Myxoid Chondrosarcoma of the Hand: An Unusual Skeletal Presentation with Review of Literature

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Abstract

Introduction: Myxoid chondrosarcoma is a distinct subtype of chondrosarcoma with several key differences from conventional chondrosarcoma of bone. We have searched the literature and have found that myxoid chondrosarcoma is typically extra skeletal in presentation. Very few cases of myxoid chondrosarcoma of the hand or wrist have been reported and they were extra skeletal in presentation.

Case report: We report a 65 year old female patient who presented to us with a recurrent swelling of the right 5th metacarpal after being misdiagnosed for chondroblastoma. After thorough investigations, a diagnosis of chondrosarcoma of the 4th and 5th metacarpal was made and wide surgical excision with reconstruction was planned. Histopathological studies confirmed skeletal myxoid chondrosarcoma.

Conclusion: The purpose of this report was to present the rarity of this tumor in the metacarpal and the use of a novel technique to reconstruct the affected ray to provide a functional hand for the patient after wide excision of the tumor.

Key words: skeletal myxoid chondrosarcoma; hand; novel surgical recconstruction

Introduction

Myxoid chondrosarcoma is a distinct subtype of chondrosarcoma with several key differences from conventional chondrosarcoma of bone. Our review of the literature and have found that myxoid chondrosarcoma is typically extra skeletal in presentation. Very few cases of myxoid chondrosarcoma of the hand or wrist have been reported and they were extra skeletal in presentation [1-10]. We report a 65 year old female patient who presented to us with a recurrent swelling of the right 5th metacarpal after being misdiagnosed for chondroblastoma. We report this case due to the rarity of this tumor in the metacarpal and the use of a novel technique to reconstruct the affected ray to provide a functional hand for the patient after wide excision of the tumor.

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Case Report

We report a 65 year old female patient who presented to us with complaints of pain and recurrent swelling on the dorsum of right hand since one year. She first noticed her swelling on the dorsum of her right hand 15 years back, which was pea sized and not associated with any pain or disability. It gradually progressed to the present size and was associated with pain and redness for which the patient went to a local hospital and underwent FNAC which revealed a diagnosis of chondroblastoma. She later underwent surgery for excision of the mass six years back. She had been asymptomatic till one year ago, when she observed a recurrence in the swelling, which was associated with pain and redness and restriction of mobility of the right hand since three months.

Clinical examination: On clinical examination a swelling of 5x6 cm over the dorsum of right hand was observed over the dorsum of 5th metacarpal encroaching up to the 4th metacarpal. It was ovoid in shape with smooth surface and well defined borders (Fig. 1). The fingers seemed to be well vascularized and a previous surgical scar was seen. The swelling was of variable consistency firm to hard and was tender. There was irregular thickening of the 5th metacarpal.

Investigations: X-ray of the right hand AP and oblique: It showed irregular moth eaten appearing permeative destructive lesion of the 5th metacarpal. There was suspicion of 4th metacarpal involvement also due to surrounding soft tissue changes (Fig. 2).

MRI of the hand: A well defined lobulated non- homogenously enhancing soft tissue mass lesion involving the 5th metacarpal with irregular bony destruction (Fig. 2)

Surgery: Ray amputation of 5th metacarpal and excision of 4th metacarpal with reconstruction of 4th

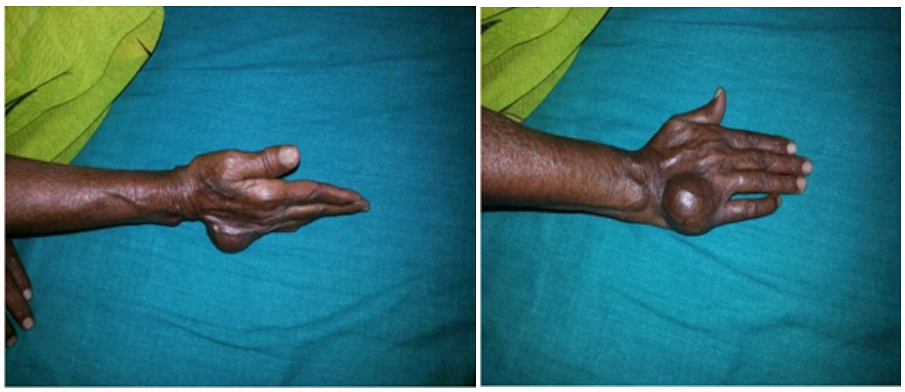

Figure 1: Clinical photograph

metacarpal (Fig. 3, 4, 5).
Histopathology: Macroscopy: Well encapsulated lesion seen with areas of hemorrhage and myxoid deposition. Microscopy: Tumor cells arranged in sheets of myxoid stroma. Cells have ill defined borders and pleomorphic nuclei with rhabdoid cells, tumor cells seen infiltrating subcutaneous tissue. Features are suggestive of myxoid chondrosarcoma (Figure 6).

