

# Skeletal Myxoid Chondrosarcoma of the Fibula

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## ABSTRACT

Chondrosarcoma of the bone is second most common malignant bone tumor next only to osteosarcoma. The most common type of chondrosarcoma is the conventional chondrosarcoma (90%). The myxoid variant is a rare occurrence and is associated with a worse outcome as opposed to conventional type. There are only a few references of this tumor in literature. The most common location for skeletal myxoid chondrosarcoma is the femur. There is similarity in light microscopic features in extraskeletal chondrosarcoma and skeletal myxoid chondrosarcoma there exist elementary differences at ultrastructural and molecular levels. Skeletal myxoid chondrosarcoma presents as a well-defined osteolytic lesion with endosteal scalloping, cortical invasion and expansion on radiography. Myxoid chondrosarcoma of the bone has a more aggressive clinical course than conventional intramedullary chondrosarcoma, with patients commonly developing distant metastases and local recurrence. The purpose of our article is to report this rare myxoid chondrosarcoma of the proximal fibula, which has not been reported at this anatomical site previously with an emphasis on its radiological features and histopathological features.

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## INTRODUCTION

Chondrosarcoma is a ubiquitous entity of the skeletal system and constitute 9% of primary malignancies of bone, an incidence about half that of osteosarcoma. They are the second most common malignant tumors of the bone after osteosarcomas. These are generally low to intermediate grade tumors with a predisposition towards men in age group of 20 to 60 years. The most common type is the conventional chondrosarcoma (90%). The myxoid variant is a rare occurrence and is associated with a worse outcome as opposed to the conventional type. The former has frequently been found and reported to be an extra skeletal tumor. They are so similar histologically that often radiology helps to make the final diagnosis. However, number of case reports describing skeletal myxoid chondrosarcoma (SMC) is scant. We here present a rare case of skeletal myxoid chondrosarcoma of proximal fibula.

## CASE REPORT

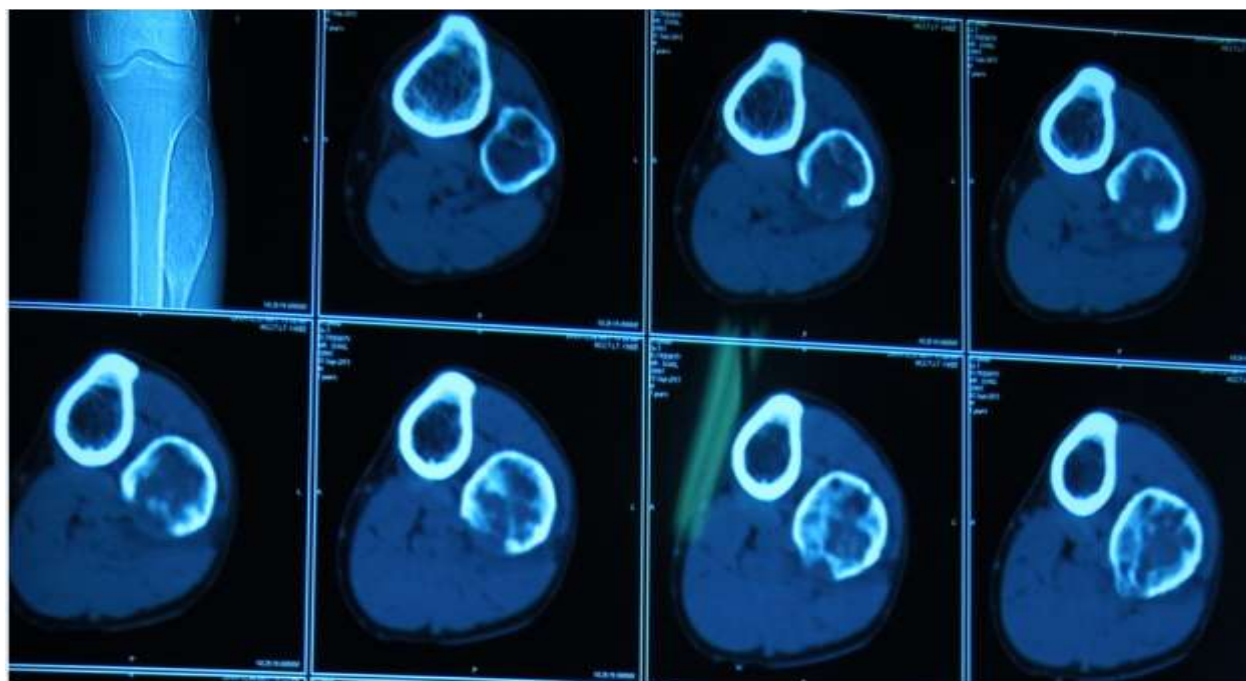
A 21 year old male presented with a slowly growing globular lump on his left upper leg, just below the knee. Although the swelling had been present for last 15 years, it had started to grow in size and had started to pain for the past few weeks. The pain was of insidious onset, dull in nature and it would increase on bearing weight. There was no history of ant preceding trauma. The swelling was not associated with restriction of movements, symptoms suggestive of distal neurovascular compromise or any loss of function. There were no symptoms suggestive of any infective etiology.

