

Clear Cell Sarcoma - A Rare Case Report

Dr. Sujata S. Giriyan¹, Dr. Chethana H.D²

¹Professor and HOD, Department of Pathology, Karnataka Institute of Medical Sciences, Hubballi, Karnataka, India

²Post Graduate, Department of Pathology, Karnataka Institute of Medical Sciences, Hubballi, Karnataka, India

Abstract: Clear cell sarcoma of tendon and aponeuroses is extremely rare tumour of melanocytic origin, arises in deeper soft tissues, bound to tendons or aponeuroses. Despite histological similarity with cutaneous melanoma, it is distinct from it clinically, genetically. It is mainly seen between the ages 20 to 40 yrs and rarely occurs in children. Here we are presenting a 12yr old girl with clear cell sarcoma arising in left thigh.

Keywords: Clear cell sarcoma, Malignant melanoma of soft parts, Soft tissue sarcomas, Paediatric tumours.

1. Introduction

Clear cell sarcoma of tendon and aponeuroses is a rare malignant tumour, accounting for 1% of all soft tissue tumours [2,6,7]. It occurs preferentially in adolescents and young adults with predilection for lower extremities in close association with tendons, aponeuroses [1,2,6].

Because of histologic and immunohistochemical similarity with conventional malignant melanoma, Chung and Enzinger proposed a name "malignant melanoma of soft parts" [3,6]. However it lacks junctional changes and rarely involves epidermis [2].

Cytogenetic analysis showing characteristic translocation t(12;22) (q13;q13) has been considered pathognomonic for CCS involving ATF1 gene on chromosome 12 and EWS gene on chromosome 22 [1,2,6,8]. This translocation is absent in malignant melanoma, hence these tumours are considered as different entities [6].

Most cases present as slow growing painless mass in in lower limbs followed by dissemination to lymphnodes and lungs.

2. Case Report

A 12 yr old girl presented with pain and swelling over lower end of left thigh since 3 months, which was tender and fixed to underlying structures. No history of trauma.

On examination a firm, tender mass measuring 8×4 cms noted in distal end of left thigh. Provisional diagnosis of "SOFT TISSUE TUMOUR OF LEFT THIGH" was made.

X-RAY: Normal.

M.R.I. :- Showed lobulated mass measuring approximately 7.7×3.7×4.1 cms in left quadriceps tendon causing its expansion and extending to involve vastus medialis muscle. Diagnosis of **Giant cell tumour of quadriceps tendon sheath** was given.



Figure 1: MRI of left thigh showing lobulated mass in quadriceps tendon

FNAC : Revealed cellular smear showing oval to spindle cells in aggregates with tapered cytoplasm. Nuclei were vesicular with prominent nucleoli.

Impression of *spindle cell tumour* was made.

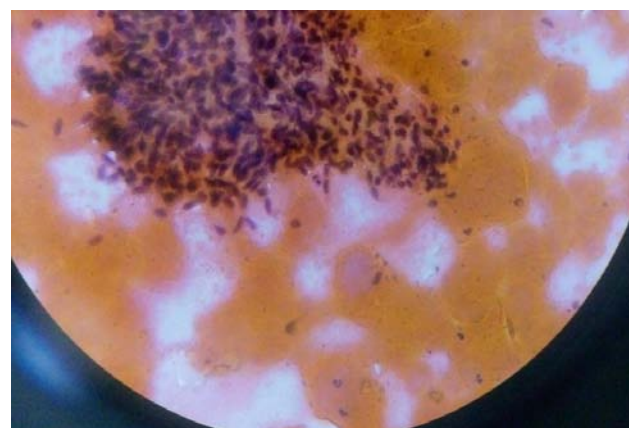


Figure 2: FNAC smear showing spindle cells in nests (40x10)

Patient underwent surgical excision and specimen submitted for histopathological examination. Grossly it revealed multiple, irregular, tissue pieces, Largest measuring 7×5.5×3.5 cms. Cut section showed grey white to grey brown areas.

Microscopy: H & E revealed tumour cells arranged in nests, groups and fascicles. These tumour cells were round to

spindle shaped with pale eosinophilic cytoplasm and vesicular nuclei with prominent nucleoli. Few cells showed clear cytoplasm. The cell nests were surrounded by thin delicate fibrous septae.

Diagnosis of malignant spindle cell tumour possibly Clear cell Sarcoma of soft part was given.

