Large Predominantly Solid Variant of Aneurysmal Bone Cyst in the Diaphysis of Femur: A Rare Entity

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Abstract

Aneurysmal Bone Cyst (ABC) is a non neoplastic solitary lesion which commonly affects long tubular bones and spine. Most lesions are predominantly cystic, and affect the metaphyseal region in the long bones with extension into the diaphysis but purely diaphyseal lesions are uncommon. In this case report, we describe X-ray and magnetic resonance imaging (MRI) findings in a pathologically proved case of predominantly solid variant of ABC involving the diaphysis of femur.

Keywords: Diaphysis, magnetic resonance imaging, solid, cyst

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Introduction

Aneurysmal Bone Cyst (ABC) is a relatively common non neoplastic bone lesion. Jaffe and Lichtenstein [1] in 1942 first described this lesion as blood-filled cysts in which tissue from the cyst wall contained conspicuous spaces, areas of hemosiderin deposition, giant cells, and occasional bone trabeculae. The lesion was further defined by both of these authors [2,3] and became known as Jaffe-Lichtenstein disease. It occurs in patients between 5 and 20 years of age in 75% of the cases and approximately 60% occur in females [4,5].

The cause of this lesion is unknown. Changes in local hemodynamics related to venous obstruction or arteriovenous fistula have been implicated to play an important role. Some investigators believe that the lesion may be caused by a trauma. The lesion may arise de novo in bone or may be associated with various benign lesions like giant-cell tumor, osteoblastoma, chondroblastoma, chondromyxoid fibroma, fibrous dysplasia) or malignant lesions like osteosarcoma, fibrosarcoma, or chondrosarcoma.

ABC is predominantly a cystic lesion and tends to involve metaphysis with or without diaphyseal extension in the long bones. Purely diaphyseal location is uncommon. Also ABC is a predominantly cystic lesion with only a small solid component. In this case report, we describe a predominantly solid variant of ABC affecting purely the diaphyseal region of distal femur which is rare entity.

Case Report

A 24 years old male presented to the orthopaediacian with painful swelling over the middle aspect of right thigh since one month. No history of trauma was seen. On examination, swelling was tender and overlying skin was mildly red. Erythrocyte sedimentation rate and complete blood counts were normal. For further evaluation X-ray and MRI were advised. X-ray AP (Figure 1A) and lateral (Figure 1B) view of right thigh showed a large well defined central radiolucent expansile lesion with thinning of cortex, internal septations and scalloped margins involving the posterior aspect of mid diaphysis of femur. Associated soft tissue component was seen. MRI revealed a large lobulated lesion arising from posterior aspect of mid shaft of right femur, slightly hyperintense to muscle on T1 weighted images (Figure 2A). On T2 weighted images (Figure 2B and 2C), the lesion was heterogeneously hyperintense to

muscle with presence of few cystic areas in it (white arrows). No fluid-fluid or blood-blood level was seen. A provisional diagnosis of ABC was made. On histopathology, ABC was confirmed with presence of large solid component and mild cystic changes.

