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Management of aneurysmal bone cyst of proximal femur with a pathologic intertrochanteric femur fracture with an intramedullary nail and bone graft: A case report

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Abstract
Aneurysmal bone cyst (ABC) is a benign tumor like lesion that is an expanding osteolytic lesion consisting of blood-filled spaces of variable size separated by connective tissue septa containing trabeculae or osteoid tissue and osteoclast giant cells. Nearly 95% occur in the first three decades. Aneurysmal bone cysts are benign active or aggressive bone lesions that commonly arise in the metaphysis of long bones, especially the femur, tibia and humerus. They are extremely rare, occurring in 1.5 per one million people per year. And usually present with pain, swelling or fracture.
We report a case of a giant aneurysmal bone cyst in the proximal femur with a pathologic Inter-trochentric femur fracture in a 23 year old male patient which was treated with a proximal femur nail and curettage and bone grafting of the lesion in the proximal femur. At 18 months follow-up, the lesion had subsided and patient was walking pain-free with full range of motion without any deformity. We suggest this method of treatment to be worthwhile for a pathologic fracture of an ABC at proximal femur.

Keywords: Aneurysmal bone cyst, proximal femur, intramedullary nail

Introduction
Aneurysmal bone cyst (ABC) is a benign tumor like lesion that is an expanding osteolytic lesion consisting of blood-filled spaces of variable size separated by connective tissue septa containing trabeculae or osteoid tissue and osteoclast giant cells[1]. Seventy-five percent of the lesions occur in the first two decades of life, and almost 95% occur in the first three decades [2]. The long tubular bones are the most common sites for ABCs, followed by the spine and the flat bones. These three areas account for 80% of all ABCs. In a published review of 897 cases of ABC, the following rates of occurrence were reported [3]: Tibia–17.5% Femur–15.9% Vertebra – 11.2% Humerus – 9.1%.
The aetiology of aneurysmal bone cysts is unknown, but numerous authors have proposed that they form in response to vascular disruption in the bone due to a pre-existing primary tumor or a traumatic insult. ABCs may arise de novo; those that arise without evidence of another lesion are classified as primary ABCs. Most primary ABCs demonstrate at (16; 17) (q22; p13) fusion of the TRE17/CDH11-USP6 oncogene [4]. This fusion leads to increased cellular cadherin-11 activity that seems to arrest osteoblastic maturation in a more primitive state [5]. Such ABCs in the presence of another lesion are called secondary ABCs, and treatment of these ABCs is based on what is appropriate for the underlying tumor. The true aetiology and pathophysiology remains unknown.
Patients with an aneurysmal bone cyst (ABC) usually present with pain, a mass, swelling, a pathologic fracture, or a combination of these symptoms in the affected area. Pathologic fracture occurs in about 8% of ABCs. Radiographic findings usually consist of an eccentric or, less commonly, a central or sub periosteal lesion that appears cystic or lytic. Images may show expansion of the surrounding bone with a blown-out, ballooned or soap-bubble appearance, the cystic spaces, rarely, partially ossified septa can be seen [6].

Jaffe and Lichtenstein first described ABC as its own entity in 1942, when they noted "a peculiar blood-containing cyst of large size" [7]. Two cases were reported in which a lesion with a "soap-bubble" appearance on radiographs [7]. The same characteristics are demonstrated on computed tomography (CT) as on plain radiography; however, CT scans also show internal septation (calcified rim, eggshell appearance), which may be completely or partially intact. Fluid-fluid levels can also be seen [8]. Findings from magnetic resonance imaging (MRI) are similar to those from CT, but MRI can more specifically reveal blood within the lesion, as well as expansion into the soft tissues [6]. The mainstay of treatment has been intralesional curettage [9]. Other surgical options include en-bloc resection or wide excision, selective arterial embolization, and curettage with locally applied adjuvants such as liquid nitrogen, argon beam photocoagulation, or phenol.

The prognosis for an ABC is generally excellent, though some patients need repeated treatments because of recurrence. The overall cure rate is 90-95% [10, 11]. Differential Diagnoses include: Chondroblastoma, Fibrous Dysplasia, Giant Cell Tumor, Telangiectatic osteosarcoma, Unicameral Bone Cyst. Histologically the gross appearance of the ABC is that of a blood-soaked sponge. A thin subperiosteal shell of new bone surrounds the structure and contains cystic blood-filled cavities. The tissue within shows brownish intertwining septa.

Case report

A 23 year old male patient presented with a history of trivial fall at home complaining of right hip pain with restricted range of motion. On examination patient had localised swelling and tenderness at proximal thigh and had an external rotation deformity of right lower limb with no neuro-vascular deficit. On X-ray a pathologic Inter-trochentric femur fracture having giant aneurysmal bone cyst in the proximal femur was seen as an eccentric metaphyseal lesion that appeared cystic with expansion of the surrounding bone with a soap-bubble appearance with the cystic spaces partially ossified septa.

