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Case Report

Extra Skeletal Myxoid Chondrosarcoma of Hand – A Case Report with Review of Literature

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Abstract

Extraskeletal myxoid chondrosarcoma (EMC) arising from soft tissues is a rare neoplasm morphologically characterized by ill-defined nodular masses composed of cords and strands of small eosinophilic cells separated by fibrous septae along with abundant myxoid stroma. We report a middle-aged man presenting with swelling on his hand for the last 11 years. He reports occasional pain in the swelling without any increase in its size or change of color. Radiologically, it mimicked a giant cell tumor of the tendon sheath. We received a white gelatinous tumor mass showing myxoid and hemorrhagic areas on cut section. Histo-morphological examinations suggested a diagnosis of EMC. Immunohistochemistry showed a strong expression of S100 and NSE. This slow-growing mass mimicking a benign giant cell tumor of tendon sheath warranted an urgent examination for metastasis and further treatment.

Keywords: Chondrosarcoma, NR4A3 gene, Myxoid, pazopanib

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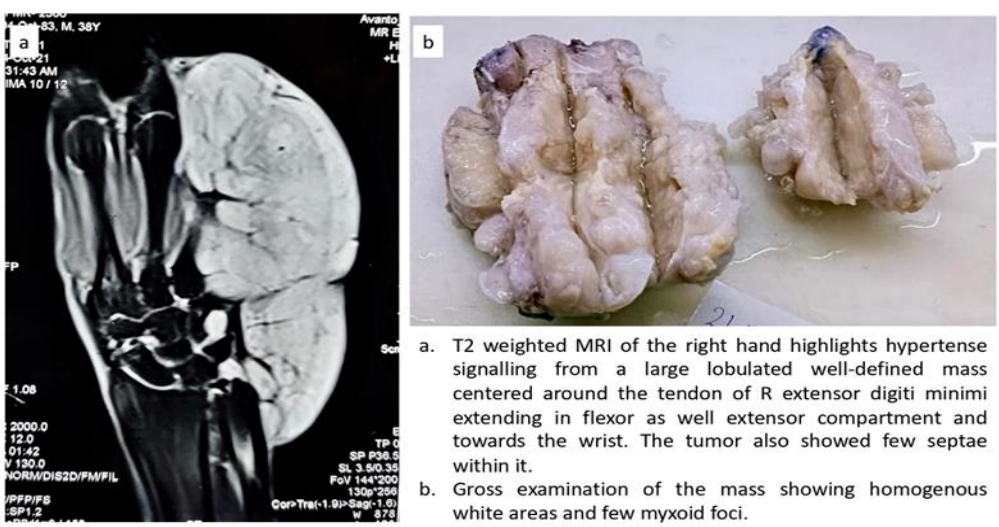
INTRODUCTION

Extraskeletal Myxoid chondrosarcoma arising from soft tissues is a comparatively rare neoplasm morphologically characterized by ill-defined nodular masses made up of cords and strands of tiny eosinophilic cells that are separated by an abundance of myxoid stroma[1] . The cells' cartilaginous origin was suggested by their resemblance to developing chondroblasts, the mucoid ground substance staining reactions indicating the presence of large amounts of sulfated mucopolysaccharides, and the electron microscopic findings but this remains uncertain.[2] EMC is defined by the rearrangement of NR4A3 gene, which typically (62–75%) fused with EWSR1 gene as a result of a t(9;22) (q22;q12)[3]

CASE REPORT

A 38-year-old male presented to the opd for consulting a swelling on his right hand that had been present for the last 11 years. The patient complained of occasional pain in the swelling that did not affect the quality of his life. He did not report any increase in its size or change of color. Radiological examination of the mass suggested the differentials of giant cell tumor of the tendon sheath or a soft tissue mesenchymal tumor. MRI of the lesion showed a large lobulated soft tissue mass centered around the tendon of extensor digiti minimi, suggesting a tendon sheath giant cell tumor (Figure 1a).

Grossly, the solid tumor measured 13 cm in its largest dimension with white gelatinous outer surface, while the cut section showed creamish white and myxoid areas along with hemorrhagic spots (Figure 1b). Low power histopathological examination on H&E stained sections showed a multinodular architecture divided by fibrous septa with hypocellular and hypercellular areas. These hypercellular areas showed round to oval cells having mild pleomorphism, vesicular chromatin, and abundant eosinophilic cytoplasm. Hypocellular areas showed an abundant myxoid matrix with cells arranged in cords, trabeculae, and reticular patterns. These cells were oval to spindle in morphology with mild pleomorphism, elongated nuclei, and cytoplasmic vacuolation (Figure 2 a,b). Thus, histomorphology was consistent with malignant spindle cell neoplasm, and the differentials of extra-skeletal myxoid chondrosarcoma and myxoid liposarcoma were made. Immuno-histochemical evaluation both with S100 and NSE (Neuron-specific Enolase) showed strong cytoplasmic positivity (Figure 2 c,d). The patient was kept in close follow-up for metastasis, but none were detected.



a. T2 weighted MRI of the right hand highlights hypertense signalling from a large lobulated well-defined mass centered around the tendon of R extensor digiti minimi extending in flexor as well extensor compartment and towards the wrist. The tumor also showed few septae within it.
b. Gross examination of the mass showing homogenous white areas and few myxoid foci.

