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Case Report

Extraskeletal Myxoid Chondrosarcoma - A Rare Case

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ABSTRACT

Background: Extraskeletal myxoid chondrosarcoma is a rare tumor comprising 2.5% of soft tissue sarcomas. The tumor presents as a painless mass in the proximal extremities and trunk, is histologically, immunohistochemically & genetically distinct from osseous chondrosarcoma.
Case History: We present a case of Extraskeletal Myxoid chondrosarcoma in a 63 year old female patient. She presented with a painless soft tissue mass along the lateral aspect of right knee since six years which was slowly increasing till present size of 10x10cm. Wide excision was done. Histopathological examination aided with immunohistochemistry confirmed the diagnosis of Extraskeletal Myxoid chondrosarcoma. The patient did not reveal metastasis at the time of diagnosis. Three months follow up is uneventful.
Conclusions: 1) Extraskeletal myxoid chondrosarcoma is a rare malignant soft tissue tumor. Definite diagnosis is made by histopathological examination with the support of immunohistochemistry.
2) Extraskeletal myxoid chondrosarcoma is a phenotypically and genotypically distinct entity than osseous chondrosarcoma and needs long term follow up because of its protracted clinical course.

Keywords: Soft tissue tumor, extraskeletal myxoid chondrosarcoma, histopathology.

INTRODUCTION

Extraskeletal myxoid chondrosarcoma (EMC) is a rare soft tissue tumor composed of spindle and epitheloid cells set in a myxoid matrix that biologically and histochemically resemble cartilage. These tumors are histologically, immunohistochemically and genetically distinct from osseous chondrosarcoma. [1] We present a case of EMC in a 63 year female patient to highlight its rarity, and distinct gross, microscopic and immunohistochemical characteristics.

CASE HISORY

A 63 year female patient was referred to our institute with the complaint of gradually increasing swelling of 10x10cm over lateral aspect of right knee since six years with no evidence of pain or discharge. There was no history of trauma.

Investigations:

X ray AP and lateral view of right knee showed Soft tissue tumor over distal aspect of right knee with no connection to bone (as seen in fig 1-A).

MRI revealed a well defined lobulated lesion in anterolateral aspect in the

subcutaneous plane of distal aspect of right thigh with no connection to bone with areas of haemorrhage and necrosis suggestive of sarcoma. (as seen in Fig 1- B)

X-ray chest and other investigations were within normal limits, suggesting no evidence of metastasis. Excision of the lesion with overlying skin was done and the specimen was sent for histopathological examination.

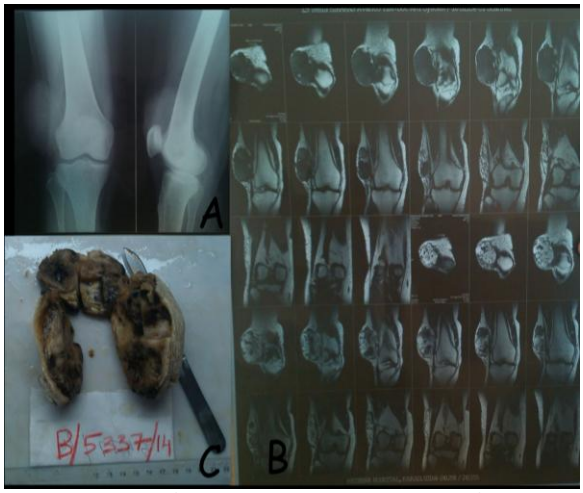

Fig 1- A-X ray- Soft tissue tumor over distal aspect of right knee with no connection to bone.
B- MRI- Lobulated lesion in subcutaneous plane of distal aspect of right thigh suggestive of sarcoma.
C- Gross- Gray brown solid cystic tumor with multiple nodules.

Pathological features:

Gross examination revealed a single lobulated grey brown soft tissue tumor measuring 9x7x2cm covered with skin. Cut surface showed multiple grey white to brown coloured tumor nodules with intervening areas of cystic change, hemorrhages and lot of gelatinous tissue. (as shown in Fig 1-C)

Microscopy revealed cords, nests and clusters of spindle and stellate cells with uniform dark staining nuclei, indistinct nucleoli and scanty to moderate amount of amphophilic cytoplasm(fig 2-A,B &C). These cells were dispersed within a myxoid stroma which was positive for alcian blue at pH 4.0 and 5.0 suggesting the presence of chondroitin sulphate.

