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Case Report

Clear Cell Carcinoma - A Diagnostically Challenging Presentation at Usual Site

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Abstract

Clear cell sarcoma of soft tissue is an aggressive malignant mesenchymal neoplasm which was previously also called as Malignant Melanoma of soft parts but now the latter terminology is no longer recommended. CCS is an exceedingly rare malignant tumor that originates from neural crest cells and is histologically characterized by clear cells representing intracellular glycogen accumulation. The sarcoma typically involves the deep soft tissues of extremities particularly the distal lower extremities such as ankle and foot where it is usually associated with tendons or aponeurosis. We hereby present a case of Clear Cell Sarcoma in an elderly woman who was initially managed as a case of cellulitis and benign cutaneous ulceration of left heel with antibiotics and topical medications and was only later the diagnosis was confirmed by histopathology and immunohistochemistry.

Keywords: Clear Cell Sarcoma; Neck; Trunk

Introduction

Clear cell sarcoma of soft tissue is a malignant mesenchymal neoplasm which was previously also called as Malignant Melanoma of soft parts but now the latter terminology is no longer recommended. Clear cell sarcoma of soft tissue is an exceedingly rare malignant tumor that originates from neural crest cells and is histologically characterized by clear cells representing intracellular glycogen accumulation [1]. The sarcoma typically involves the deep soft tissues of extremities particularly the distal lower extremities such as ankle and foot where it usually associated with tendons or aponeurosis [2]. The tumor has also been reported in the head and neck, trunk and viscera such as lungs and gastrointestinal tract. CCS resembling Melanoma both clinically and histopathologically have been described [3]. We hereby present a case of Clear Cell Sarcoma in an elderly woman who was initially managed as a case of cellulitis and benign cutaneous ulceration with antibiotics and topical medications and was only later the diagnosis was confirmed by histopathology.

Case Report

A 62-year-old female presented with pain and swelling over the left heel of 3 months' duration and gradually progressed to ulceration. The patient was initially managed with antibiotics and wound dressing but in vain. She was then taken up for the evaluation of a recalcitrant ulcer. She underwent CT scan of the left foot which revealed a focal ulcerating enhancing soft tissue lobulated mass lesion in subcutaneous tissue of heel region with no underlying bone involvement.

An incisional biopsy of the lesion was carried out and submitted for histopathological examination in the department of lab sciences. Haematoxylin and Eosin stained section revealed tumor cells arranged in sheets and nests separated by fibrovascular septae. The cells are polygonal with round to oval pleomorphic nuclei, vesicular chromatin, prominent eosinophilic nucleoli and eosinophilic to clear cytoplasm.

A completion surgery with Wide Local Excision was carried out. The tissue submitted for histopathological examination revealed a single skin lined grayish white tissue measuring 5 x 4 x 2 cm with an ulcer measuring 1.5 x 1.8 x 1 cm. Cut surface of the specimen showed a lobulated tumor mass with a myxoid appearance infiltrating the underlying stroma. No areas of pigmentation were noted. Haematoxylin and Eosin stained sections revealed an ulcerated epithelium with tumor predominantly in the subepithelial stroma comprising of tumor cells arranged in lobules separate by fibrovascular septae. The microscopic examination revealed the features of Clear Cell Carcinoma with margins clear of tumor cells. Focal areas of necrosis were noted. No pigmentation/melanin was appreciated.

Immunohistochemistry on the tumor sections showed the neoplastic cells stained positively for HMB45, S100, Vimentin while stained negatively for SMA, PanCK, p40, Desmin. Ki67 proliferation index was found to be 40-45%. Immunohistochemistry for DNA mismatch repair factors showed the lesion to be microsatellite stable tumor. A diagnosis of Clear cell Carcinoma of left heel was made with the pathological stage classification of pT1NxMx.

