumours representing about 1% of all bone tumours and affecting mostly those younger than 20 years of age. Typically found in the metaphysis of long bones, pelvis and posterior vertebral elements. They may have physeal extensions leading to growth abnormalities. The current mainstay of treatment following diagnosis is curettage with or without bone graft and use of adjuvant therapy according to the surgeon's preference. In spite of the number of techniques reported in literature, there remains a recurrence rate of 5% to over 40%.

This is a case report of a 4 year old boy who presented with 6 months history of recurrent right shoulder pain, associated with swelling and reduced range of motion. Following blood work ups, physical exam and radiological assessment, a preoperative impression of proximal humerus Juxta-physeal ABC was made. Open curettage, biopsy and bone substitute was performed through the deltopectoral approach. Histopathology results confirmed ABC. Seven months of follow up has demonstrated clinical and radiological remission.

Key words: Aneurysmal Bone Cyst, Curettage

INTRODUCTION

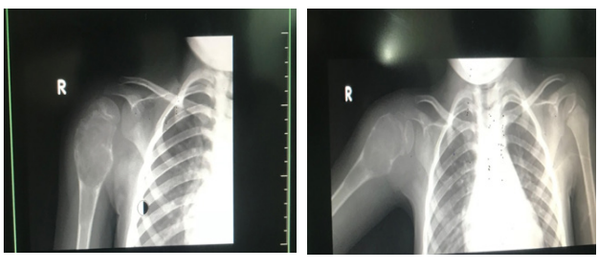
Aneurysmal Bone Cysts (ABC) are among a group of lesions described by CA Helms in 2013 who coined them as "Don't touch lesions"(1). Once radiologically diagnosed, they need no further attention. They include ABC, simple cysts, bone infarcts, cortical desmoids, geodes etc. ABC is a misnomer as these lesions are neither aneurysmal nor truly cystic due to lack of an endothelial wall (2). They are benign expansile lesions that produce cavities within bone then fill with blood. These cavities are lined by proliferative fibroblasts, giant-cells, and trabecular bone. Although first described in 1942 by Jaffe and Leichstein, their aetiology is still unknown. Historically they are believed to have resulted from increased venous pressure. Cytogenetic studies have shown specific translocations of the Ubiquitin-Specific Protease (USP) 6 gene but only in primary lesions (3). ABCs comprise of about 1% of all primary bone tumours and 80% occur in people younger than 20 years of age. Typically, they affect the metaphyses of long bones, pelvis, and posterior vertebral elements. They may have physeal extensions resulting in growth plate disturbances (4). As much as ABCs are considered as "Don't touch lesions" hence treated mostly conservatively, this case exhibited aggressive behavior with encroachment of the proximal humeral physis where most of the longitudinal growth of the upper limb occurs from. In order to curb future growth complications, surgery and biopsy was indicated. We therefore report a case of a 4 year old boy who underwent open curettage, biopsy and cavity filling with bone substitute. Eight months of follow up has demonstrated clinical and radiological remission.

CASE REPORT

We present a 4 year old school-going boy who reported experiencing recurrent painful right shoulder for the past 6 months. He had no other relevant medical or surgical history. His general condition was unremarkable. Physical examination demonstrated a moderately swollen, warm and tender right shoulder with reduced range of motion. Complete blood work-up was normal. Radiographs of the right shoulder revealed an expansile lytic lesion abutting the proximal humeral physis and circumscribed by a thin layer of cortical margin having areas with probable cortical destruction. Intralesional septae were also evident as shown in Figure 1 .

Figure 1

Preoperative radiographs of the right shoulder demonstrating the lesion on the proximal humerus in AP and lateral views



The patient was therefore prepared for an open curettage, biopsy and packing with Calcium TriPhosphate (CTP) bone substitute. This was done under general anaesthesia in a supine position through the conventional deltopectoral approach. A 5cm incision was made to reach the medial aspect of the cystic lesion in order to avoid injury to the axillary nerve laterally. The lesion was incised longitudinally to drain about 40 milliliters of bloody fluid followed by curettage all round but avoiding the superior physeal region. Saline washout was done then the cavity was packed with 10 grams of CTP and wound closed in layers following hemostasis. The contents were collected for histopathology and the report is displayed in Table 1.