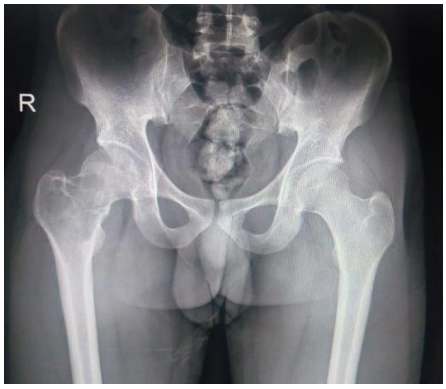

Fig 1: Plain Radiograph of the Lesion

Investigations revealed Hb-10.9 gm/dl, with normal total and differential WBC counts, ESR 20 mm in the 1st hour, Hematocrit 33.3%, and platelet count 2,96,000/cu mm. Routine urinalysis was normal. Blood biochemistry including Blood sugar, urea and creatinine was normal. The serum calcium was 9.1 mg/dl and serum phosphate was 4.1 mg/dl. Liver function test revealed serum albumin 3.2 gm/dl, globulin 3.9 gm/dl, total serum bilirubin 0.5 mg/dl, SGOT 36 IU/ml, SGPT 40 IU/ml, and serum alkaline phosphatase 179 IU/l. MRI was done suggestive of ill-defined expansile lytic lesion involving the proximal femur with adjacent soft tissue edema.

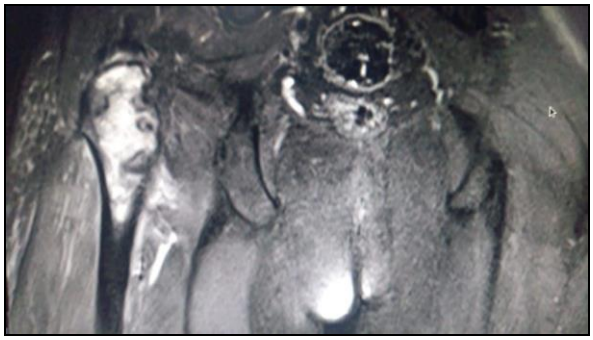

Fig 2: MRI Image of the Lesion

Consent was obtained from the patient and his relatives prior to surgery. Pre-operative planning was done and excision of the lesion with proximal femur intramedullary nail fixation for the fracture and the created bone gap was to be filled up by autologous bone graft from iliac crest and some allograft also was used intra operatively to fill the void after excising the lesion in the proximal femur.

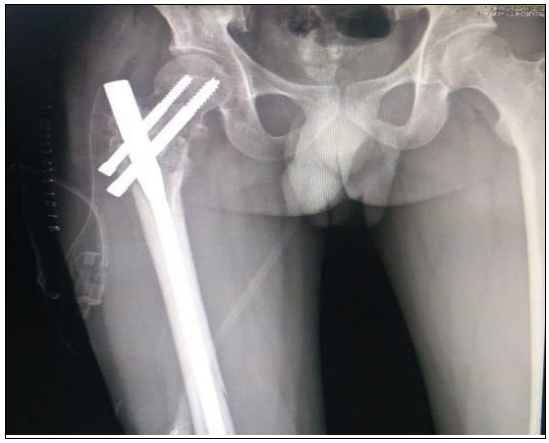

Fig 3: Post Operative Radiograph AP view

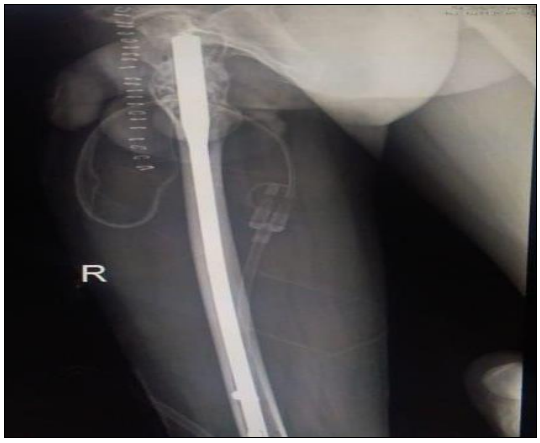

Fig 4: Post Operative Radiograph Lateral view

Post operatively histopathology reports confirmed Aneurysmal bone cyst and the patient was advised nil weight bearing for initial six weeks post op and partial weight bearing till 3 months. Static exercises were started after 3 weeks post operatively. He was followed up at six weeks, three months, and six months after surgery. At the end of six weeks he regained full range of movements of at hip joint. Series of x rays were done at 6 weeks and three months. At 18 months follow-up, the lesion had subsided and patient was walking pain-free with full range of motion with a LLD of 2cm.

