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Clear Cell Sarcoma of Tendon And Aponeurosis – A Rare Case

KEYWORDS: Clear cell sarcoma, Immunohistochemistry, tendon and aponeurosis, S-100.

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ABSTRACT *Clear cell sarcoma of soft tissue is a tumor of melanocytic origin involving tendons and aponeurosis, which lacks skin involvement. The incidence is higher in the third and fourth decades of life and the most common primary sites are distal extremities. It is clinically, genetically and biologically distinct from cutaneous melanoma despite certain histologically similarities. It usually presents as a slowly enlarging mass. The clinical course is difficult to predict with distant metastasis to regional lymph nodes and lungs. We present a case of clear cell sarcoma of tendon and aponeurosis presenting as a soft tissue mass in the upper extremity.*

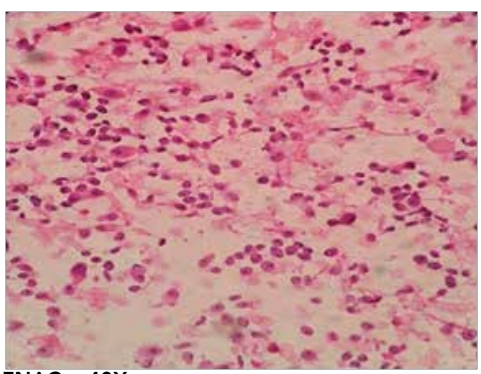
INTRODUCTION:
Clear Cell Sarcoma of Tendon and Aponeurosis also referred to as melanoma of soft parts is a rare malignant tumor of soft tissue. Accounts for less than 1% of malignant tumors in adults[1] arising from soft tissues involving tendons and aponeurosis of distal extremities (lower & upper), rarely bone, GIT, head and neck. It is thought to be derived from neural crest cells.[2]

CASE REPORT:
A 40 yrs lady presented with a painless swelling over flexor aspect of right wrist since 3 months which was firm, non-tender measuring 3×2 cm. There were no other swellings over the body. A provisional diagnosis of soft tissue sarcoma was made.

FNAC revealed oval to spindle shaped cells arranged in loosely cohesive clusters and singly scattered. Some of the cells are large and pleomorphic with altered N: C ratio. The cells have moderate cytoplasm, vesicular nuclei with prominent nucleoli against hemorrhagic background. Myxoid areas and few binucleate cells were also noted. Features were suggestive of spindle cell lesion.

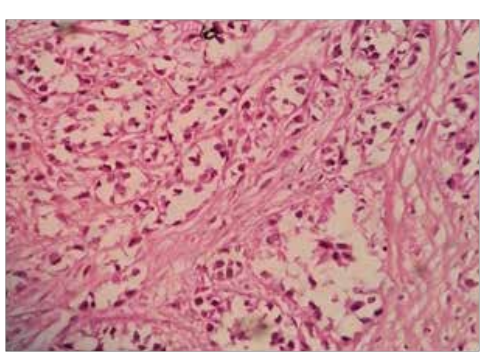
The patient underwent surgical excision and specimen sent for histopathology examination. Gross examination reveals a firm, well circumscribed, grey-white mass with a gritty sensation when cutting.

H& E slides showed tumor cells arranged in compact nests, alveolar & fascicular pattern separated by fibrous septa. Cells were round to spindle with clear/eosinophilic cytoplasm and vesicular nuclei with prominent nucleoli. Multinucleated tumor giant cells were seen. No evidence of hemorrhage or pigment was made out. The sections tested positive for S100 and negative for Desmin. These pathological findings were consistent with the diagnosis and confirmed by immunohistochemistry. Cytogenetic study could not be done in the present case.



FNAC – 40X

GROSS IMAGE



H& E biopsy – 40X

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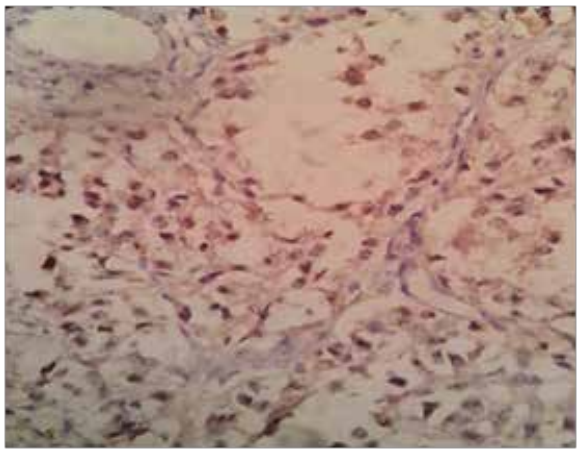

S100 POSITIVE

DISCUSSION:

Clear Cell Sarcoma of Tendon and Aponeurosis is a rare tumor and was first described by Enzinger in 1968, it affects distal extremities of young adults (20-40 yrs; slight predilection for women) and rarely in children. Usually found attached to tendons, aponeurosis and behaves like a high grade sarcoma[3].

Clinical symptoms are soft tissue swelling accompanied by pain or tenderness or may be asymptomatic and a history of antecedent trauma is not unusual. Gross pathologic examination reveals a localized, tan-grey, firm, circumscribed mass measuring between 0.4cm to 14.5cm. The tumor shows infiltration into tendons and aponeurosis with a histological pattern of haphazard arrangement into small compact nests and fascicles of uniform neoplastic cells dissecting along dense fibro connective tissue. These cellular clusters are divided into lobules by fibrocollagenous septa.

The neoplastic cells are polygonal to fusiform with clear or pale eosinophilic cytoplasm and centrally located round to oval vesicular nucleus with prominent nucleoli. The neoplastic cells show no or minimal pleomorphism. Scattered multinucleate giant cells are present and areas of necrosis may be noted. Variants have been described including cases of myxoid change, alveolar pattern of growth because of loss of cellular cohesion.

On cytological specimens features include varied cellularity that shows cohesive groups and single cells. The individual cells are epithelioid, round to polygonal with moderate to abundant cytoplasm. Multinucleate and binucleate tumor cells may also be observed. Melanin is present in half of the cases. IHC shows tumor cells react with S100, NSE and HMB-45. Desmin and Cytokeratin are usually negative. Most cases show reciprocal translocation t (12; 22) (q13; q12) involving EWS-ATF1 fusion gene[4]. Metastasis occurs to regional lymph nodes and lungs with frequent recurrences[5]. Poor prognosis is indicated by large tumor size and necrosis[6]. The mainstay of treatment is excision and MRI plays an important role.

CONCLUSION:

Clear cell sarcoma of tendon and aponeurosis is a slow growing tumor. About 22% are seen in upper extremity, the location in the present case[7]. Cytologically tumor simulates other soft tissue sarcomas (MPNST, synovial sarcoma, fibrosarcoma) and melanin producing tumors (melanoma, pecoma). Malignant peripheral nerve sheath tumor is usually associated with a large peripheral nerve or with manifestations of neurofibromatosis.

Malignant melanoma which is the most important mimic of clear cell sarcoma of tendon and aponeurosis in the soft tissues occurs in patients who generally have a preceding history of cutaneous melanoma and absence of reciprocal translocation not found in melanoma. Synovial sarcoma can be differentiated by characteristic chromosomal translocation. The distinctive cytological features of clear cytoplasm, prominent nucleoli and Immunophenotyping and cytogenetic sets clear cell sarcoma apart from other soft tissue sarcomas. There is evidence for melanocyte differentiation by Immunohistochemistry, ultra structural and genomic profiling studies [8].

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