

Aneurysmal bone cyst of the lateral end of clavicle in a twelfth year old girl

Case study

Mohammed S. Al-Iedani, F.I.C.M.S. *; Ghadeer H. Majeed, F.I.C.M.S. *; Basher A. Rashid, F.I.C.M.S. **

ABSTRACT

Introduction: An aneurysmal bone cyst are enigmatic lesion of unknown cause and presentation and are difficult to distinguish from other lesions, it is a benign, but expansible tumor like lesion that generally occurs in the long bones. An aneurysmal bone cyst arising from the flat bone like clavicle is rare .

Case presentation: We report a 12-year-old girl child with an aneurysmal bone cyst of the lateral third of left clavicle treated with enblock resection. The pathologic findings confirmed the diagnosis of aneurysmal bone cyst. The patient has been followed up for one year with no evidence of recurrence .

Conclusion: En bloc resection can be curative and provide good results for this rare type of clavicle tumour.

Keywords: Aneurysmal bone cyst, clavicle, enblock resection.

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*Assistant professor in Al- Kindy College of Medicine - University of Baghdad.

**General surgeon, Al-Kindy teaching hospital.

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Corresponding to Dr. Mohammed S. AL-Ideani

Aneurysmal bone cyst (ABC) was first reported by Jaffe and colleagues in 1942. They used this term to describe the "blow out" radiographic appearance and blood filled contents of the cystic spaces ¹. ABC is a non-neoplastic bone tumor that occurs predominantly in children and young adults, mainly involving the long bones and the vertebrae. The tumor considered more of a descriptive term since there is no concrete evidence as to the direct origin, causation, or pathogenesis of the lesion itself ².

Primary bone tumors of flat bones like clavicle are rare. True benign tumors are much uncommon than metastatic or malignant lesions. Aneurysmal bone cyst (ABC) is a benign but locally aggressive lesion of the bone which accounts for 3% of all bone tumors. Its histology is characterized by multiloculated cystic tissue filled with blood. Etiology and pathogenesis of this lesion remain unclear ³. It is a disease mainly of the young with a peak incidence in the second decade. However, it may on occasion occur in the elderly and the very young ⁴. ABC may involve almost any bone but the most frequent sites are long tubular bones and vertebrae. Despite very characteristic radiological features, the uncommon site led to diagnostic difficulties in the present case. The clavicle is a rare site for these lesion and not many have been reported in the literature. Smith in 1965 could find only 25 cases in the medical literature, textbooks and atlases ⁵. Because of these factors, this report is felt to be of interest. We report a case of ABC of the left clavicle treated with enblock resection.

Case report: A 12 years old girl presented with swelling in his left clavicular region that had been increasing in size progressively since last six months to reach the size of a lemon. The swelling at the acromial end had distinct edges, was smooth surfaced, figure 1, and overlying skin temperature was normal. The mass was bony hard, non-tender and the skin over the swelling was pinchable.

Swelling was immobile. No functional impairment, skin changes or dilated veins or signs of neurological deficits and lymphadenopathy was noted. There was history of trauma one year before and non-displaced fracture of mid shaft clavicle treated uneventfully at that time by an arm sling. There was no history of recurrent fever and loss of weight. No other lumps or swellings were present anywhere else.

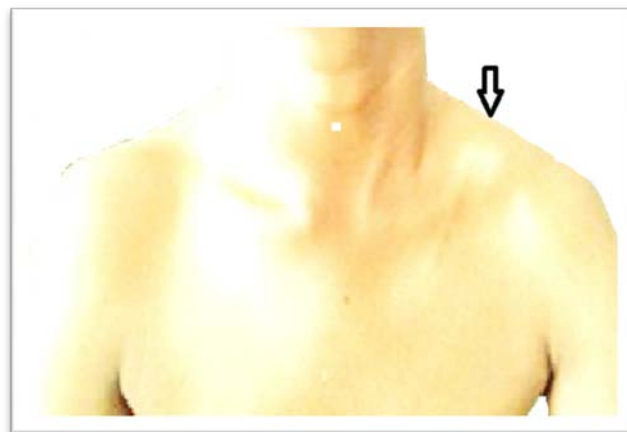


Figure 1: Photograph depicting swelling at the left clavicular region.

Radiograph showed a cystic expansile lesion of the lateral end of the left clavicle bounded by a thin layer of bone, Figure 2, based on the appearance various possibilities including simple bone cyst, ABC, eosinophilic granuloma, and enchondroma were considered.

Basic hematological work up including complete blood count, ESR, CRP and alkaline phosphatase were within normal limits. The lesion was further studied with MRI left shoulder, Figure 3, and fine needle aspiration cytology.

