Aneurysmal bone cyst: An unusual cause of pathological intertrochanteric fracture in an eight year old boy

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Citation

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Abstract

Aneurysmal bone cysts are rare benign tumors of bone. Most of the patients are in the second decade of life and most cysts are clinically silent. The authors report an ABC in the proximal femur of an eight year old boy presenting as a pathological intertrochanteric fracture.

INTRODUCTION

Aneurysmal Bone Cysts are rare, accounting for 1 – 6 % of all the primary bone tumours. The usual age of presentation is 10 – 20 years. We report an interesting case of an ABC in the upper femoral metaphysis presenting as a pathological intertrochanteric fracture in an eight year old boy.

CASE HISTORY

An eight year old boy presented with chief complaints of pain in the right hip since the last 2 months. The pain was insidious in onset, mild to moderate in intensity and did not show any radiation or any diurnal variation.

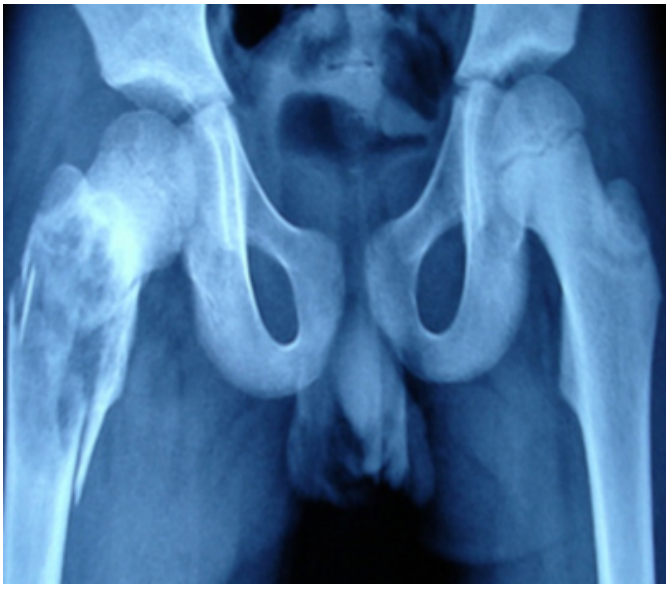
One day back, he had a trivial fall while playing following which the pain became severe and continuous. He was unable to bear weight on the right limb.

On examination, the right lower limb was in an attitude of full external rotation. There were no swellings, scars, sinuses or abnormal veins in the region of right hip. The greater trochanter was tender and all the movements of hip were painfully restricted. Distal neurovascular status was within normal limits.

Radiographs of pelvis with both hips revealed a well defined, homogenous and expansile lytic lesion in the region of the right upper femoral metaphysis. The width of the lesion was more than maximum width of the growth plate. The cortex surrounding the lesion had been thinned out leading eventually to a pathological intertrochanteric fracture.

Figure 1

Figure 1: X Ray of Pelvis with both hips AP showing a ABC in the proximal femoral metaphysis with an intertrochanteric fracture.



Serum biochemistry studies were within normal limits.

CT guided FNAC was done but was inconclusive.

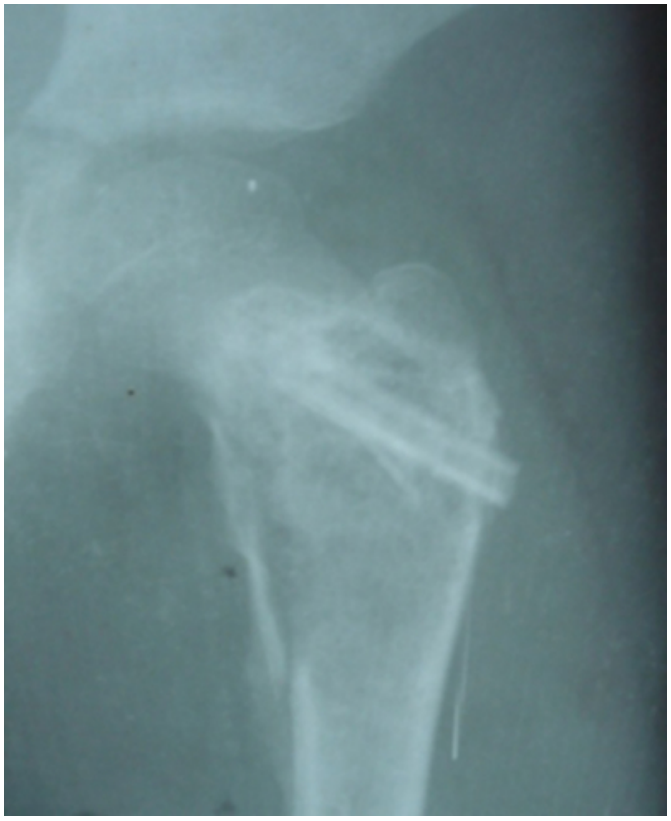
A provisional diagnosis of Aneurysmal bone cyst was made based on the roentgenographic findings. The diagnosis was confirmed subsequently on histopathology.

