

Huge chondromyxoid fibroma of proximal third tibia masquerading as an aneurysmal bone cyst: A rare case report

Sir,

Chondromyxoid fibroma (CMF) was described by Jaffe and Lichtenstein^[1] as a distinctive entity in 1948. Jaffe HL, Lichtenstein L^[1] in the masterly paper, not only set out a system of grading based on stromal differentiation, but also emphasized that the so-called variants have little in common with genuine giant-cell tumors or with one another. These “variants” have subsequently been defined as the benign chondroblastoma, CMF and aneurysmal bone cyst, while the so-called giant-cell tumors of tendons and synovia are probably granulomata rather than neoplasms.^[2-4]

It occurs mostly in the second and third decades with no difference in the sex incidence. It presents like most benign bone tumors with pain, slowly increasing localized swelling, sometimes accompanied by the presence of a palpable tumor mass and tenderness on palpation. In long bone, the tumor is found in or near the metaphysis and sometimes crosses the epiphyseal line only rarely and in advanced cases. Proximal end of tibia is by far the most common site reported.

Radiologically the lesion is radiolucent, eccentric, space occupying, and is located in the metaphysis, margins are usually well-defined with surrounding sclerosis with ill defined margins between sclerosis and the host bone. The cortex is expanded considerably, peripheral bony margins often become hazy and poorly defined. Unlike other cartilaginous neoplasms calcification within the lesion is very uncommon. Computed tomography (CT) scan imaging may be necessary to delineate a cortical margin in the expanded soft tissue mass. The radiologic differential diagnosis includes giant cell tumor, aneurysmal bone cyst, unicameral bone cyst, chondroblastoma, and fibrous dysplasia.^[5]

CMF resembles fibrocartilage grossly. It has a sharp border often with an outer surface of thin bone or periosteum. The glistening grayish white lesion is firm and lobulated. It may also have small cystic foci or areas of hemorrhage.

Histopathologically can be confused with chondrosarcoma.^[2,4]

Recognition of the tumor depends largely upon the histological appearance; it consists of mixture of fibrous myxoid and chondroid areas of varying maturity with

increased cellularity at the periphery. There are occasional foci of calcification, giant cell, and irregular nuclei. The sharp borders of each lobule and the lesion itself help to differentiate it from chondrosarcoma.

The patient, a girl aged 15 years when first seen, presented with a swelling of the upper end of the right tibia, which she had noticed 1 year back. No medical advice was taken and the swelling gradually increased, which made her report to our hospital. She did not complain of any discomfort or pain due to the swelling but an increase in size was the question of worry. Her general health was good and she had not suffered any previous illness of significance. The family history was likewise negative with no such complaints to anyone in the family.

On examination a diffuse oval swelling of 16 × 8 cm was present on the upper end of right tibia, the overlying skin was normal and the swelling was nontender on palpation [Figure 1]. The swelling appeared to be fixed to the underlying bone with no localized rise of temperature. General examination revealed no other abnormality except pallor, and routine investigations were all normal except for mild anemia with hemoglobin of 8.1 g%.

Radiographic examination showed osteolytic radiolucent eccentric lesion with extensive expansion noted in the metaphysis at the medial border of proximal tibia [Figure 2]. Lateral border of the lytic lesion had sclerotic margins with no evidence of periosteal new bone formation or radiologically evident calcification. Cortex was thinned out, however, there was no radiological breach. Zone of transition was narrow and there was no evidence of calcification in the lesion.

A diagnosis of an aneurysmal bone cyst was suggested by the radiologist.

An incisional biopsy under spinal anesthesia was performed from the right proximal tibia and a diagnosis of an aneurysmal bone cyst was suggested by the



Figure 1: Preoperative clinical picture showing swelling over the proximal end of right leg

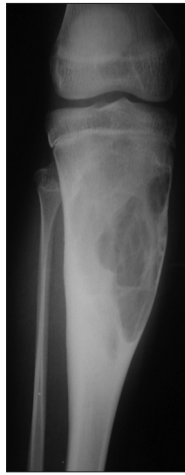


Figure 2: Preoperative radiographs showing an osteolytic lesion at the upper end of right tibia in the metaphyseal region

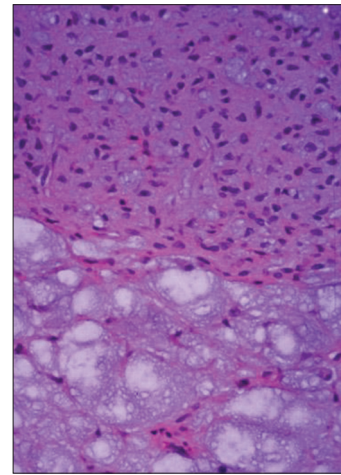


Figure 3: Histological picture of chondromyxoid fibroma



Figure 4: Postoperative clinical photograph after 1 year



Figure 5: Radiograph of the patient during follow up showing complete healing of lesion

pathologist.