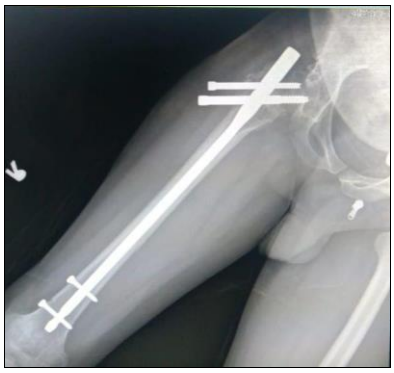

Fig 5: Plain AP radiograph at 18 month

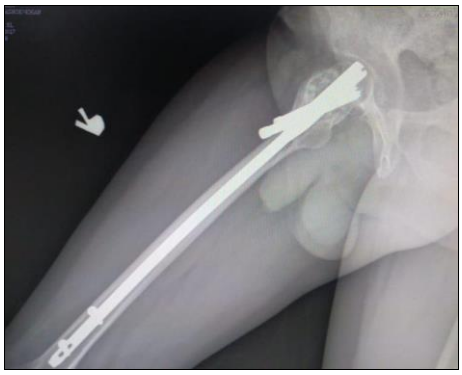

Fig 6: Lateral View Radiograph at 18 months follow-up

Discussion

Aneurysmal bone cyst (ABC) is a benign, expansile, non-neoplastic lesion of the bone, characterized by channels of blood and spaces that are separated by fibrous septae. Because ABCs may affect any bone in the body, the relevant surgical anatomy necessarily varies with location. The long tubular bones are the most common sites for ABCs, followed by the spine and the flat bones. These three areas account for 80% of all ABCs. When present in long tubular bones, ABCs tend to be eccentrically located in the metaphysis. Giant ABC is an uncommon condition and can be difficult to handle because of the destructive effect of the cyst on the bones and compressive effect on the nearby structures, especially in the weight-bearing bones of the body.

Table 1: Morphologic types of ABC on the basis of radiographic findings

Type I	Central metaphyseal presentation; well contained within the bone, with the bone profile intact or with slight expansion
Type II	ABC that involves the entire segment of bone; an inflated appearance with cortical thinning
Type III	Eccentric metaphyseal location; no or minimal expansion of the cortex
Type IV	Subperiosteal extension; no or minimal cortical erosion; rare in the diaphysis
Type V	Metadiaphyseal location; inflation of periosteum toward the soft tissues; penetration of the cortex; extension into cancellous bone

A younger age, open growth plates, and a metaphyseal location all have been associated with an increased risk of recurrence [10]. The stage of the ABC has not been shown to influence the rate of recurrence; however, Capanna[12] described the five morphologic with types I and II recurring more often than types III, IV, and V.

ABCs may rise de novo; those that arise without evidence of another lesion are classified as primary ABCs. ABCs are also seen to arise in an area of previous trauma[2]. ABCs may also be caused by a reaction secondary to another bony lesion - This theory has been proposed because of the high incidence of accompanying tumors in 23-32% of ABCs; although giant cell tumors of bone are most commonly present, many other benign and malignant tumors are found, including fibrous dysplasia, osteoblastoma, chondromyxoid fibroma, nonossifyingfibroma, chondroblastoma, osteosarcoma, chondrosarcoma, unicameral or solitary bone cyst, hemangioendothelioma, and metastatic carcinoma. Such ABCs in the presence of another lesion are called secondary ABCs, and treatment of these ABCs is based on what is appropriate for the underlying tumor. Although ABC can be diagnosed on an x-ray, in some situations, distinguishing an ABC from a giant cell tumor of bone or a Telangiectatic osteosarcoma is difficult [13]. MRI can distinguish ABCs from telangiectatic osteosarcoma (TOS).

Reported primary recurrence rates have varied greatly. Some studies have reported recurrence rates in curettage and bone grafting had (30.8% recurrences) [3]. Small studies have shown a benefit to using selective arterial embolization [14], and some authors advocate it as a first-line treatment [15]. Some authors argue that not enough data on selective embolization exist and that surgery is the first-line treatment [16]. Intralesional excision and bone grafting has the most data to suggest that it is a safe and effective method [17].

So our choice was to use non-vascularized autogenous cancellous bone grafts as they are technically easier to harvest and provide excellent structural bone support with successful rigid internal fixation with an intramedullary proximal femur nail. In the present case, the final construct obtained was stable and allowed progressive weight-bearing without any recurrence of the lesion.

Conclusion

Treatment of ABCs with associated pathologic fracture, especially a fracture of the hip, is a challenging problem and is always an indication for intervention, so we recommend that the best treatment includes excision of the lesion and bone grafting by autologous bone graft from iliac crest and allograft to fill the defect and rigid internal fixation for the fracture with intramedullary nailing.

Conflict of interest: There are no conflict of interest declared.

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