MRI pointed to the possibilities of ABC, Giant cell tumor and

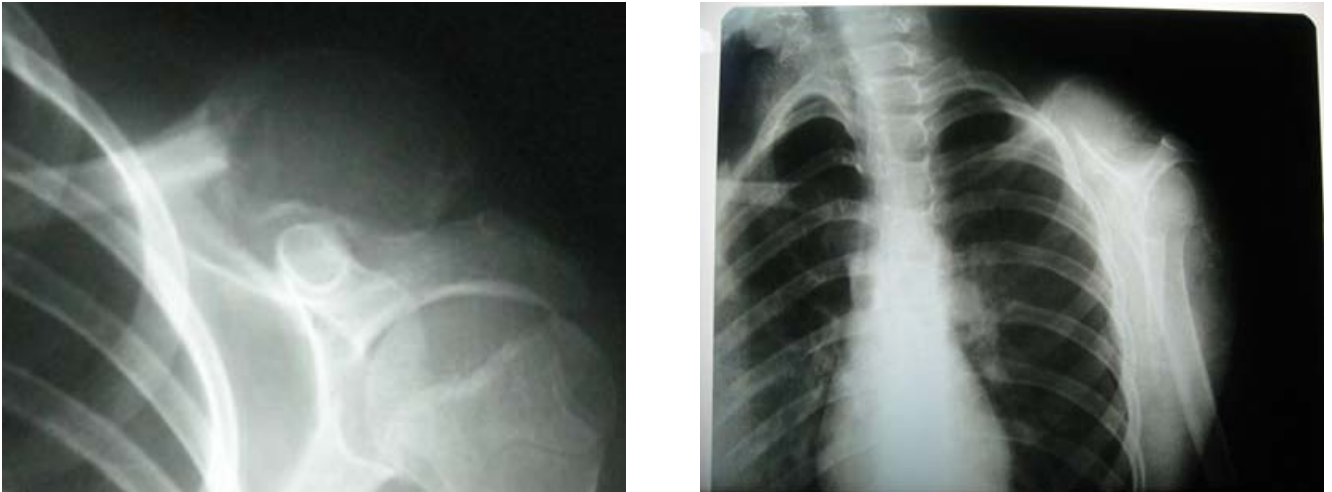


Figure 2: Radiograph showing cystic expansile lesion bounded by a thin layer of bone at the lateral end of the left clavicle.

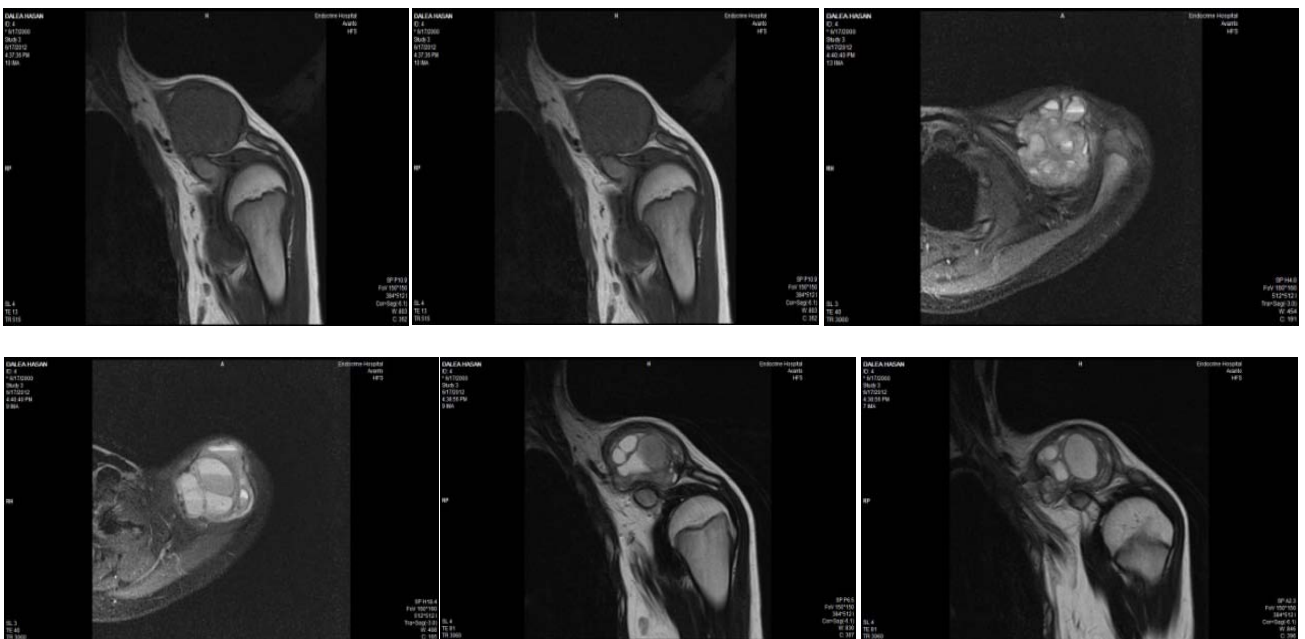


Figure 3: MRI Left shoulder (Plain and contrast) Scan showing multilobulated T1 hypointense, T2 hyperintense arising from end of left clavicle with destruction of lateral end.



Figure 4: Intra operative photograph showing blood-fluid in the cyst.



Figure 5: Intra operative photograph of ABC.

chondroblastoma. FNAC report came out to be inconclusive.