On clinical examination the lump was non tender and located in the region of the upper third of the left leg on lateral side. It was globular in shape and measured 12 cm in length and 11 cm in breadth. The overlying skin was normal with no signs related to presence of any underlying inflammation. On palpation the swelling was not mobile, was bony hard in consistency, non pulsatile, non reducible and non compressible. The distal neurovascular status was normal. There was no restriction of ipsilateral knee and ankle joint movement. Differential diagnosis of giant cell tumor, bone cyst and parasitic cyst were kept in consideration.

The radiographs (of left knee, ankle and leg – antero-posterior and lateral views) revealed fusiform, lytic defect with scalloping of the inner cortex with a periosteal reaction in the head of fibula measuring about 11 cm in length and 9 cm in breadth. On computed tomographic scan it was that there was no involvement of the surrounding soft tissues. Complete blood count, erythrocyte sedimentation rate, renal and liver function tests, and other basic haematological investigations were done and were within normal limits. [Figure - 1 and 2]



**Figure -1: Radiographs of the knee and leg showing the expansile lytic lesion in proximal fibula. The lesion is fusiform with scalloping of inner cortex.**

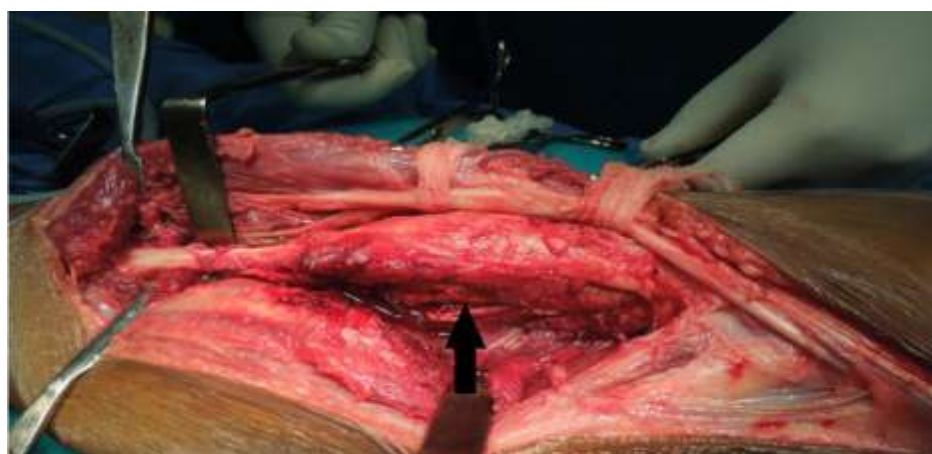


**Figure -2: CT scan of part showed no involvement of the surrounding soft tissues.**

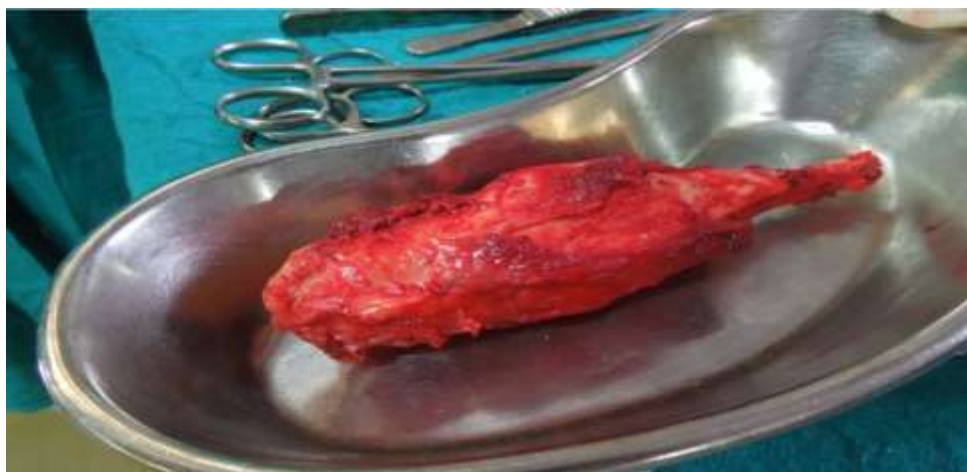
An excision biopsy was planned for further evaluation. Per-operative findings showed a lobulated multinodular mass, well circumscribed by a distinct fibrous capsule which was excised en-mass with the head of the fibula. The size of the resected specimen was 10 cm in length and 8 cm in breadth. [Figure - 3 to 6]



**Figure -3:** Peroperative picture shows that lateral peroneal nerve has been safely retracted away from the lesion. Black mark shows the nerve and red mark shows the tumor mass.



