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Case Report

Extraskeletal Myxoid Chondrosarcoma of Left Thigh: A Case Report

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ABSTRACT

Extraskeletal myxoid chondrosarcoma is a rare soft tissue sarcoma which is readily distinguishable from skeletal myxoid chondrosarcoma. It is characterized by distinctive morphological and cytogenetic features. As its name implies, EMC was believed to represent a variant of soft tissue chondrosarcoma owing to its histological resemblance to chondroblastic tissue in early stages of development or chondroid tumors such as skeletal chondrosarcoma. A case of an extraskeletal myxoid chondrosarcoma in a 45-year- old male localized in the left thigh is presented. The histological, histochemical, and ultrastructural features of this rare tumor are described and compared with those of the pertinent literature. The value of histochemical methods as an aid in differentiating this tumor from other malignant myxoid tumors of soft tissue is emphasized.

***Key words**:* Extraskeletal chondrosarcoma, myxoid sarcoma, bone

INTRODUCTION

Extraskeletal myxoid chondrosarcoma (EMC) is a relatively rare but well-recognized neoplasm which is locally aggressive and have metastasizing potential. It has distinctive clinical, light microscopic, immunophenotypic, cyto-genetic and ultrastructural features. [1] Conventionally it is regarded as a neoplasm showing chondroid differentiation but arising out the skeleton. [2] It was first described by Stout and Verner in 1953 in a small series of cases which arose in the extremities of adults. [3]

CASE REPORT

We report a case of 45-year old male patient presenting with swelling in the left thigh since 5-6 months. The swelling was deep seated located in anterior and lower part of left thigh, measuring 7cmx5cm.non tender. No signs of inflammation or fixity of swelling to overlying skin were noted. The swelling was operated with wide local excision and specimen was sent to pathology department for histopathological examination. Radiologically the mass was seen separate from femur, situated in anterior and lower compartment of left thigh.

Grossly the tumor was lobulated mass, well circumscribed with the distinct fibrous

capsule. The size of the tumor was 7x5x4.5cm. Cut surface was multinodular with glistening surface. Microscopically the tumor had distinct multinodular configuration delineated by fibrous connective tissue. (Fig.1) The tumor cells were arranged in delicate intersecting strands in most of the areas. The myxoid matrix was abundant. Individual tumor cells were round or slightly elongated with acidophilic cytoplasm, embedded in an abundant myxoid matrix. (Fig.2) the cells showed minimal features of chondroblasts and differentiated cartilage cells were not seen.

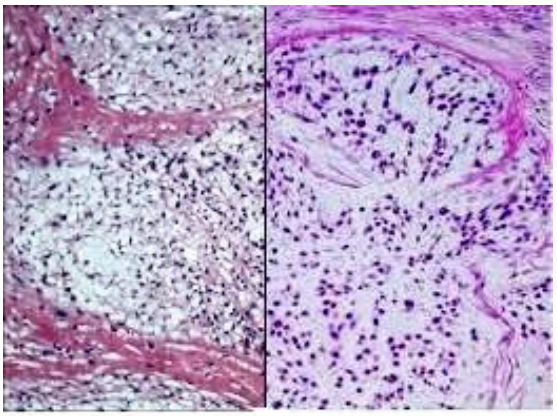

Fig. 1. Lobulated tumor mass, well circumscribed by a distinct fibrous capsule. (HE, 40X)

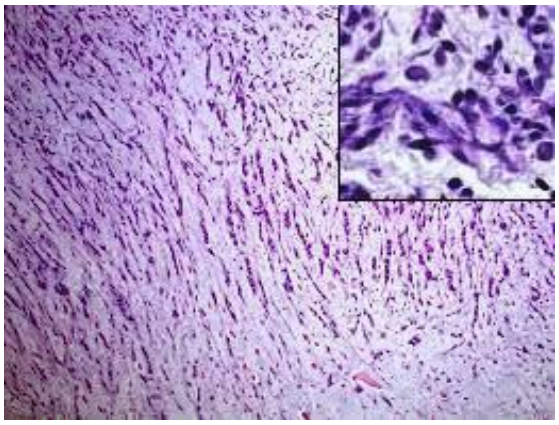

Fig 2. Thin anastomosing strands of tumor cells surrounded by an abundant myxoid matrix, (HE, 100x),) (Inset, 400x)

Immunohistochemical analysis was performed in this case. The tumor expressed vimentin, S-100 protein and focal positivity for neuron-specific enolase. It was negative for EMA and desmin. (Fig.3) Hence, the diagnosis of extraskeletal myxoid chondrosarcoma of left thigh was offered.