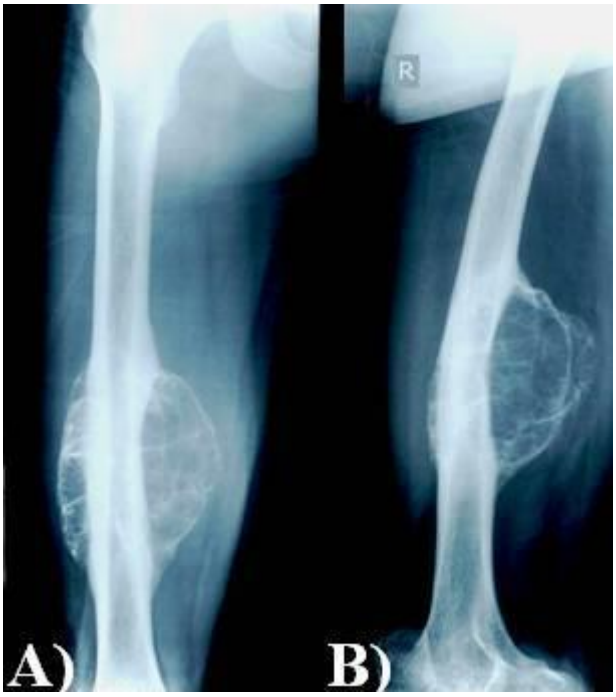

Figure 1. A) AP and B) Lateral X-ray views of right femur showing a large well defined central radiolucent expansile lesion with thinning of cortex, internal septations and scalloped margins involving the posterior aspect of mid diaphysis of femur.

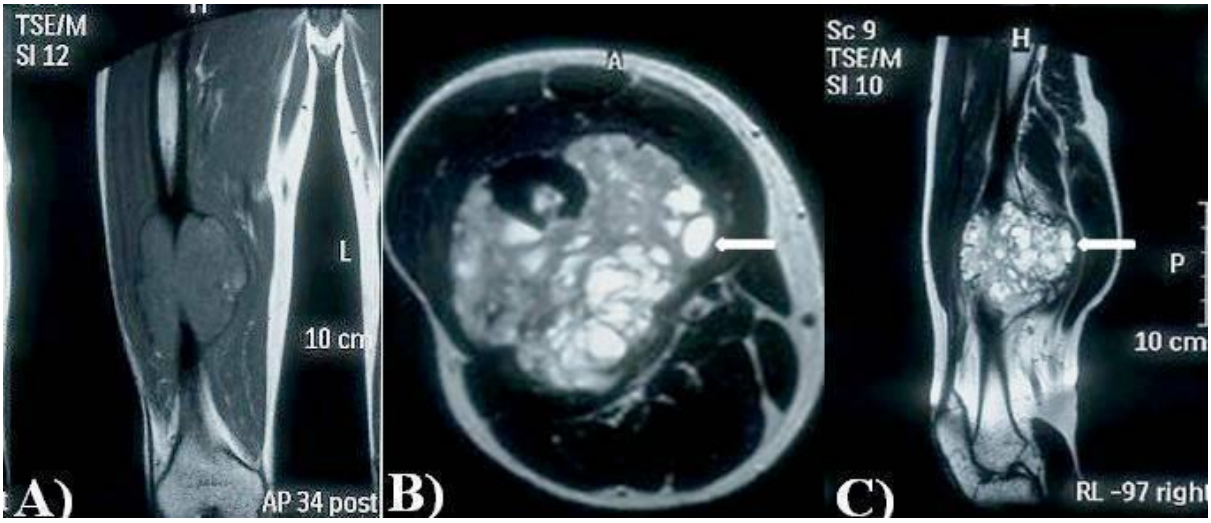

Figure 2. A) Sagittal T1 weighted MRI showing large lobulated lesion arising from posterior aspect of mid shaft of right femur, slightly hyperintense to muscle. B) Axial and C) Sagittal T2 weighted images showing a heterogeneously hyperintense lesion with presence of few cystic areas in it (white arrows).