Figure 1: MRI of the right hand and gross of the tumor mass

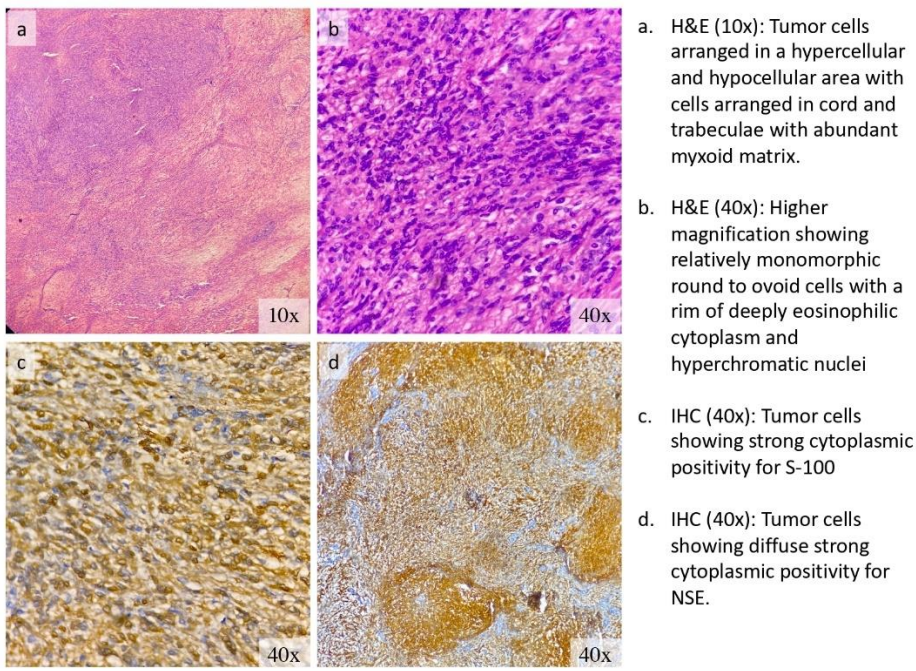


Figure 2: Microscopic and Immunohistochemical examination of the tumor mass

DISCUSSION

Extra skeletal myxoid chondrosarcoma is a rare pathological entity comprising <1% of soft tissue sarcomas. [1,2] The origin of these tumors has been debated in literature since the report of its first case in 1972 by Enzinger and Shiraki and still remains uncertain. [2] Most cases have been reported in adults, with the median age being 50 years. [4, 5] Deep soft tissues of the thigh are the most common site, whereas fewer cases have also been reported in trunk, head, and rarely in fingers, cranium, and retroperitoneum. [6-8] Microscopically, these have a multinodular architecture with fibrous septae dividing the tumor into hypercellular and hypocellular areas with myxoid matrix. The tumor cells may connect to form cord, tuberculae, and cribriform arrays. [1,2] Meis-Kindblom studied a collection of 117 cases of these tumors and found this classical appearance in all cases in at least one foci, if not in all. [4] Some cases also identified a few cellular foci reminiscent of chondroblastoma, Ewing's sarcoma, fibrosarcoma, monophasic and poorly differentiated synovial sarcoma, and rhabdoid tumor. [4]

The EMCs have not been defined by immunohistochemistry. S-100 positivity has been seen in about 20% of cases and C-kit in about 30%. Expression of NSE and synaptophysin has also been reported in some. Molecular studies of these tumors have shown NR3A4 gene rearrangement in about 90% cases. [5] These fusions have not been identified in any other sarcomas and are currently considered the tumor's hallmark. The positivity for NR4A3 rearrangement is, therefore, helpful in differentiating them from morphologically similar tumors like myoepithelioma and myoepithelial carcinoma. [3] EMC, originally believed to be a low-grade sarcoma, has been shown to have a unique clinical course. Close long-term analysis of these tumors has shown a high rate of local recurrence and metastasis. In one such study, local recurrences were seen in 48% out of which 58% had multiple local recurrences, while metastases occurred in 46%.[4] After 5, 10, and 15 years, the projected survival rates were 90%, 70%, and 60%, respectively.[4] Metastasis, when present, is

usually pulmonary. Extra-pulmonary sites and disseminated diseases have also been noted. [9] Over time EMC has shifted its place to high-grade sarcomas, with the mainstay treatment being wide-local excision with negative microscopic margins following the algorithm of all soft tissue sarcomas. The standard treatment for metastatic/advanced disease is chemotherapy based on anthracycline or trabectedin or target therapy with pazopanib or sunitib in case of treatment failure. [10] EMC is a slow-growing tumor, and surgical treatment of isolated metastases may improve outcomes as these metastatic growths remain stable for many years.

CONCLUSION:

EMC is a slow-growing tumor, and surgical treatment of isolated metastases may improve outcomes as these metastatic growths remain stable for many years. This case report depicts the distinctive morphological features of the rare tumor.

Authors Contribution

Apoorva S: Manuscript editing, Literature search, data collection

Varun W: Manuscript grammar and drafting

Vaishnavi R: Data Analysis, manuscript drafting

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Conflict of interest

The authors have nothing to disclose or any conflicts of interest.

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