Curettage was performed and the resultant cavity filled with a fibular strut graft and cancellous iliac bone graft. Immobilization was achieved in a hip spica and the fracture united without complications in 12 weeks. On follow up at 16 weeks, the patient had no complaints with non tender,

smooth, healed scar at the incision site.

Figure 2

Figure 2: Post operative X Ray demonstrating curettage of the cyst and insertion offibular strut graft and cancellous iliac bone graft.



DISCUSSION

One of the non-neoplastic bone lesions which mimic primary bone tumors, Aneurysmal bone cysts are actually masses of vascular spaces encased within periosteal new bone. This innocuous description, however, does not undermine their ability to destroy normal bone and extend into the surrounding soft tissues. The condition was first described by Jaffe and Lichtenstein in 1942.

ABCs are rare, accounting for 1-6% of all the primary bone neoplasms. Patients are usually below 20 years of age, but it is rare in very young children. Most of the studies show a slight female preponderance.[1, 2]

Aetiology of these tumours is unknown. Most of the lesions arise de novo and are termed as 'Primary ABCs'. They may also arise in conjunction with other lesions viz. fibrous dysplasia, osteoblastoma, chondromyxoid fibroma, nonossifying fibroma, chondroblastoma, osteosarcoma, chondrosarcoma, unicameral bone cyst, hemangioendothelioma, and metastatic carcinoma. Such tumours are designated as 'Secondary ABCs'[3].

ABCs can involve any bone; the favoured sites of involvement being the long tubular bones, the spine and the pelvis. The lesion is invariably metaphyseal though rarely, it may be subperiosteal where it tends to form soft tissue masses. Vertebral ABCs most commonly involve the arches; lesions in the vertebral body are rare.

Pain and swelling, which may have existed for months or even years, is one of the modes of presentation. Alternatively, a history of trauma which precedes the above symptoms is elicited. Other features are site specific, such as limitation of joint motion and signs of spinal cord or nerve root compression.

The affected bone characteristically appears cystic and ballooned outward. The lesion is metaphyseal, lytic, eccentric and extends outward to destroy the cortex. A thin rim of periosteal new bone may be demonstrable, if mineralized.[4]

Capanna et al classified ABCs into 5 morphologic types based on the radiographic findings[5]:

- 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

Computed tomography scanning: Internal septation may be seen, which may be partial or complete. Fluid-fluid levels can also be seen. The fluid-fluid levels are caused by the separation of cellular material and serum within the cystic spaces.[6] To see these levels best, the patient must remain in the position in which they are imaged for at least 10 minutes to obtain enough separation of the materials of different attenuation. The CT scan views then must be acquired in a

plane that is perpendicular to the fluid levels. ABCs characteristically donot show enhancement on contrast studies. The differentials include telangiectatic osteosarcoma, simple bone cyst, chondroblastoma, fibrous dysplasia, giant cell tumor, malignant fibrous histiocytoma, synovial sarcoma and soft tissue hemangioma. These lesions show contrast enhancement, an important point in differentiating them from ABCs. MRI shows a low signal rim separating the cyst from the medullary cavity of the affected bone and well defined margins separating the ABC from adjacent soft tissues. The cysts are heterogeneous in signal intensity,attributable to the fact that blood is in various stages of breakdown in different parts of the same cyst. The lesions show a low signal on T1W and high signal on T2W images.

Bone scans show increased uptake at the periphery of the lesion, attributable to active new bone formation in this region. The histopathologic features are cavernous blood filled spaces, lacking an endothelial lining. These spaces are enclosed in fibro-osseous septae. The supportive connective tissue contains multinuclear giant cells and calcium deposits. In contrast to the tumor giant cells, the nuclei of these giant cells are smaller. Periosteal new bone formation is seen in the peripheral rim. Aneurysmal bone cysts may regress spontaneously and ossify, a fortuitous event. The modalities of active treatment include:

Curettage with bone grafting: Since recurrence and even persistence of the lesion following curettage is known, the procedure should be supplemented with cauterization of the cyst wall with phenol and alcohol followed by bone grafting[7]. Liquid nitrogen cryotherapy is an alternative. Transcatheter arterial embolisation of cysts; which is done in case of cysts located about the pelvis[8].

Synthetic Bone Substitutes are being increasingly used following curettage; most of these are calcium phosphate ceramics[9].

Irradiation: Is reserved for inoperable cysts. It increases the risk of sarcomatous change, hence not routinely used.

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. Owing to the high rates of recurrence, curettage should be supplemented with bone grafting.

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