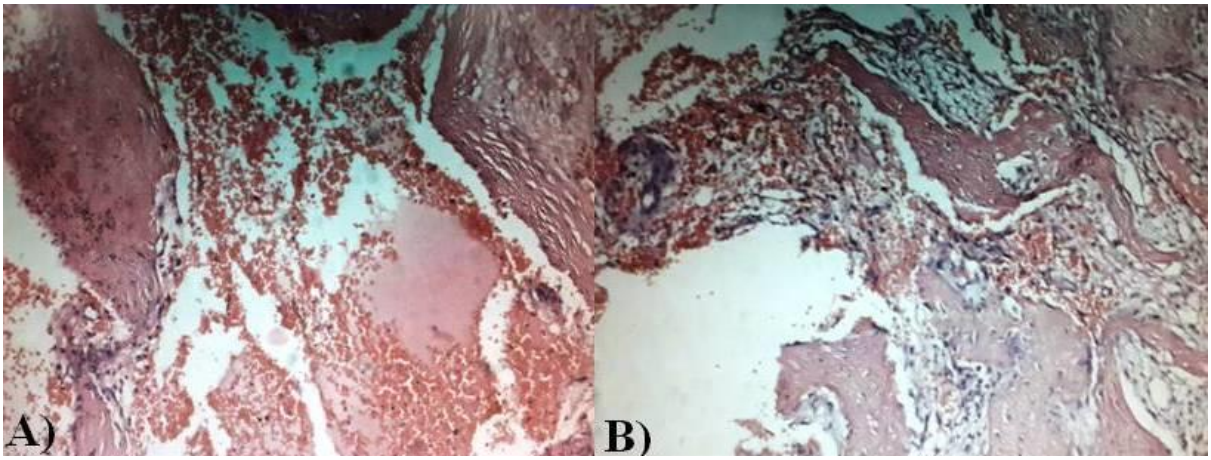

Figure 3. Photomicrographs A) and B) H&E Stain showing areas of haemorrhage, presence of osteoclasts like multinucleated giant cells at places and no endothelial lining. Intervening reactive bone formation is also seen.

Discussion

ABC is not an uncommon bone lesion. It is a non neoplastic bone lesion. Presenting complaints of patients with ABC is acute onset of pain at the affected site. Spinal ABC may produce neurological deficits because ABC has a predilection for the neural arch which might result in spinal stenosis and an extradural block. Systemic signs and symptoms are generally absent.

Some authors believe that ABC is actually a result of hemorrhagic degradative events occurring in patients with other lesions including giant cell tumor, hemangioma, chondroblastoma, osteoblastoma, nonossifying fibroma, fibrous dysplasia, chondromyxoid fibroma, eosinophilic granuloma, and other tumors [1,5,6,7,8-12].

The lesion is commonly seen in the first two decades of life. The lesion is mostly metaphyseal with diaphyseal extension and located eccentrically within the bone. Some cases may be purely diaphyseal. Eccentric location is an important distinguishing radiographic feature from unicameral bone cyst [13].

X-ray findings include the presence of blowout lesion showing marked thinning of the cortex with only minimal bone formation [13]. In the long bones, the lesion tends to be eccentric, creating a saccular protrusion with multiple fine septae internally and sharply demarcated, bulging, scalloped borders. On CT and MRI several fluid levels (fluid-fluid) can be seen within the multicystic lesion [14,15].These are probably due to settling of degraded blood.

Sanerkin et al. [16] described a variant of aneurismal bone cyst in which the predominant histology was that of the solid material of a cystic aneurysmal bone cyst; those authors used the term “solid variant of aneurismal bone cyst”. The histologic features of “solid” aneurysmal bone cysts and giant cell reparative granulomas are similar, and the terms have been used interchangeably [17]. Solid variant of ABC are primarily seen in the craniofacial and small tubular bones of the hands and feet [17].

Solid ABC appears on MR images a predominantly solid lesion with admixed cystic areas and fluid–fluid levels in some of them. In a study by Ilaslan H et al [17], most lesions (7/8) were slightly hyperintense to muscle on T1- weighted images and were heterogeneous, with

predominantly high signal intensity with scattered areas of low signal intensity on T2-weighted sequences, similar to our case.

Surgical curettage with bone chip replacement is the usual mode of therapy. Recurrence rates are high and range between 20-50%. Radiation therapy creates sclerosis of the vessels, helping reduce recurrence rates.

Thus we conclude that although predominantly metaphyseal, ABC can sometimes also occur purely in the diaphyseal location and may have solid appearance on MRI and therefore must always be considered a differential diagnosis whenever a diaphyseal predominantly solid lesion in long tubular bone is seen in young individuals.

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