**Figure -4:** Peroperative picture shows that the lesion/ tumor has been completely dissected out. Black arrow shows the tumor mass.

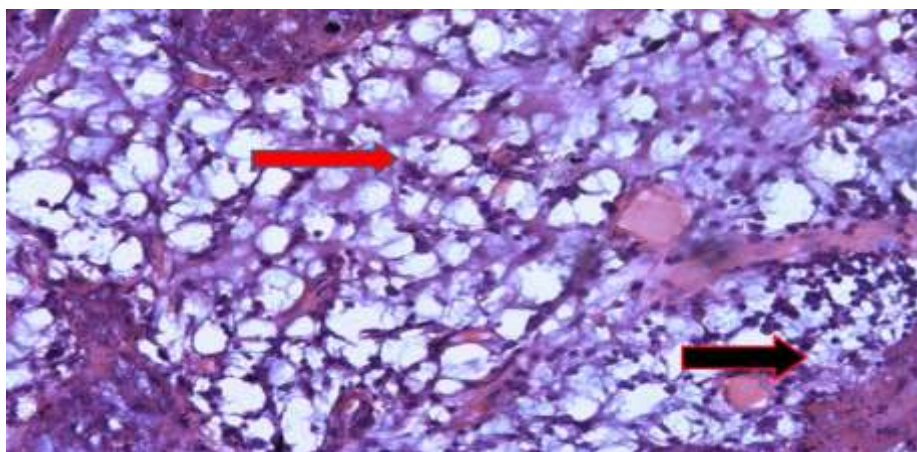




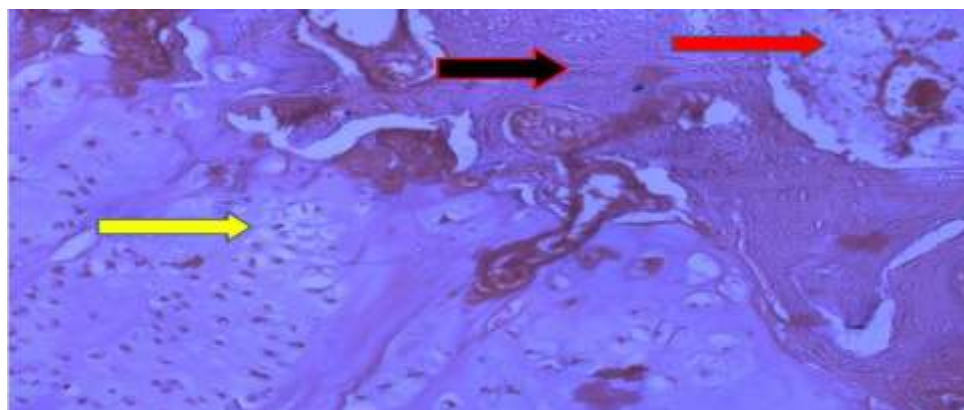


**Figure- 5 and 6: Excised tumor mass with the proximal fibula.**

On histopathological examination with haematoxylin and eosin staining, it showed typical tumour nodules, composed of round and slightly elongated cells, with features of chondroblasts, separated by mucoid substance. There were occasional differentiated cartilage cells. Additionally, there was presence of typical stellate cells with basophilic myxoid matrix. [Figure -7 & 8]



**Figure 7: Chondrosarcoma (20x) with myxoid change, represented by black arrow and stellate cell surrounded by frothy basophilic matrix, represented by red arrow.**



**Figure 8: Mxyoid chondrosarcoma (4x) with cartilage cells, represented by yellow arrow, cartilage cap, represented by black arrow, and lamellar bone represented by red arrow.**

## DISCUSSION

Myxoid chondrosarcoma is a rare, low-grade, indolent tumor that can occur in soft tissues and bone. It is, however, capable of distant metastases. Primary SMC was not fully evaluated initially as a distinct clinic-pathologic entity, and the term was/used interchangeably to designate either a conventional chondrosarcoma with prominent myxoid degeneration or a myxoid sarcoma that is identical histologically to extraskeletal myxoid chondrosarcoma (EMC). [2] Previous cytogenetic data have showed a translocation, t (9; 22) (q22-31; q12), occurring in about half the cases of the extra skeletal variant of the disease. [2] The skeletal myxoid chondrosarcoma further is an exceedingly rare neoplasm with a distinct histopathology and to the best of our knowledge this has not yet been reported in fibula.