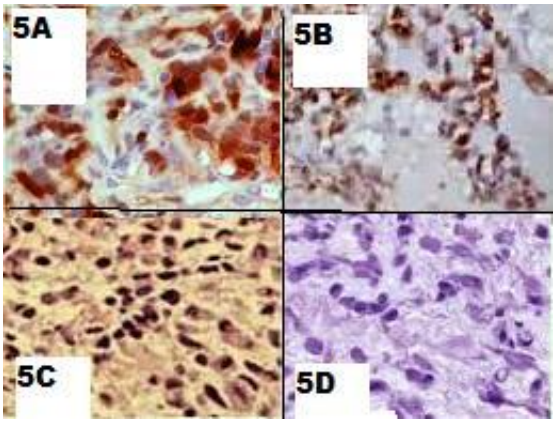

Fig.3. Tumor showed positivity for S-100 protein(5A), vimentin(5B), and neuron-specific enolase(5C) & negativity for desmin(5D).

DISCUSSION

EMC is an uncommon entity occurring primarily in the deep soft tissue of the extremities, distinct from primary skeletal chondrosarcoma with myxoid alteration. This tumor has potential to metastasize and recur. [4]

Epidemiologically it is a rare tumor contributing 2.3% of soft tissue sarcomas. Mean ages reported in various series range from 46 to 57 years, this tumor being exceptional in children and adolescent. Males are affected about twice as often as females. [5] Although multiple predisposing factors have been identified such as genetic diseases, radiation exposure, certain chemical agents, lymphedema, and trauma, no specific etiology has been found. [6]

EMC was first described in 1953 by Stout and Verner, who reported a small series of morphologically similar appearing cases, all of which arose in the extremities of adults. [3] However, in 1972 Enzinger and Shiraki, in a series of 34 cases, first established extraskeletal myxoid

chondrosarcoma or Chordoid sarcoma as a distinct clinicopathological entity. [2]

EMC has distinctive clinical, light microscopic immunophynotypic, cyto-genetic and ultrastructural features. [3] It is located in deep soft tissues of lower extremities in about 75% of cases, especially in the thigh, the popliteal fossa, and the buttock; occasionally a bone involvement may exist, as a major component. [5]

Macroscopically the tumor presents as lobulated or multinodular mass, generally well circumscribed by a distinct fibrous capsule. The size of the tumor at the time of diagnosis may vary from 1 to about 20 cm (mean size: about 7cm). On histology, typically the tumor nodules are composed of round or slightly elongated cells, with minimal features of chondroblasts, separated by mucoid substance; differentiated cartilage cells are rare. Histological diagnosis may be very difficult in highly cellular forms devoid of myxoid matrix. [4,5]

Immunohistochemically the tumor cells show positivity for vimentin, S-100 protein, occasionally for EMA, and negativity for cytokeratin. A subset of tumors display neural or neuroendocrine differentiation as shown by positive immunohistochemical reactivities to neural or neuroendocrine markers such as neuron specific enolase, synaptophysin, chromogranin A, and PGP9.5; tumors are mostly negative for markers (collagen type II, X, proteoglycan aggrecan) for the chondrocytic cell lineage. [5,7]

Ultrastructurally, at least one third of the tumors demonstrate microtubular aggregates within dilated rough endoplasmic reticulum; neurosecretory granules (80-170 nm in diameter) are occasionally identified. [3,5]

The histogenesis of EMC is still disputed. In spite of the absence of well differentiated cartilage by light microscopy, the tumor has always been viewed as being of chondroblastic origin based on interpretation of histochemical in ultrastructural data. One report suggested an origin from synovial intimal cells as extraskeletal myxoid chondrosarcoma frequently arises in close proximity with tendons and ligaments. [3,4]

It was initially thought to be a low grade sarcoma of cartilage deviation and to contain a reciprocal t(9;22), resulting in fusion of the *EWS* and *CHN* genes.(2) Cytogenetic studies have demonstrated the presence of recurrent translocation t(9;22)(q22:q12); it results in the fusion of the EWSR1 gene on chromosome 22 with NR4A3 (TEC, CHN, or NOR1) gene on chromosome 9. [4,5]

Treatment for EMC is surgical excision with adjuvant chemotherapy in case of lymph nodes or metastasis. [5,7] This is an aggressive neoplasm which recurs locally and has high rates for metastasis, especially to lungs, over the long term. Because of its protracted but resilient nature, a tenacious and long –term follow is necessary for any patient. [4,7]

CONCLUSION

Extraskeletal myxoid chondro-sarcoma is a rare soft tissue sarcoma characterized by distinctive morphological, immunohistochemical and cytogenetical features. EMC has high potential of local recurrence and metastasis, and high disease-associated death rate hence, frequent and long term follow-up is required.

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