

Clear Cell Sarcoma of Soft Parts –A Rare Paediatric Entity

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Abstract: Clear cell sarcoma of tendon and aponeuroses is extremely rare tumour, 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 report a case of clear cell sarcoma of soft tissue arising in the left foot of a 12 year old child.

Keywords: Clear Cell Sarcoma, Soft Parts, Paediatric.

INTRODUCTION

Clear cell sarcoma of tendons and aponeuroses is a rare, high grade malignant soft tissue tumor resembling melanoma and soft tissue sarcomas, and accounts for 1% of all soft tissue sarcomas. It is particularly uncommon in children. We report a case of clear cell sarcoma of soft tissue arising in the left foot of a 12 year old child.

CASE REPORT

A 12 year old boy presented with pain and gradually progressive swelling over left ankle of 3 months duration, along with pain and swelling over left groin of 2 weeks duration. On examination child had a firm, non-tender, non-mobile swelling involving the entire circumference of left ankle joint with bulk of the swelling posteriorly and on both sides of ankle joint obliterating both- medial and lateral malleoli, along with palpable left inguinal nodes 6x6 cm and left popliteal lymph nodes 1x1 cm.

MRI left lower limb showed an infiltrative lesion 5x7 cm in the retrocalcaneal region, eroding posterior cortex of posterior malleolus and posterior aspect of lower femoral epiphysis, along with large left inguinal nodes infiltrating into the vessels (Figure1).

18 F-FDG Whole body PET/CT Scan showed metabolically active malignant mass lesion left ankle retrocalcaneal region with erosion of left fibula and left calcaneum, metabolically active metastatic lymph nodes involving left popliteal and left inguinal nodes.(Figure2) Child was referred as Ewings Sarcoma after biopsy of ankle swelling, and FNAC of inguinal lymph nodes. Pathology review at our institution showed ovoid/polygonal epithelioid rhabdoid cells in nests separated by collagenous stroma with pleomorphic hyperchromatic/vesicular nuclei, prominent nucleoli and moderate amount of eosinophilic cytoplasm.(Figure3) Fine needle aspiration from inguinal lymph node showed malignant cells. (Figure4)

IHC-showed Cytokeratin- (AE1/AE3)-Negative. S-100-diffuse moderate to strong positive. HMB 45-diffuse and strong positive. Melan A-diffuse moderate positivity, desmin-negative.(Figure 5a,b,c) Though histomorphology and immunomarker profile was S/o Malignant Melanoma, in view of the absence of an overlying cutaneous melanocytic lesion, with the swelling confined to the soft tissue, the possibility of Clear Cell Sarcoma with lymph node metastasis was favoured. (Molecular genetic study was not available in-house & hence could not be done) CT-Scan Thorax was normal. Bone scan showed increased localization at left ankle joint. Child was started on neoadjuvant chemotherapy with VAIA (vincristine, actinomycin-D, ifosfamide, and doxorubicin) protocol. Initially there was a reduction in size of the primary lesion, however the lesion began to progress after 5 cycles of chemotherapy and parents opted for alternative forms of treatment.

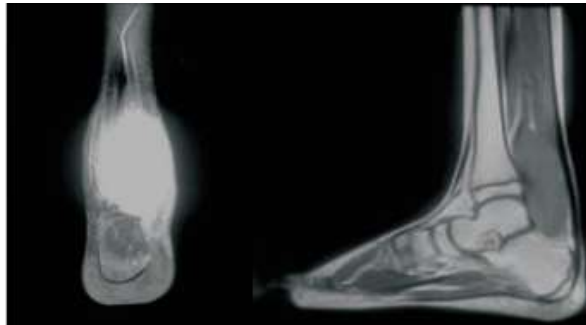


Fig-1: MRI -Irregular relatively well defined altered signal intensity lesion (T2 and STIR hyperintense, T1 iso to hypointense) noted in the posterior compartment of left leg distally, extending posterior to the left ankle joint. Post contrast lesion shows intense enhancement



Fig-2: 18 F-FDG Whole body PET/CT Scan – Metabolically active malignant mass lesion left ankle retrocalcaneal region and metabolically active metastatic lymph nodes involving left popliteal and left inguinal nodes

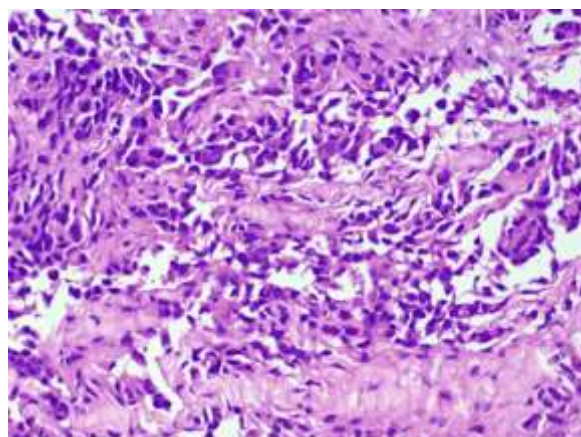


Fig-3: Clear cell sarcoma of soft tissue: Ovoid and polygonal epithelioid-appearing cells in nests separated by collagenous stroma. H&EX200