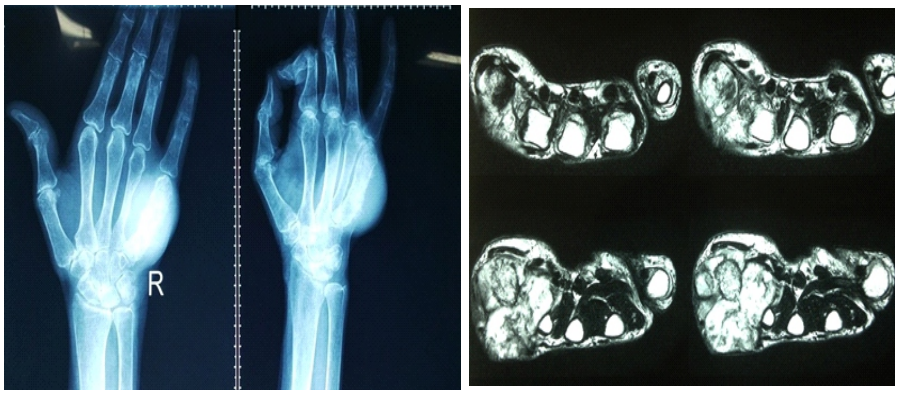

Figure 2: Radiographic photos of the hand showing the tumour

Discussion:

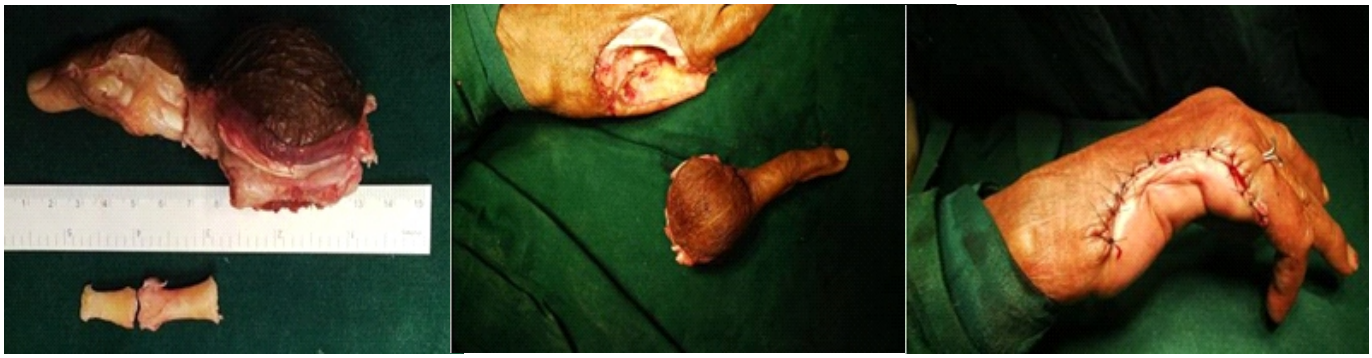

Figure 3: Intraoperative picture of the tumour

In our review of literature only thirty-two cases of skeletal myxoid chondrosarcoma that were studied [1-10], sixteen were in male patients, fourteen were in female patients, and two had no data on sex. The age at the time of diagnosis ranged from nine to seventy years, with a mean of forty-two years. The femur was the most common site of involvement.
Only three cases of myxoid chondrosarcoma in the hand or wrist have been reported, and all were extra skeletal. All three patients were treated with ray resection and were disease-free at the time of the latest evaluation. Wide surgical resection remains the most accepted treatment. Little success has been documented with use of chemotherapy and radiation in controlling myxoid chondrosarcoma tumors [11, 12].

Myxoid chondrosarcoma is a distinct

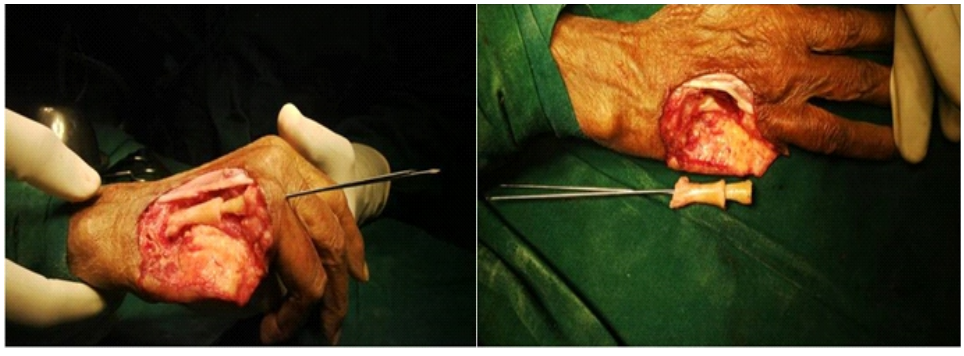

Figure 4: Surgical reconstruction of the 4th metacarpal using the 5th finger phalanges with K-wire