Table 1

Histopathology results of the sample collected

Microscopic examination	Sections show abundant blood clot and bony trabeculae with fibrous septae lined by fibroblasts, myofibroblasts and hystiocytes, however, no endothelium or epithelium was seen. Osteoclast-like multinucleated giant cells with loose spindled stroma to cellular stroma and reactive bone is seen. Areas of degenerating calcifying fibromyxoid tissue is also noted. There are no overt features of malignancy.
Conclusion	Morphological features are most consistent with Aneurysmal Bone Cyst. Correlation with clinical, intra operative and radiological findings is advised.

Post operatively, the boy recovered well and was discharged from the hospital the following day on an arm sling. Post-op radiographs and follow up at weeks 3, 7, 15 and 32 are shown in Figures 2 - 5 respectively.

Figure 2

Post-op radiographs at 3 weeks

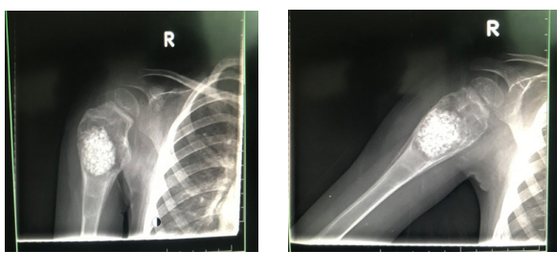


Figure 3

Post-op radiographs at 7 weeks

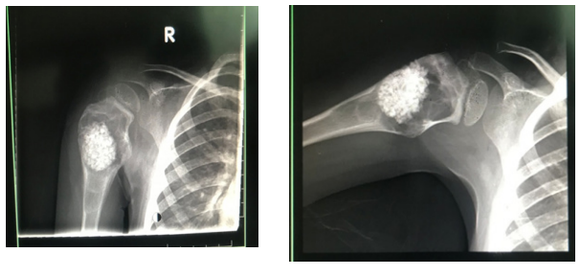


Figure 4

Post-op radiographs at 15 weeks

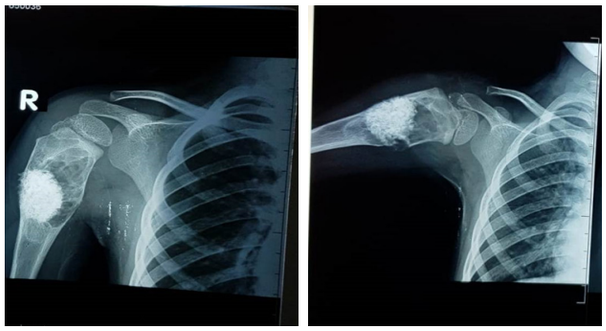
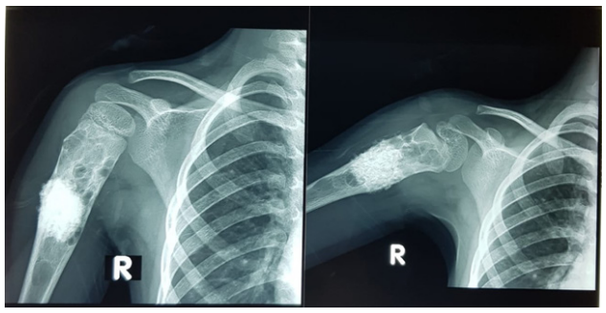


Figure 5

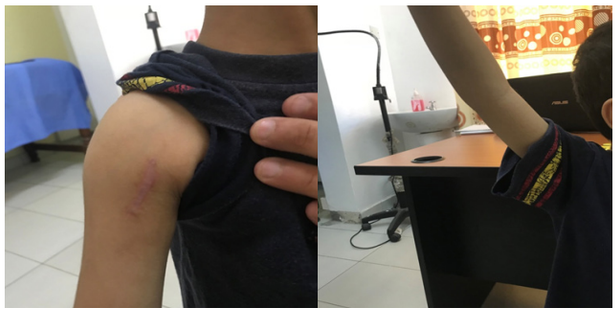
Post-op radiographs at 32 weeks



Clinical and radiological remission was noted to have been achieved. These were demonstrated by full range of motion at the affected shoulder associated with no pain and radiographic features of cortical bone formation within the cavity as demonstrated in Figure 6.