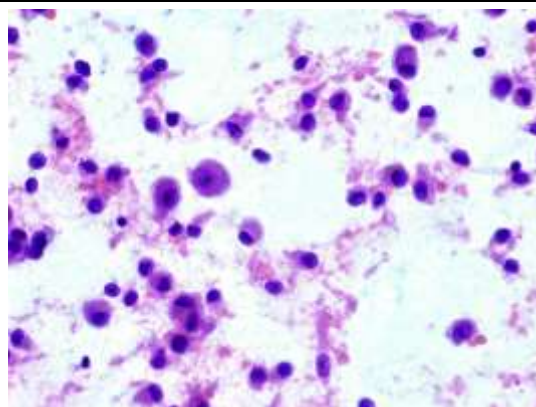


Fig-4: FNA from inguinal lymph node showing malignant cells. Pap stainX400

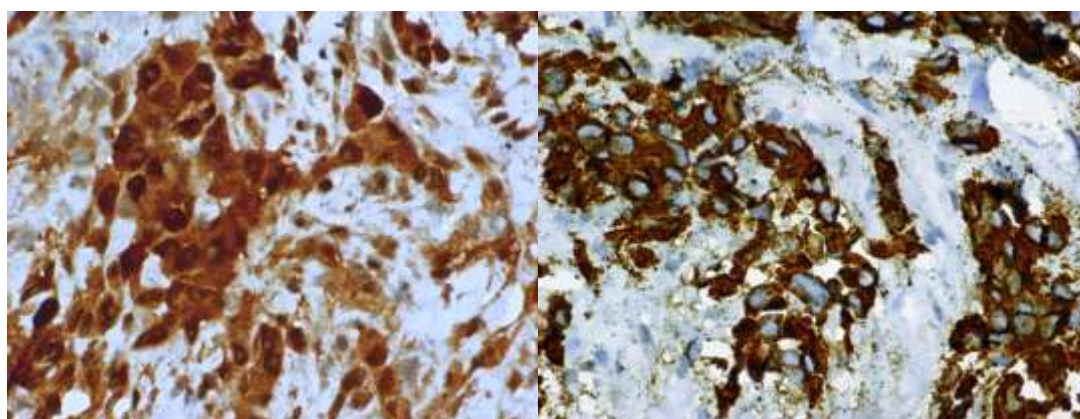


Fig-5a

Fig-5b

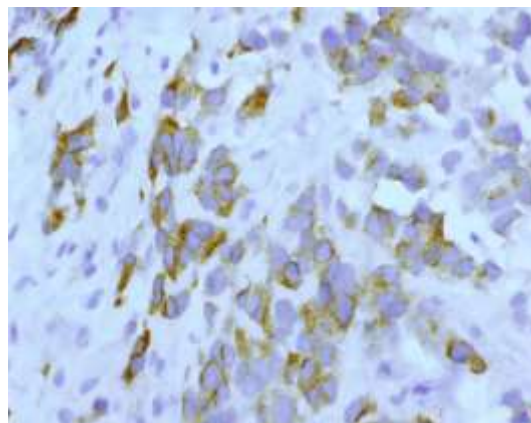


Fig-5c

Fig-5: (a,b,c) Clear cell sarcoma of soft tissue: Diffuse and strong expression of S100 protein(a), HMB45(b), and Melan A(c). IHCX400

DISCUSSION

Clear cell sarcoma (CCS) accounts for less than 1% of all soft tissue tumors[1]. CCS occurs preferentially in adolescents and young adults with predilection for lower extremities (about 70% of cases), in close association with tendons, aponeuroses [2]. The age of patients ranged from 7 to 83 years, with a median age of 27 years, 2% occurs in children younger than 10 years[3]. Due to the histologic features overlapping with malignant melanoma, CCS was originally considered to be a melanoma of soft tissue origin and was called “malignant melanoma of soft parts.”

Compared to melanoma, clear cell sarcoma occurs in much younger patients, is deeply located, associated with tendons or aponeuroses, lacks epidermal involvement, tends to display less cellular pleomorphism, these features are unusual for melanoma. The specific chromosomal translocation t(12;22) (q13; q12) involving DNA transcription factors ATF-1 on chromosome 12 and the EWS gene on chromosome 22 has been detected in 60 –75% of CCS cases[4,5]. This translocation is absent in patients with malignant melanoma.[6,7] The EWSR1-CREB1 gene fusion is less commonly observed in soft tissue CCS but

is more common in gastrointestinal CCS. A normal precursor cell corresponding to the neoplastic elements of CCS has yet to be identified. Clinically, it often presents as a painless slowly enlarging mass that has been present for several months or even years.[8] 10-15% of children with CCS develop regional lymph node metastasis the majority of these patients will develop distant metastasis, most often to lung and bone. Radiologically, CCS typically show a benign looking, well defined, homogenous mass[9].

Complete surgical resection remains the mainstay of treatment for CCS patients. The prognosis for patients with unresected and large tumors is poor, also in relation to the scarce efficacy of adjuvant treatments. In the largest reported series of CCS of tendons and aponeuroses in pediatric patients, 5 year OS and EFS rates were 68.9% and 62.7%, respectively [10]. Long-term follow-up is necessary because of late local recurrences and regional lymph node or distant metastases. Tumor size together with the quality of surgical resection is the most important predictor of survival.

CONCLUSION

Clear cell sarcoma is a rare malignant soft tissue tumour particularly uncommon in children. Complete tumor resection represents the main treatment modality and is the sole treatment required for patients with small tumors. Though rare in children CCS may also be considered in the differential diagnosis of a child presenting with distal extremity tumour with LN metastasis. Awareness of this rare tumor is crucial because it can be mistaken for other types of soft tissue tumors, especially metastatic malignant melanoma, an entity with an entirely different prognosis and management.

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