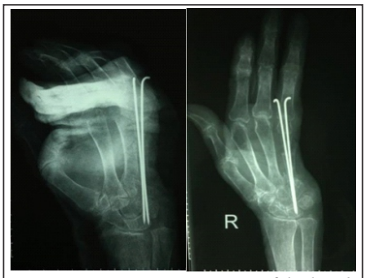

Figure 5: Post operative X-ray of the hand

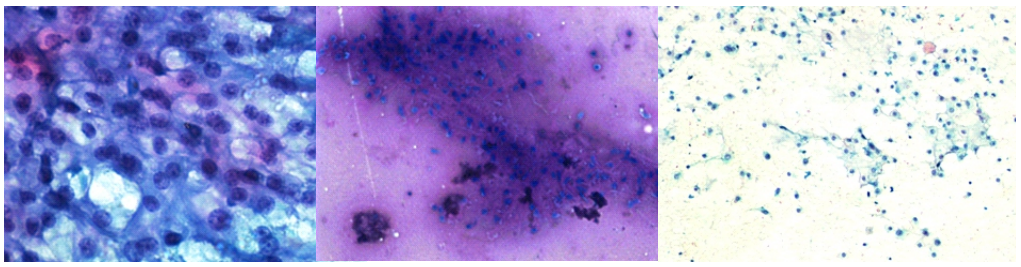

Figure 6: Histopathology photograph a- Photograph Demonstrating the tumor cells. b,c -Photograph demonstrating the tumor cells with myxoid matrix

subtype of chondrosarcoma with several key differences from conventional chondrosarcoma of bone. First, myxoid chondrosarcoma tumors does not contain substantial amounts of hyaline cartilage or mineralized matrix, traits characteristic of conventional chondrosarcoma [8, 13, and 14]. Rather, myxoid chondrosarcoma is composed of cords and strands of small eosinophilic cells within an extensive myxoid stroma, with a lace-like configuration [13, 14, 15, 16, and 21]. However, myxoid chondrosarcoma can also contain foci of extensively pleomorphic and anaplastic cells [17, 18]; and rhabdoid cells [1, 7, 18, 19] as were observed in our case.

Second, myxoid chondrosarcoma has characteristic immunohistochemical properties. The myxoid stroma of myxoid chondrosarcoma contains sulfated acid mucopolysaccharides, demonstrated by strong alcian blue staining resistant to hyaluronidase pretreatment, and by metachromatic staining with toluidine blue [5, 11, 12, 16, 6, and 20]. Extra skeletal myxoid chondrosarcoma tumors have shown strong reactivity for vimentin in almost all
tumors studied [13, 15].

Third, myxoid chondrosarcoma has unique genetic markers that differentiate it from conventional chondrosarcoma. The EWS translocation is the most common translocation seen in cases of extra skeletal myxoid chondrosarcoma, t (9; 22) EWS-CHN [22]. Our patient was from an economically poor background and hence these studies could not be performed. We did not find that establishing genetic confirmation of myxoid chondrosarcoma would affect the treatment and outcome in any way.

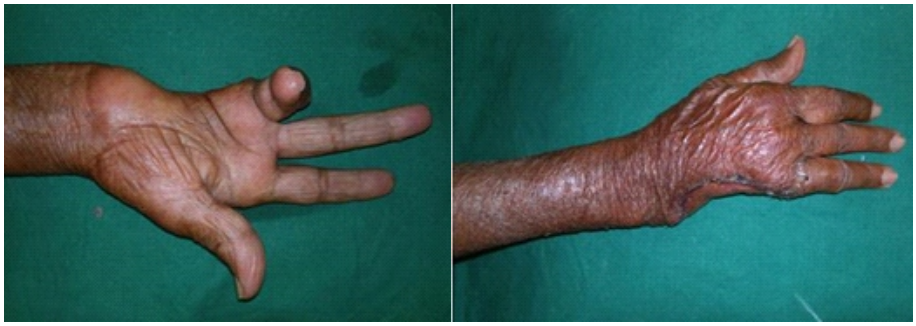

Figure 7: Functional outcome after 3 months

The treatment of low-grade myxoid chondrosarcoma is controversial with good results after extended curettage and the use of intraoperative adjuvant treatments. Resection is the treatment of choice for the rest. For resection, a wide operative margin is suggested, as it will reduce the prevalence of local recurrence.

Chemotherapy has no role in the treatment of conventional chondrosarcoma. Radiotherapy likewise has a limited role in treating surgically inaccessible areas. [13, 15]

In our patient, the tumor was involving the 4th and 5th metacarpal with surrounding muscles, precluding curettage and salvation of the metacarpal; hence wide resection (Ray amputation of 5th metacarpal and excision of 4th metacarpal with reconstruction of 4th metacarpal was done with reconstruction using the phalanges of the 5th digit) was opted for treating this patient.

Since our surgical technique could provide a functional hand, the patient could continue his job (farming) after six months. After a one year follow up patient had good range of movements of his fingers. His hand grip was 75% of the contralateral hand (Figure 7).

Myxoid chondrosarcomas are slow growing tumors and their early diagnosis and management is critical. Long-term follow-up with regular radiographs of the operative site and the chest is imperative so that treatment can be initiated promptly in the event of a recurrence. The treatment of choice remains wide surgical excision of the tumor.

Conclusions

The purpose of this report was to present the rarity of this tumor in the metacarpal and the use of a novel technique to reconstruct the affected ray to provide a functional hand for the patient after wide excision of the tumor.

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