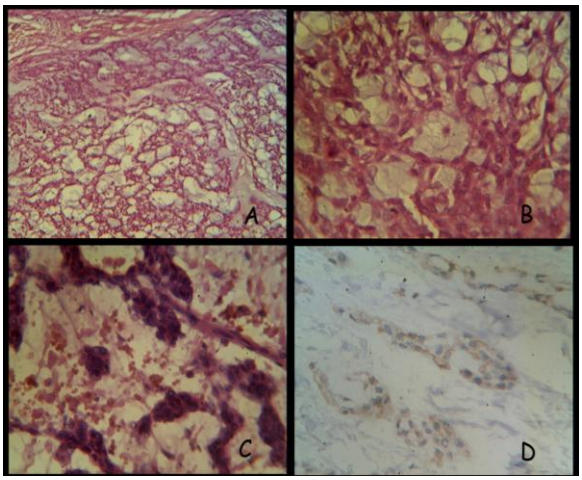

Fig 2- (A,B,C)- Cords and nests of spindle to stellate cells set in myxoid stroma. (100x & 400x H & E). D- Immunopositivity for CD117.

The tumor was immunopositive for CD117 (fig 2-D) and immunonegative for EMA, synaptophysin, S100 protein, CD68, CD10, SM4, desmin and MDM2.

Considering all the points, the diagnosis offered was "Extraskeletal myxoid chondrosarcoma" arising from the soft tissue along the lateral aspect of right thigh.
Three months follow up of patient is uneventful.

DISCUSSION

Extraskeletal myxoid chondrosarcoma is an uncommon tumor accounting for less than 2% of all soft tissue sarcomas. [2] It was first described by Stout and Eroer. [3] The tumor affects adult males with median age in the fifth decade [4] and occurs in the deep soft tissues, especially in the lower extremities. The tumor presents as painless, slow growing soft tissue mass and rarely with lung metastasis with an occult primary. [1] Though our case was a female patient, the other clinical findings were exactly same as mentioned in the literature.

These tumors are distinct from osseous chondrosarcoma on the basis of clinical presentation, histology, immunohistochemistry, cytogenetics as well as prognosis. Unliike most chondrosarcomas

of bone, mature chondromatous tissue is a rare feature in these tumors. [3]

Radiological features: Extraskeletal myxoid chondrosarcomas reveal non specific soft tissue masses and does not give accurate diagnosis with radiological studies, [5] the similar experience was shared with our case.

Pathological features: Extraskeletal myxoid chondrosarcomas appear as lobulated soft tissue mass >5cm, with grey, soft and gelatinous areas. Microscopy reveals cords, nests and clusters of spindle shaped cells dispersed in a gelatinous matrix. Cellular and matrix poor tumor reveal nuclear atypia with frequent mitosis. [1]

Immunohistochemistry: Extraskeletal myxoid chondrosarcomas are usually immunoreactive for S100, vimentin, neuron specific enolase and synaptophysin and CD117. [6,7] Our case revealed positive immunoreactivity to CD117.

Cytogenetics: Majority of extraskeletal myxoid chondrosarcomas show t(9:22) (922,912) translocations involving EWS and CHN genes. [8,9]

Differential diagnosis: Differential diagnosis of extraskeletal myxoid chondrosarcomas include soft tissue chondroma, soft tissue osteosarcoma and osseous chondrosarcoma with soft tissue extension and ossifying fibromyxoid tumor. [1]

Prognosis and diagnosis:

Treatment of extraskeletal myxoid chondrosarcoma is wide surgical excision. Chemotherapy and radiotherapy can provide palliative benefits to patients with widespread metastases.[10] The tumor has high overall recurrence rate with 5 year survival of 65-80%.[1,10] Histological grading does not correlate with prognosis. Older patient, larger tumor size and tumor location in the limb girdle or proximal extremity are adverse prognostic factors. Metastasis also adversely affects the survival. These patients need long term follow up due to protracted clinical course and high rate of recurrence. [11] Our case also revealed bad prognosis due to advanced age, larger tumor size and tumor site although metastasis was not evident.

CONCLUSION

Extraskeletal myxoid chondrosarcomas are rare soft tissue tumors which are diagnosed with histology and with support of immunohistochemistry.

These tumors have distinct biologic behaviour and pathological features than osseous chondrosarcoma.

Long term follow up is essential due to high rate of recurrence and metastasis.

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