A preoperative diagnosis of an aneurysmal bone cyst was thus made and patient was operated for excision and extensive curettage with bone grafting. Intraoperatively the lesion contained yellowish gray material, which was friable on handling and removal in one piece was not possible. Hence, the lesion was curetted and was packed with bone graft harvested from the contralateral fibula. Due to the extensive curettage and the suspicion of creating a pathological fracture, an external fixator was applied to provide stability. The wound was closed in layers and a negative suction drain was applied.

The intraoperative findings were not consistent with those of an aneurysmal bone cyst and the curetted material was again sent for histopathological examination, which was reported by the pathologist as a CMF [Figure 3]. At 2 months, there was no recurrence of the lesion and the patient was able to mobilize, bearing full weight on the affected limb with no deformity [Figure 4]. Fixator was removed after 3 months. The tumor was completely replaced by normal bone by 6 months. No evidence of

recurrence was noted on X-ray taken 1 year after surgery [Figure 5].

Clinical presentation of CMF is almost similar to any benign tumor arising from bone. As evident from this case report, there do exist some striking similarities between CMF and aneurysmal bone cyst, which on many occasions lead to misdiagnosing this rare tumor as an aneurysmal bone cyst, which is more common in occurrence.

Takenaga *et al.*^[6] reported two cases of CMF presenting as subperiosteal lytic lesion. They have recommended that subperiosteal CMF, although rare, should be included in the differential diagnosis of a painful, radiographically inactive lytic lesion on the surface of a long bone. Other differential diagnosis can be giant cell tumor, aneurysmal bone cyst, unicameral bone cyst, chondroblastoma, and fibrous dysplasia. Magnetic resonance imaging (MRI) examination is not helpful to confirm the diagnosis but helps in knowing the extent of spread of tumor.^[5]

CMF and aneurysmal bone cyst resemble histologically due to the presence of numerous giant cells, which are

consistent features of a CMF but the highly vascular structure of an aneurysmal bone cyst may be absent in a cyst with more of solid material, the basic pattern of a CMF shows wide variations even within the same tumor and the final diagnosis depends upon the examination of several sections.^[2,4] Histologically it has appearance very similar to chondrosarcoma. Presence of organized matrix and less cellularity differentiate it from chondrosarcoma. The difference in opinion regarding the diagnosis in this case may be due to inadequacy of material as provided by an incisional biopsy.

It is benign tumor with high rate of recurrence when treated with curettage of lesion. Curettage and bone grafting has very low rate of recurrence. Gherlinzoni *et al.* reviewed the records of 27 patients with the diagnosis of CMF at the Bone Tumour Center of the Istituto Ortopedico Rizzoli. Surgical treatment was evaluated in 22 patients with a follow-up of longer than 2 years; their average follow-up was 117 months (range, 24-380 months). Seventeen patients had been followed up for at least 5 years. There was an over-all rate of recurrence of 27%. With curettage alone there was an 80% recurrence rate, but when curettage was combined with corticocancellous bone-grafting the recurrence rate decreased to 7%. The recurrence rate was not found to be dependent on the age of the patient or on an atypical histological appearance.^[3] On the basis of this experience, we believe that thorough curettage combined with corticocancellous bone-grafting remains the treatment of choice for patients with chondromyxoid fibroma.

Malignant transformation is very rare.^[2,7] There are only occasional reports of malignant transformation. Scaglietti and Stringa^[7] have drawn attention to the locally aggressive behavior of this tumor in the young and have suggested a more radical form of local resection in its treatment.

With the above discussion, we emphasize on the need

to include this lesion in the differential diagnosis of a painful, radiologically lytic lesion of a bone and the need for correlation between the pathologist and the clinician for the correct diagnosis of this rare benign tumor as was done in this case. Treatment with curettage and bone grafting is successful with very low incidence of recurrence.

Rajesh Dulani, Samir C. Dwidmuthe,

Sandeep Shrivastava, Pradeep Singh, Sharad Gupta

Department of Orthopaedics, Acharya Vinoba Bhave Rural Hospital and Jawaharlal Nehru Medical College, Datta Meghe Institute of Medical Sciences (Deemed to be University) Sawangi (M), Wardha, Maharashtra, India

Correspondence to: Dr. Samir Chandrakanth Dwidmuthe,

E-mail: samirdwidmuthe@gmail.com

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