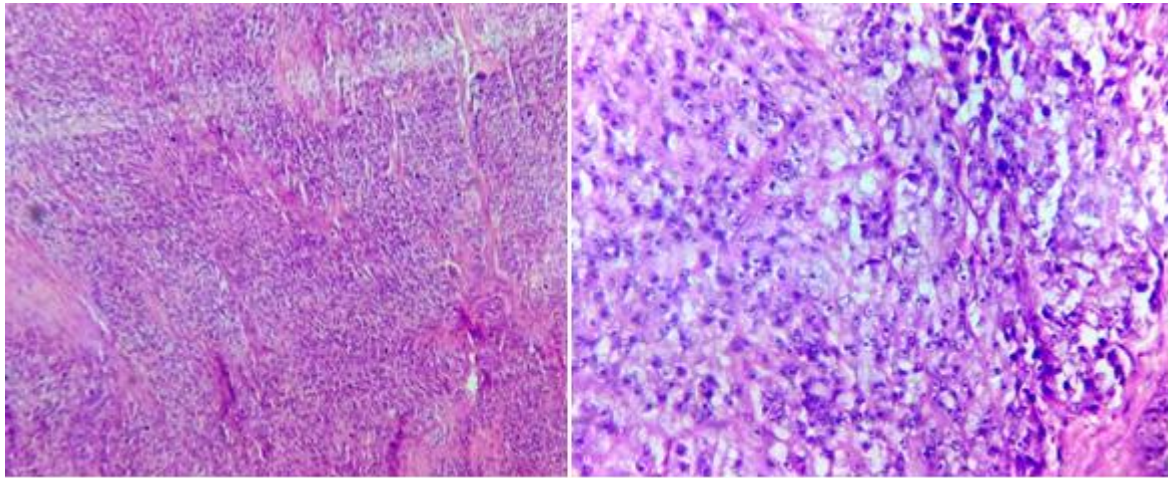


Figure 3a: Cells arranged in fascicles and sheets (H&E,4x10). 3b, Round to spindle cells in groups having vesicular nuclei and prominent nucleoli (H&E, 40x10).

On IHC, cells were positive for vimentin, S-100 and HMB-45, negative for Cytokeratin, Desmin, SMA. Diagnosis of CLEAR CELL SARCOMA – MALIGNANT MELANOMA OF SOFT PARTS was given

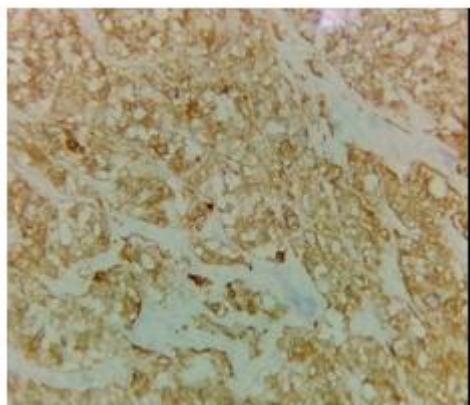


Figure 4 (a): Cells showing positivity for S100

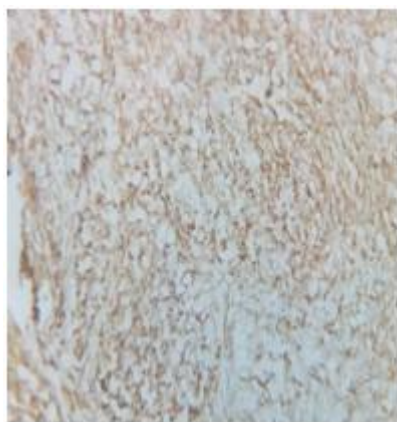


Figure 4 (b): Cells positive for vimentin

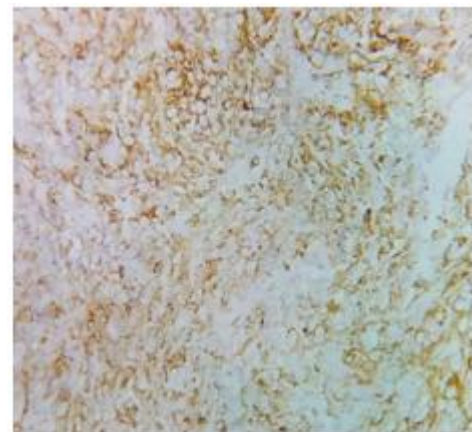


Figure 4 (c): Cells positive for HMB 45

3. Discussion

The case presented here fulfils the histologic criteria of Clear cell sarcoma of tendons and aponeuroses which was first described by Enzinger in 1968^[1,5].