Chondrosarcoma of the soft tissues, originally reported by Stout and Verner in 1953, was described to show similar histologic features to those arising in bones.[3] Based on two cases, Kilpatrick et al. were the first to attempt to separate the primary SMC into two distinct categories: conventional chondrosarcoma with marked myxoid degeneration, and the so-called "chordoid sarcoma".[4] The conventional osseous chondrosarcoma with myxoid change appeared microscopically to have neoplastic cells set in distinct lacunar spaces, suggestive of hyaline cartilage formation. No quantitative assessment was provided regarding the amount of myxoid change that had occurred.

Antonescu et al. reported the most common location for SMC to be in the long bones of the lower extremity. In half of the cases, the tumors were clustered around the hip joint, with 30% in the pelvic bones and 20% in the proximal femur; and in only 15% of cases the tumor located around the knee joint. Fifteen percent of cases presented in the shoulder girdle involving proximal humerus, clavicle, and scapula. Only one case reportedly involved the craniofacial bones (the maxillary antrum). [2] SMC and EMC do not constitute a single entity arising in two different locations. Although they do have overlapping microscopic features there exist significant differences at the ultrastructural level and especially at the molecular level. Specifically, the t(9;22) found in EMC has never been found in a clear case of SMC. [2]

Microscopically, SMC is composed almost entirely of a myxoid matrix, with only minimal hyaline cartilage formation [5]. The tumor has very high water content, related to the extensive myxoid stroma and better-differentiated areas of hyaline cartilage. The conventional osseous chondrosarcoma with myxoid alteration appears microscopically to have neoplastic cells set in distinct lacunar spaces, suggestive of hyaline cartilage formation. On the other hand, the SMC was described to be distinct from the usual chondrosarcoma of the bone, most probably representing the skeletal counterpart of EMC [4]. In some references, skeletal and extraskeletal myxoid chondrosarcoma are described to be morphologically identical, and imaging studies are necessary to confirm the bone or soft tissue origin of the tumor [6]. In another study, skeletal myxoid chondrosarcoma is said to be morphologically distinct from the extraskeletal counterpart by virtue of the lobulated, multinodular appearance, comprised of a uniform population of rounded to slightly spindled cells. [3]

SMC on radiographic examination shows up as a hazy, rarefied, destructive lesion, mostly devoid of intralesional calcifications with endosteal scalloping, cortical invasion and expansion. Concentric cortical thickening due to periosteal reactive bone formation has also been noted in most of the long bone tumors but is less apparent in those of the pelvic bones. All these observations correlate with the presentation in our case. [4] Histologically, CMF seems quite similar to chondrosarcoma. They are so similar histologically that often radiology helps to make the final diagnosis. SMC may be distinguished from chondromyxoid fibroma mainly by the sharply demarcated radiographic area of bone destruction and histologically by the lack of stellate and spindle cells randomly arranged within fibromyxoid stroma and accompanied by multinucleate giant cells. The sharp borders of each lobule and the lesion itself help to differentiate it from chondrosarcoma. [7]

The treatment of choice for chondrosarcomas is wide or radical resection or amputation. In tumours in expendable location primary wide resection is advised without a biopsy to decrease the risk of tumour contamination. After wide resection rate of recurrence is 10 %. Kawaguchi et al. reported that wide excision for myxoid chondrosarcoma was effective for achieving local tumor control. Chemotherapy and radiation therapy have limited roles in cases of chondrosarcoma, as the disease is generally insensitive to these treatments. Although, radiation therapy may be used if a tumor has been excised incompletely or is located in surgically inaccessible region. [7]

To conclude, we report a case of skeletal myxoid chondrosarcoma of proximal fibula which has been reported to have a more aggressive clinical course than conventional myxoid chondrosarcoma, with patients commonly developing distant metastases and local recurrence [3].

## **REFERENCES**

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- [7]. Kawaguchi S, Wada T, Nagoya S, Ikeda T, Isu K, Yamashiro K, et al. Extraskeletal myxoid chondrosarcoma: a Multi-Institutional Study of 42 Cases in Japan. *Cancer*. 2003; 97:1285–1292. Figure 1: Chondrosarcoma (20x) with myxoid change, represented by black arrow and stellate cell surrounded by frothy basophilic matrix, represented by red arrow.