

Case Report

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Juxtacortical Chondromyxoid Fibroma of Tibia

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Abstract

Introduction: Chondromyxoid fibroma, CMF, is the least common benign cartilaginous tumor composed of chondroid and myxoid matrix. It is usually located eccentrically in distal femur or proximal tibia metaphysis. Juxtacortical Chondromyxoid fibroma which may be seen in unusual places such as intracortical, or sub periosteal locations is very unusual.

Case report: A 16-year-old boy presented to us with a mildly painful distal tibial mass for one year. Imaging studies showed a protruded mixed radio-opaque, radio lucent lesion at posterior surface of distal tibial metaphysis, without soft tissue involvement. A biopsy was performed which showed Chondromyxoid fibroma. Curettage and bone grafting was done as the therapeutic modality.

Conclusion: Chondromyxoid fibroma is usually not considered in the differential diagnosis of a painful, superficial lesion on a long bone. Other tumors such as periosteal chondroma, parosteal or periosteal osteosarcoma, parosteal myxoma, sub periosteal ganglion cyst, or periosteal osteoid osteoma are usually considered. Our case along with similar cases has proved that Juxtacortical CMF should be included in the differential diagnosis of a surface bony lesion.

Keywords: Chondromyxoid fibroma; Juxtacortical; Tibial metaphysis

Introduction

Chondromyxoid fibroma CMF was first described by Jaffe and Lichtenstein [1]. It is a rare tumor which comprises less than 1% of all benign bone tumors, and it is the least common benign cartilaginous tumor of bone. It usually presents during the second and third decades of life, and has a tendency for the metaphyseal region of the distal femur and proximal tibia [2]. Chondromyxoid fibroma may occasionally appear as a surface lesion. This includes tumors which are intracortical, sub periosteal, periosteal, or parosteal. Intracortical involvement is more common [3-5]. As it is not always possible to determine the precise origin of these surface lesions, the term Juxtacortical includes comprehensively all of these surface locations [6]. Periosteal Chondromyxoid fibroma has been reported to have imprint cytology as a method of confirmation [7]. We here present a new case of Juxtacortical Chondromyxoid fibroma.

A 16-year-old boy presented to us with a painful swelling above his right ankle. This swelling had been gradually increasing in size, and present for year. On physical examination, the patient had a tender, fixed, bony hard swelling on the distal part of the right tibia. The lesion was not fixed to the skin and was not associated with any ulceration, rise of temperature or sinus formation. The swelling was diffuse and large. Neurovascular status of the right lower extremity was normal. The patient did not have any other symptoms or any abnormal findings on systemic physical examination. His past

medical, family, allergy and drug, and social histories were not relevant. The laboratory tests including CBC, ESR, and CRP were normal. Plain radiographs of the right tibia and ankle in antero-posterior and lateral views showed a protruded superficial calcified mass in the posterior aspect of distal tibia (Figure 1A).

MRI of the right ankle and tibia showed T2 image with lobulated high signal lesion at the posterior aspect of distal tibia compressing posterior soft tissues including the Achilles tendon. The lesion was seen as a low signal lobulated mass on T1 images. There was no evidence of soft tissue component around the lesion (Figures 1B-1D). Imaging files of the patient were reviewed by an expert bone radiologist in Shafa orthopedic hospital, and together with the clinical information, a differential diagnosis of CMF, parosteal osteosarcoma, periosteal osteosarcoma, periosteal chondroma, periosteal myxoma, and sub periosteal ganglion cyst were proposed by orthopedic oncologists and bone radiologists in our hospital. Because malignant tumors were in the differential diagnosis, we decided to perform an incisional biopsy, to confirm the diagnosis before the final treatment. Histopathology exam of the biopsy specimen confirmed the diagnosis of Chondromyxoid fibroma (Figures 2A & 2B).

The patient was treated by en bloc resection of the tumor. No attempt was made for the reconstruction of the distal tibia, because three cortices were intact at the end of surgery. The patient was

asked to follow a non-weight bearing ambulation for two months. At the latest follow-up, twenty-five months after the surgery, the patient was free of symptoms and with no recurrence. This study

was approved by ethical committee of our institution and written consent was obtained from the patient and his family to report the case.

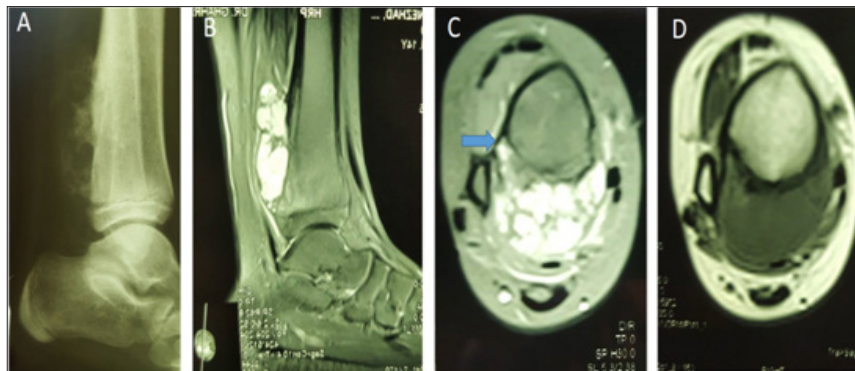


Figure 1 : A, Plain radiography of the right ankle showing a protruded radio-opaque radio-lucent lesion protruding from posterior aspect of distal tibia; B, C, and D, magnetic resonance imaging of the right ankle demonstrating a superficial lesion with high signal intensity on T2(B&C), and low signal on T1(D). The arrow shows that the lesion is within cortex, not involving intramedullary canal.