It predominantly affects young adults between 20-40 yrs of age with female preponderance, rarely occurs in children^[1,2,3,4,6]. It commonly arises in lower extremities, foot and ankle followed by thigh, toes. Head and neck, trunk are rarely involved^[1,3,6].

Clinically presents as a painless slow growing mass that has been present for several months or even years^[6]. Grossly it appears as a well circumscribed lesion measuring 0.4-14.5 cms^[2,4,7].

Microscopically, Clear cell sarcoma displays compact nests and fascicles of uniform or mildly pleomorphic tumour cells which are round to spindle shaped with clear / eosinophilic abundant cytoplasm and vesicular nuclei with prominent nucleoli separated by delicate fibrous septa^[1,2,7]. Tumour

cells may have melanin pigment in 50% cases and areas of necrosis may also be present.

On immunohistochemistry they express S100, HMB 45, melan A, MITF, NSE, Cd57. Negative for desmin and cytokeratin ^[1,2,4].

Ultrastructurally melanosomes are detected within tumour cells ^[1,7]. Clear cell sarcoma clinically and radiologically appears as benign lesion but behaves like high grade sarcoma ^[1,4]. It has tendency to recur and metastasis occurs to regional lymphnodes ^[1,6,7]. Overall prognosis is poor ^[3,6,7]. Adverse prognostic factors include tumour size > 5cm and presence of necrosis ^[1,2,7]. Complete tumour resection is the main stay of treatment ^[1,6].

4. Conclusion

Clear cell sarcoma is a rare malignant tumour particularly uncommon in children. Clear cell sarcoma shares many features with melanoma including histological, immunophenotypic, ultrastructural pattern. However, Clear cell sarcoma doesn't display any pagetoid spread of atypical melanocytes. Ultimately t(12; 22)(q13;q12) translocation involving ATF 1 and EWS genes observed in Clear cell sarcoma. BRAF mutations in Malignant melanoma.

Cytologically, Clear cell sarcoma simulates other soft tissue sarcomas like MPNST, synovial sarcoma, fibrosarcoma and melanin producing tumours like PEComa, cellular nevus. Clear cell sarcoma differs from them by its distinct cytologic features with prominent nucleoli, clear cytoplasm and immunophenotypic profile.

Differentiation from clear or spindle cell neoplasms and unusual malignant melanoma subtypes is essential for patient management. Early diagnosis and treatment of such entity may improve the prognosis.

References

- [1] Sharon w.weiss, John R Goldblum. Enzinger and weiss's soft tissue tumors, 5th edition. St.Louis, Missouri.
- [2] Haren V. Oza, Jignasa N.Bhalodia et all. Clear cell sarcoma of gluteal region; malignant melanoma of soft parts. National journal of medical research. 2013;3(2):193-195.
- [3] O. Hocar,A. Le Cesne et all. Clear cell sarcoma(malignant melanoma) of soft parts:A clinicopathologic study of 52 cases. Dermatology research and practice.2012;vol 2012, article id 984096.
- [4] Dr Syed M Rahman, Dr. KPA Chandrashekar. Clear cell sarcoma of tendon and aponeurosis- A rare case. Indian journal of applied research.2015;5(1):459-460.
- [5] Alitalo Kari , Paavolainen Pekka et all. Clear cell sarcoma of tendons and aponeuroses- report of two cases. Act ortho scand.1977;48:241-244.
- [6] Ferrari Andria , Casanova michela et all. Clear cell sarcoma of tendons and aponeuroses in paediatric patients. American cancer society.2002;94(12):3269-3276.
- [7] Bali Akshay,Roy Maitrayee et all. Cutaneous clear cell sarcoma- A rare aggressive tumour with potential

diagnostic challenge. Journal of lab physicians. 2012 jan-jun;4(1):53-55.

- [8] Poppe Bruce, Forsyth Ramses et all. Soft tissue tumours-Clear cell sarcoma.atlas of genetics and cytogenetics in oncology and haematology.2003;7(1):49-52.
- [9] Creager J. Andrew, Pitman B. Martha et all. Cytologic features of Clear cell sarcoma (malignant melanoma) of soft parts. Am society of clin pathol.2002;117:217-224