Case Report

A Rare Case of Primary Aneurysmal Bone Cyst (ABC) of Patella with Histopathological Correlation

Garg Sahil¹, Patil Abhijit², Bhalla Mitesh³, Kuber Rajesh⁴

¹Second Year Resident, ²Associate Professor, ³First Year Resident, ⁴Professor, Department of Radiodiagnosis, Padmashree DY Patil Medical College and Research Centre, Pimpri, Pune.

Corresponding Author: Garg Sahil

ABSTRACT

Aneurysmal bone cyst (ABC) is a rare rapidly growing benign bone tumour and accounts for 1-2% of all primary bone tumour. It occurs commonly in shaft of long bones (most common in distal femur and proximal tibia) and vertebral column, with less than 1 % cases occurring in patella. They are usually primary but may occur secondary to other benign or malignant bone tumours. ABC usually affects young adults and presents as a solitary expansile lytic bone lesion. Primary ABC of the patella is very rare. Herein we present a 24 year old male presenting with pain and swelling and diagnosed with primary patellar aneurysmal bone cyst on Radiograph and magnetic resonance imaging (MRI) with its histopathological confirmation.

Keywords: Aneurysmal bone cyst, Patella, MRI, curettage, Osteoclast giant cells

INTRODUCTION

Aneurysmal bone cyst was first described by Jaffe and Lichtensein consisting essentially of fibrous honey combed appearance separated by vascular spaces. [¹] It is more common in the females and usually occurs in the first or second decades of life. [²] They have the predilection for metaphysis of long bones. ABC is a benign vascular tumor presenting as an expansile lytic lesion which may expand from the affected bone and involve the overlying soft tissues. Less than 1% of the aneurysmal bone cyst occurs in patella. [³,⁴] Histologically they are expansile osteolytic lesion filled with blood filled spaces of variable size separated by connective tissue containing bone trabeculae or osteoid tissue and osteoclast giant cells.

We present a case of primary Aneurysmal bone cyst of patella in a 24 year old male diagnosed on Radiograph and MRI further confirmed on excision and histopathological examination.

CASE REPORT

A 24 year old male complained of pain in the left knee since 1 month associated with tingling sensation. The patient presented with pain and difficulty in extension. There was history of trauma 1 year back due to fall from bike. On physical examination patient had mild tenderness without any overlying soft tissue swelling.
Plain radiography of left knee AP and lateral views revealed loss of bony trabeculations with multiple lucencies within patella. The bony cortices appear intact without any evidence of soft tissue involvement. The joint spaces were well maintained without any intra-articular extension of the lesion. (Figure 1)

MRI of the left knee joint was performed without administration of intravenous contrast. On T2 Sagittal and PDFS (Proton density fat saturated) there were hyperintense multiseptated cystic lesion in the patella (Figure 2). Subtle fluid - fluid levels were seen in the cystic lesion on T2WI. The lesion was predominantly in the medial aspect of the patella and also crossing the midline to the opposite side (Figure 3). Associated thinning of the anterior as well as posterior cortex was also seen in the patella, however without any cortical breech or surrounding soft tissue component.

Total patellectomy was performed which on gross pathological examination revealed spongy haemorrhagic mass covered by thin shell of reactive bone.(Figure 4 A)

Histopathological examination revealed large cystic spaces filled with blood and separated by fibrous septa, alternating with solid areas. Cysts and septa were lined by fibroblasts, myofibroblasts and histiocytes but not endothelium. Clusters of osteoclast like multinucleated giant cells
with loose spindly stroma to cellular stroma, reactive woven bone and degenerated calcifying fibromyxoid tissue was seen with variable mitotic figures and haemosiderin. No malignant osteoid and no atypia were noted. (Figure 4 B and C) These findings were consistent a primary aneurysmal bone cyst.