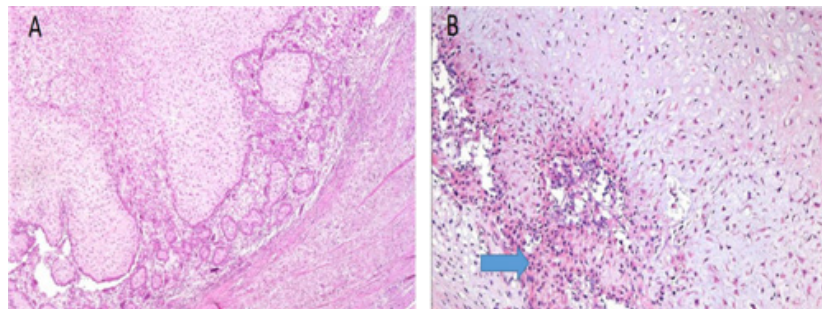


Figure 2 : A, Photomicrographs of the resected tumor showing lobulated architecture of CMF (H and E x 10); B, Higher magnification of a nodule showing stellate cells in loose myxoid stroma at center and increased cellularity at the periphery (arrow) (H and E x 40).

Discussion

There are some unique demographic and morphologic features attributed to Juxtacortical CMF, which are different from classic CMF. In a series of patients which include six sub periosteal CMF; four out of six patients were older than 30 years [4]. In another series, 13 out of 20 patients with Juxtacortical CMF were older than 30 years [8]. This is in contrast to the conventional CMF which usually presents before the age of 30 [2]. Juxtacortical CMF very commonly occurs in male gender [4]. In a study including 20 patients with Juxtacortical CMF, only 40% of them were female [8]. In another study including six patients with superficial CMF were male [4]. Our patient was also a male adolescent, so was another patient in a similar case report [9]. In classic CMF this male predilection is very slight (1.1:1) [2]. The most common anatomic location of Juxtacortical CMF is in long bones of lower extremity, especially in tibia. Ten out of 20 patients with Juxtacortical CMF reported by Allyson C. Baker et al. had their tumor in tibia, and 8 of them in femur, making these two bone the host of 80% of Juxtacortical CMF in that series [8]. In another series, and 4 out of 6 cases (66%) of sub periosteal CMF occurred in tibia [4]. Our case was also located in tibia. Although long bones are the most common place of presentation for conventional CMF, this preference is not as strong as it is in the Juxtacortical variant. Only 46.6% of conventional CMF have been found in long bones, and 30.3% of them is found in flat bones [2].

Radio logically; there is prominent calcification within Juxtacortical Chondromyxoid fibroma. This feature is seen much more commonly in the Juxtacortical than the conventional subtype of CMF [10]. This presentation is in contrast to the traditional view held by many radiologists that CMF does not show distinctive internal calcification [8] as conventional CMF is typically purely lytic, with occasional instances of intralesional calcifications [2,4,11]. The presence of prominent calcification may be the result of the sub periosteal location of this tumor and the resulting irritation of the periosteum during tumor growth. Histologically, the classic Chondromyxoid fibroma has stellate or spindle shaped cells arranged in lobules on a Chondromyxoid background. Within the lobules, the neoplastic cells tend to be localized more toward periphery. The lobules are separated by fibrous bands that contain blood vessels and sometimes multinucleated giant cells [4,11-13].

Many of these lesions could be treated by simple curettage. However, incomplete removal may lead to recurrence. En block excision is another mode of therapy, allowing complete removal of the lesion [11,14]. This is a necessity when facing aggressive tumors. Radical procedures should be avoided because it is a benign lesion [11,14]. If the tumor is near physis, it seems logical that the procedure be delayed until the tumor grows away from the physal area.

Conclusion

When confronted with a superficial lesion on a long bone, orthopedic surgeons, pathologists, and radiologists are more likely to consider osteochondroma, periosteal chondroma, periosteal myxoma, sub periosteal ganglion cyst, or sub periosteal osteoid osteoma in younger age group of patients. In older age group, more aggressive cartilage containing neoplasms, namely, periosteal chondrosarcoma, periosteal osteosarcoma, and parosteal osteosarcoma are included in the differential diagnosis. However, our case and the previously reported cases showed that CMF should also be included in the differential diagnoses of a surface neoplasm's of bone [4,8]. When one is aware of this lesion and the characteristic radiographic findings, a biopsy of the tumor can be diagnosed with more precision.

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