Figure 6

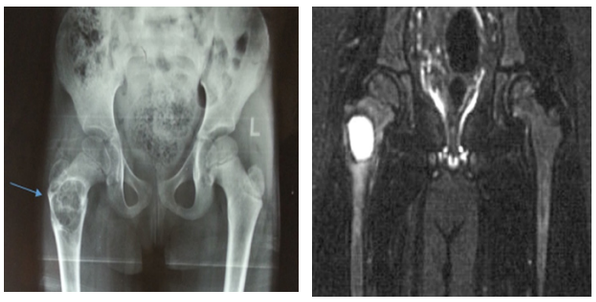
Photographs demonstrating the post operative range of motion of the shoulder



DISCUSSION

Comprising of about 1% of all primary bone tumours and mostly occurring in the skeletally immature patients (4). ABCs have a slight female predilection with a male to female sex ratio of 1:1.16. They are commonly located in the metaphyses of long bones, pelvis, and posterior vertebral elements. ABCs were first described by Doctors Jaffe and Lichenstein in 1942 when they reported pelvic and spine lesions that when exposing the lesion and opening into its thin wall, the surgeon was immediately confronted by a large hole containing bloody fluid (2). These lesions present with pain, swelling and sometimes with impending or pathologic fracture. They may affect the range of motion due to their location or produce mass and pressure effect for example nerve impingement in the spine. Growth plates can be affected leading to limb deformity and length discrepancies (5). Radiological diagnosis of ABC is characterized by eccentrically located radiolucent cystic lesions circumscribed by a thin cortex. A soap bubble appearance indicates fluid-levels within septae which may be confirmed by MRI as shown in Figure 7.

Figure 7
Radiological features of ABC and Magnetic Resonance Image of the proximal femur(5)



Grossly, the lesions appear as spongy, haemorrhagic masses covered by a thin shell of the reactive bone. Microscopically, red blood cells and often pale brown haemosiderin filling cyst-like spaces bounded by septal proliferations of fibroblasts, mitotically active spindle cells, osteoid calcifications, and scattered multi-nucleated giant cells are exhibited (2). Management of benign bone tumours differ depending on the histological type and their behavior ranging from non-operative management with observation of the lesion, intra-lesional injection or curettage with or without grafting, or adjuvant therapy. Marginal or even wide resections may be needed in aggressive lesions which have soft tissue involvement. All forms of treatment aim to relieve pain promote healing and prevent complications like recurrence and pathological fractures (6). Treatment of ABCs is largely conservative through observation and serial monitoring. Surgical treatment is opted when patients demonstrate pathological fractures or locally aggressive benign tumours associated with recurrent pain or exhibiting a restricted range of motion (7). This aggressive nature may lead to physeal extensions resulting in growth plate disturbances and subsequent development of deformities hence surgery is considered (5). Surgical treatment options include En bloc excision which has been found to have the lowest rates of recurrence though at the expense of high morbidity (2). Currently, curettage with or without use of bone-graft is favored. The surgeon may consider use of adjuvant to reduce possibility of recurrence which include bone cement, high-speed burr, argon beam, phenol, and cryotherapy. In cases of recurrence or inoperable lesions, radiotherapy, sclerotherapy using polidocanol and arterial embolization have also been employed (8). Other pharmacological agents that have been used and still undergoing studies include doxycycline, bisphosphonates and denosumab (2). Endoscopic curettage of different types of intra-osseous benign bony lesions has proved to be an effective treatment modality with promising oncologic outcome, improved functional scores, and fast functional recovery (6). We used calcium phosphate cement product in this case. These come in pellet form or injectable biocompatible bone substitutes. Though arguably costly, once available, it has been shown to be friendlier during handling, high mechanical strength and good osteoconductive biological properties. The study also showed a recurrence rate of 6% when it was used for more aggressive Giant cell tumours. In general, recurrence is common ranging between 5% to over 40% especially in young males with proximal femur lesions (7).

Declaration: I declare that this was original work and for any cited information, relevant references have been provided. The report was not funded by any individual or organization.

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