DISCUSSION
Aneurysmal bone cyst (ABC) are commonly primary than secondary with a ratio of 2:1 and associated with no other lesions. Secondary ABC occur with other benign or malignant lesions such as giant cell tumour, chondroblastoma, osteoblastoma and fibrous dysplasia which show haemorrhagic cystic changes. Secondary ABC is usually aggressive and have high recurrence rate. Primary aneurysmal bone cysts are less aggressive and are associated with history of trauma, pain and swelling. They are vascular benign solitary expansile lesions of the bones. They are more common in females than males and may affect any bone but mostly affect the metaphysis of long bones, commonly lower end of femur, upper end of tibia, humerus, flat bones and posterior elements of vertebral body with very rare location in the patella. They occur commonly in first or second decades of life and are found to be associated with people of African descent.

In our patient history of trauma was present and no history of associated neoplasm was present so primary ABC was considered rather than secondary aneurysmal bone cyst which was further confirmed on histopathological examination. In 1941, Jaffe HL and Lichtenstein L were first to describe aneurysmal bone cyst and the disease came to be known as Jaffe-Lichtenstein disease. According to them they used this term in relation to the blow out radiographic appearance and cystic spaces filled with blood. Many etiologies have been proposed but most commonly accepted pathogenetic mechanism of Aneurysmal bone cyst was found to be local circulatory disturbance causing increased venous pressure leading to development of dilated and enlarged delayed vascular bed in the affected bone. Recently chromosomal abnormalities have been proposed to be a causative mechanism. A recurrent t (16; 17) (q22; p13) gene has been identified as the causative chromosomal abnormality. Over expression of insulin like growth factor 1 has also been proposed as a causative mechanism.

Radiologically Aneurysmal bone cyst gives a ‘blown out appearance’ appearing as eccentric expansile lytic lesion with cortical expansion or expansion into the soft tissues. Peripherally the trabeculae appear coarse but become delicate at the centre of the lesion. Osteolytic lesion are surrounded by bony septa with formation of periosteal new bone formation at the surface of intraosseous border.

Histopathologically primary ABC shows blood filled cystic spaces separated by fibrous septa. The fibrous septa consists of moderately dense cellular proliferation of fibroblasts with scattered multinucleated osteoclast type giant cells and erosive woven bone rimmed by osteoblasts.

Most common differential diagnosis includes chondroblastoma and giant cell tumour and can be differentiated on basis of radiography, MRI, CT imaging and Bone scanning. Radiographically, chondroblastoma appears as an osteolytic, round or lobulated lesion with sclerotic rim and giant cell tumour occurs as an eccentric, expansile lesion with multiple septations and geographical pattern of destruction involving more than 75% of patella. CT and MRI helps in better delineation of expansion of bone, mineralization of bone, changes in cortex, internal architecture of tumour and associated soft tissue swelling.
Some authors suggest that plain radiography alone could diagnose the case of aneurysmal bone cyst without the aid of CT and MRI. [6]

Currently the Enneking staging system is used for standardization of operative procedure of patellar tumours. Stage I lesions have a well defined cortex, Stage II lesions have a thinned cortex, which may be partly broken but limited to the periosteum and Stage III lesions penetrate the cortex with small breaches around the perimeter. In our case there was thinning of cortex without any break in the bony cortex, so stage II was given pre operatively.

Intralesional curettage with bone grafting is the preferred treatment in stage I and II and total patellectomy with patellar prosthesis is preserved for aggressive stage III lesions. In a case study by Gibbs et al on 40 patients treated for ABC excision, 34 out of 40 patients were treated by curettage. Of the 22 patients who had cancellous bone autogenous grafting 12% had local recurrence. [10] Recurrence rate is higher in patients treated with curettage, bone grafting, cryosurgery, radiation or en bloc resection than total patellectomy. [3] To prevent the recurrence our patient was treated with total patellectomy.

CONCLUSION

In conclusion, ABC is a rare benign expansile osteolytic lesion with its rare location in patella with difficulty in its differentiation from other lesions radiologically and clinically. Detailed MRI examination with histopathological correlation helps in early diagnosis, adequate surgical planning and better evaluation of its prognosis and sequelae.

REFERENCES

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