



Case Report

A rare case of chondromyxoid fibroma of distal phalanx of great toe

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ABSTRACT

Chondromyxoid fibroma (CMF) is rare benign tumour of bone which are due to incomplete cartilage differentiation and first described by Jaffe and Lichtensein in 1948. CMF's constitutes for less than 1% of all bone tumors. Histopathology is diagnostic and CMF show lobulated areas of spindle shaped or stellate cells with hypocellular center of chondroid or myxoid material and periphery is hypercellular with abundant fibromyxoid tissue and few multinucleated osteoclasts like giant cell. CMFs are commonly seen in metaphysis of long bones including proximal tibia or distal femur. Short tubular bones of hand and feet are uncommon sites and toes constitutes less than 5% of the tumors. We present a case of chondromyxoid fibroma of distal phalanx of left great toe in 35 years old male patient.

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1. Case Report

We here present a case of 35 years old male who comes to OPD with chief complaints of pain and swelling of the left great toe which was gradually progressive for few months. The pain was mild in intensity and was associated with difficulty while walking. Local tenderness was present and overlying skin was normal. X-ray showed lytic lesion on great toe. (Figure 1) On MRI of left foot there was presence of altered signal intensity lesion in the great toe along plantar aspect and distal phalanx and causing marginal erosion of distal phalanx. (Figure 2) The diagnosis of Giant cell tumour of tendon sheath was considered on radiological findings. The mass was excised and sent for histopathology.

Grossly excised specimen was single grey white to pearly white mass measuring 3×4×1cm. It was firm in consistency and cut section was grey white to pearly white.

Histopathological examination showed lobules of chondrocytes in lacunae, stellate cell in myxoid background showing oval to spindle shaped nuclei with mild pleomorphism, occasional mitosis, hypocellular in the centre and hypercellular in the periphery with occasional giant cells separated by spindle cells. (Figure 3 a, b)

Curettage has been done in our case. The patient was followed up for six months and there is no complain of pain or tenderness and follow up X-ray did not reveal any defect and scar has healed well.

MRI images in coronal plane show a lobulated expansile lesion with geographical bone destruction involving the distal phalanx of the big toe with a well-defined low signal intensity sclerotic margin. The lesion appears iso to hypointense on T1W1 Figure 2 a and heterogeneously hyperintense on T2WI Fig.2b with intense post-contrast enhancement Figure 2 c.

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Figure 1: Anteroposterior and Lateral view X-ray of right foot showing lytic lesion on great toe

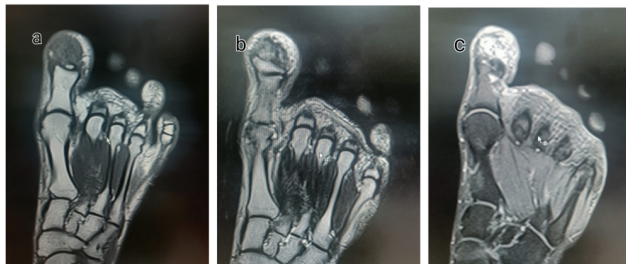


Figure 2:

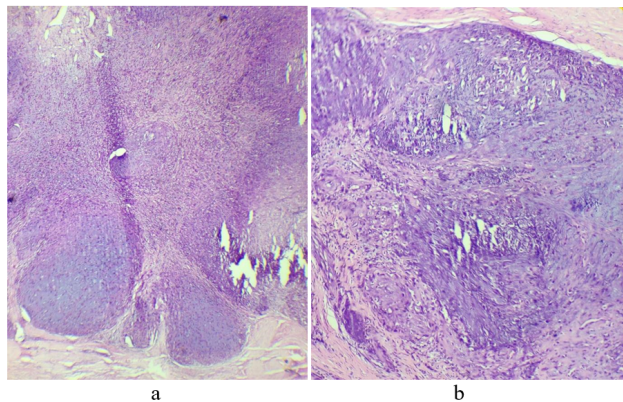


Figure 3: a): Microphotograph showing lobular growth; b): Lobules showing hypocellular centre with myxoid material and condensation of cells towards periphery

2. Discussion

Chondromyxoid fibroma (CMF) is a rare benign tumor of cartilaginous origin. The most common age of presentation is in second or third decade of life. Males have higher incidence than females. CMF is most commonly seen in metaphysis of long bones around the knee in proximal tibia, proximal fibula or distal femur. The short tubular bones of feet are rarely involved and is commonly seen in the adults in the third decade.¹⁻⁴

The patients present with clinical complaints of chronic pain, swelling, restriction of mobility and sometimes with pathological fractures. Radiological investigations show lytic, destructive lesion which is eccentric and sharply demarcated from the adjacent normal bone. There may be thinning of cortex and the lesions can destroy trabecular bone.⁵ Definitive diagnosis of CMF is made only on histopathology. The histopathological examination shows a lobular arrangement of stellate cells in myxoid and chondroid background. The lobules are hypocellular in center with hypercellular fibromyxoid tissue in the periphery. Osteoclast like giant cells can be seen in the periphery.^{6,7}

The differential diagnosis of Chondromyxoid fibroma includes chondrosarcoma, Giant cell tumor, Enchondroma, Chondroblastoma, Aneurysmal bone cyst and Simple bone cyst.

The main differential diagnosis is chondrosarcoma and it is very important to distinguish Chondromyxoid fibroma from chondrosarcoma to avoid misdiagnosis and aggressive treatment. In chondrosarcoma extensive and irregular cortical destruction with soft tissue involvement is seen in radiology. Histopathology examination of both shows lobular pattern, myxoid stroma and peripheral hypercellularity with nuclear atypia; however, in chondrosarcoma mature hyaline cartilage is seen and chondrosarcomas are more monotonous, have larger lobules, show abundant myxoid ground substance and less fibrous component. Sometimes chondrosarcoma diagnosed as chondromyxoid fibroma were undertreated which results in worse prognosis later on.^{8,9}

Giant cell tumors lack chondroid differentiation and they show peculiar arrangement of osteoclast like giant cells with stromal fragments.

Enchondroma is another differential diagnosis and shows paucicellular cartilage with mottled calcification on histology and are commonly seen in fingers and toes.¹⁰

Low grade infections sometimes can be confused with CMF in radiology but periosteal reaction is commonly seen in infections and chondromyxoid fibroma shows commonly cortical breach.¹¹

The treatment options are curettage alone, curettage with bone grafting, en bloc excision or amputation.¹² Long term follow up is recommended to look for recurrence of tumor and malignant transformation. Amputation may be

considered in recurrences or malignant transformation.

3. Conclusion

Chondromyxoid fibroma is an uncommon neoplasm of the bone with cartilagenous differentiation commonly found in tibia or femur. Chondromyxoid fibroma of toe is extremely rare. It should be considered in the differential diagnosis of a solitary bone lytic lesion that has well-defined scalloped margins, and differentiated from other tumors, especially from chondrosarcoma and giant cell tumour to avoid aggressive treatment. Histopathology provides the definitive diagnosis. Long term follow up is necessary to look for recurrence and malignant transformation.

4. Source of Funding

None.

5. Conflict of Interests

There are no conflict of interests regarding the publication of this paper.

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