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Proximal femur pathological fracture secondary to aneurysmal bone cyst in a child

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Abstract
Aneurysmal bone cysts are enigmatic, locally destructive blood filled lesions of bone with unknown cause & unusual presentations. We report a case of an 8-year old male child who presented to us with a pathological fracture of right hip after a trivial fall. It was an aggressive aneurysmal bone cyst of the proximal femur complicated by a subtrochantric fracture. Treatment was done successfully with an intralesional curettage and implantation of a frozen cancellous cadaveric allograft of femoral head followed by internal fixation of the pathological fracture with plate and screw.

Keywords: Aneurysmal bone cyst, femur, pathological fracture

Introduction
Aneurysmal bone cyst (ABC) is a rare skeletal tumour that occurs mostly in the first two decades of life. It is an intramedullary eccentric metaphyseal & rapidly expansible benign lytic lesion with multiloculated blood filled cystic cavities [1]. It causes extensive weakening of the bony structure & impinge on the surrounding tissues. Pathological fractures secondary to benign bone tumours are difficult to treat because of the extensive bone destruction, peri-particular location and high risk of local relapse [2]. We report a rare case of aneurysmal bone cyst of proximal femur complicated by subtrochantric fracture. It was successfully treated by intralesional curettage, chemical and electrical cauterisation followed by implantation of frozen cadaveric cancellous bone graft of femoral head. Allografting per se as a choice of surgical protocol is less explored. It was followed by internal fixation of the pathologic fracture by dynamic compression plates.

Case report
An 8-year old male child presented to the orthopaedic OPD with pain & swelling in the right hip and unable to bear weight on right lower limb following a trivial fall. On examination, right lower limb was externally rotated, swelling over hip and tenderness over proximal femur was present. Radiograph of right femur was done and there was sub trochanteric fracture with eccentric lytic lesion at proximal metaphysis area (Figure 1). Computed tomography was done to confirm the lesion (Figure 2) and to know the extent of lesion which showed large lobulated predominantly cystic intramedullary lesion involving the neck & proximal shaft of right femur with neck femur fracture with deformity suggestive of benign neoplasm. Haematological examination was normal.

Surgical management
The main stay of treatment of the tumour is curettage (Figure 3, 4). It was followed by chemical and electrical cauterisation to remove the residual tumour and to achieve haemostasis, which was further followed by implantation of cadaveric cancellous bone graft (Figure 5) of femoral head mixed with 10ml of bone marrow aspirate from iliac crest.
The fracture was treated by internal fixation using dynamic compression plate (Figure 6). He tolerated the procedure well.

Fig 1 and 2: Pre operative radiographs

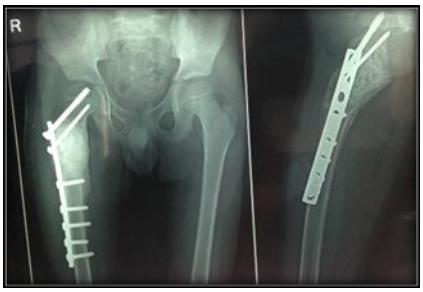

Fig 3 to 6: Intraoperative and postoperative images

Discussion

Aneurysmal bone cyst is a locally destructive osteolytic lesion which occurs at metaphyseal region of bones. These tumors consist of blood-filled septate cavities lined by fibroblast and histiocytes. Hemosiderin-laden macrophages, chronic inflammatory cells, and multinucleated giant cells also are present. It most commonly affects proximal humerus, distal femur, proximal tibia and spine [1]. It has an annual incidence of 0.14 per 1 lakh population [3] constituting 1% of all primary bone tumours. It was first described by Jaffe and Lichtenstein4 in 1942. Our case report had pathological fracture which occurs in 8% of aneurysmal bone cyst cases [3]. Exact incidence of pathological fracture among proximal femur ABC is not found in literature. Aneurysmal bone cyst is most commonly seen in 2nd decade of life, being slightly more common in females [5]. In a study which reviewed 150 patients of ABC, 9 patients had lesions in proximal femur [5]. True aetiology of these tumours is unknown. Generally, ABCs are divided into two types: primary and secondary. Most lesions arise de novo and are termed as primary ABCs and the ones which arise in other lesions like fibrous dysplasia,

Osteoblastoma, chondromyxoid fibroma, non-ossifying fibroma, chondroblastoma, osteosarcoma, chondrosarcoma, unicameral bone cyst, hemangioendothelioma and metastatic

Carcinoma, such tumours are designated as secondary ABCs [6]. Features of the current case were consistent with primary ABC. Although the pathogenesis is uncertain, it is likely that aneurysmal bone cysts result from local circulatory disturbance leading to increased venous pressure and production of local haemorrhage [7]. Radiographs typically show an eccentric, lytic lesion which is expansile, seen at metaphyseal area [1, 2] The natural history of aneurysmal bone cyst has been described as evolving through four radiological Stages: initial, active, stabilization and healing. In the initial phase, the lesion is characterized by a well- defined area of osteolytic with discrete elevation of the periosteum. This is followed by a growth phase, in which the lesion grows rapidly with progressive destruction of bone. The growth phase is succeeded by a period of stabilization, in which the characteristic soap bubble appearance develops, as a result of the maturation of the bony shell. Final healing stage results in progressive calcification and ossification, with the lesion transformed into a dense bone mass [8]. The current case was consistent with the growing phase of ABC. CT scanning can be used to define the lesion and to know the extent of lesion. It is helpful in lesions which are located at complex anatomical areas. We treated the patient successfully by intralesional curettage, chemical and electrical cauterisation followed by filling of cavity with frozen cadaveric cancellous bone graft of femoral head. It was followed by internal fixation of the pathologic fracture by dynamic compression plate. Patient was on hip Spica cast for two months and later mobilised. The literature on pathological fractures of proximal femur in aneurysmal bone cyst in benign cases is limited. Only few case reports are there in literature with proximal femur ABC with fracture [9-11].

Conclusion

Aneurysmal bone cysts are rare in the first decade of life. However, the condition must be borne in mind when the clinician is presented with a lytic bony lesion. Biopsy is the gold standard for diagnosis in such cases. For achieving bony union, fracture fixation should be supplemented with bone grafting.

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