Therapeutic options which were considered at that point were complete resection of the lesion. Intraoperatively, initially a 10cc disposable syringe was used to aspirate the contents of the cavity. The content was found to be a blood-fluid, Figure. 4. The lesion was approached by direct incision over the lateral half of the clavicle and enblock resection of the lateral third of the clavicle was obtained, figure 5. The limb was immobilized in cuff and collar sling. Postoperative period was uneventful. The preoperative diagnosis was confirmed with the histopathological examination of the resected specimen.

Discussion. Aneurysmal bone cysts were first described in 1942 by Jaffe and Lichtenstein, and later again in 1950 by Lichtenstein¹. The term aneurysmal bone cyst considered more of a descriptive term since there is no concrete evidence as to the direct origin, causation, or pathogenesis of the lesion itself². The lesion itself is usually described as a blood filled cavity within an expanded region of bone, and a nutrient artery may or may not be visualized as to the source of the fluid⁶.

The signs and symptoms of aneurysmal bone cysts can be nonspecific; 55% of the time there is mild pain and 24% of the time there is swelling, and the pain may be brought on by activity⁷. In order to properly diagnose the lesion, it is imperative to use MRI or CT scans to augment the information gained from the radiographs. Once all other possible differentials are excluded, and aneurysmal bone cyst is found to be the diagnosis, then it is important to fully understand the extent of the lesion. In order to properly treat the lesion surgical intervention is usual advised, based on the size and amount of pain the patient is in. While some lesions will resolve on their own, some are very large and to leave them alone would put the patient at risk of fracture. Surgical treatment of aneurysmal bone cysts has evolved over the years; treatment now includes curettage, bone graft/chips, sclerotherapy, marginal excision, and a

combination of the aforementioned methods⁸. However, even with proper surgical treatment the reoccurrence rate of the lesion is 15-20%⁹.

Despite the long experience of orthopedicians, radiologists, and pathologists, there is limited knowledge regarding the cause of the lesion, its natural history, and the results of treatment^{10, 11}. An interesting theory about the etiology of primary ABCs is that the lesions occur because of hemorrhage in the bone as a result of increased venous pressure. The hemorrhage is thought to lead to osteolysis. The osteolysis, in turn, causes further hemorrhage, leading to exponential growth of the tumor. This theory would perhaps explain why ABCs are uncommon in the clavicle and bones of the facial skeleton, where the venous pressure is low. On the other hand, ABCs are common in long bones, where the venous pressure is high and the marrow content is greater¹². Conventional thinking; however, ascribes these lesions either to a reaction to physical injury, which may sometimes be remote, or to a vascular disturbance. The concept that the lesion represents a vascular degenerative process for some benign bone lesions is an attractive one, but the pathologic findings, with rare exception, do not really support this proposal. Few pathologic specimens contain tissues that are highly characteristic or diagnostic of giant cell tumor, chondroblastoma, hemangioma, osteoblastoma, non ossifying fibroma, chondromyxoid fibroma & others^{11, 13, 14, 15}. Thus, it is often thought that ABC is more of a pathophysiological change in a pre-existing primary bone lesion rather than a single, unique entity¹⁶.

Difficulty can occur in diagnosing these lesions. The imaging studies, even CTs and MRIs, sometimes do not provide clearly diagnostic criteria for the diagnosis of ABC, and ABC is sometimes added on to a list of diagnoses including eosinophilic granuloma, giant cell tumor, non ossifying fibroma, unicameral bone cyst, fibrous dysplasia, chondroblastoma, chondrosarcoma, chondromyxoid fibroma, Ewing's tumor etc^{13, 14, 17}.

In the past, curettage alone was employed in the treat-

-ment, later methods like saucerisation, resection, radiotherapy, cryotherapy and vascular occlusion are being employed. Nevertheless, there is no consensus among treating physicians regarding how these methods should be used. As a result, there are quiet contradictory reports regarding results and complications¹⁸. Resection of lesion offers low recurrence rate but this option cannot be exercised everywhere. A combination of cryosurgery and curettage has been reported by few authors that reported local control after the first treatment in 82% patients¹⁹. Radiotherapy can result in radiation induced sarcomas and can cause radiation induced injury to physis¹⁸. Thus radiotherapy is reserved in cases that cannot be operated because of their location and to prevent damage to the function of important structure¹⁴. In some cases embolisation of a feeding vessel may help to decrease vascularity, making the surgical procedure less bloody, especially in difficult locations such as spine and pelvis but it is a highly demanding technique and may not be available at all centers. The recurrence rate in young children with ABC may be as high as 100%³. Autograft implantations or utilization of intercalary allografts are quite successful and for the most part, are used for patients with lesions that are large or seem to threaten the integrity of the bone. Enblock resection is other option especially in rib and clavicle without incidence of recurrence. Our patient responded nicely to this form of treatment and we feel that this case enriches existing data regarding treatment option of an ABC in an unusually young patient and in unusual location.

In conclusion, ABC is a rare, benign pathology, which should be considered in the differential diagnosis of clavicle region swelling. Complete surgical excision can be the best treatment for cure.

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