Discussion

Clear cell sarcoma is an uncommon soft tissue malignancy that mainly affects young adults between third and fourth decades of life with a slight female predominance [2,4]. The genetic hallmark of the tumor is reciprocal translocation t(12;22)(q13;q12) which is present in 70-90% of the cases [2]. The said translocation results in EWSR1-ATF1 chimeric fusion protein which constitutively activates the promoter of MITF, a target of MSH pathway leading to melanocytic differentiation and growth of Clear cell sarcoma. On immunohistochemistry, the neoplastic cells of Clear cell sarcoma consistently express melanocytic markers including S100, SOX10, Melan-A, HMB45 and MITF [2]. Almost all patients with Clear cell sarcoma present with a palpable mass of months' or even years' duration. Pain and tenderness are reported in only one third to one half of the cases.

Our patient, an elderly woman, presented with palpable mass which progressed to cutaneous ulceration in the region of left heel. The tumor was initially managed as cellulitis and benign cutaneous ulceration but on further evaluation with imaging, the lesion was found to be a neoplastic mass lesion. The lesion was finally diagnosed as a case of Clear Cell Sarcoma of left heel on histopathology assisted by immunohistochemistry.

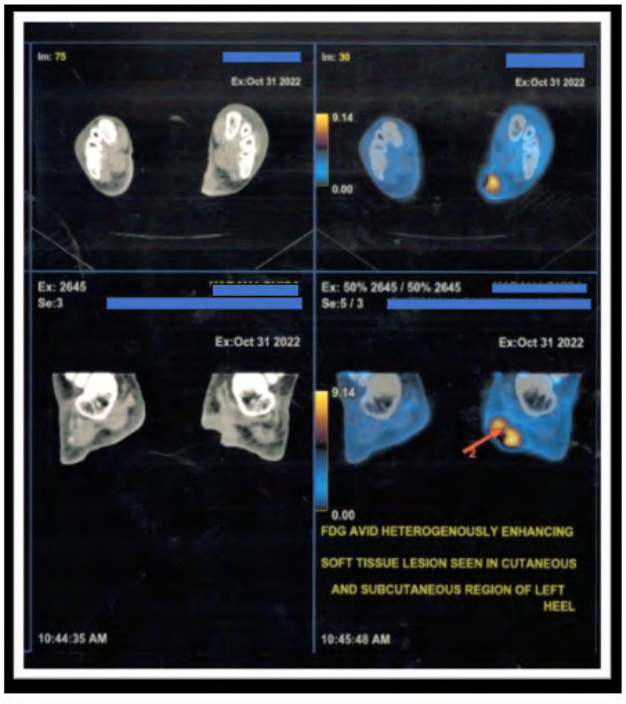

Figure 1: Whole body PET-CT Scan: Metabolically active heterogeneously enhancing soft tissue density lesion noted in cutaneous and subcutaneous region of left heel.

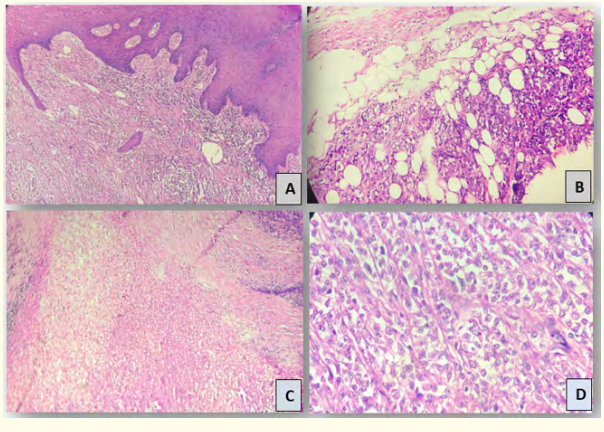

Figure 2: Microphotograph of Hematoxylin and Eosin stained sections of the lesion: A, B- Neoplastic cells in lobules separated by delicate fibrous septae seen infiltrating the subepithelium and the subcutis. C- Foci of necrosis noted. D- Neoplastic cells are polygonal with round to oval pleomorphic nuclei, vesicular chromatin, prominent eosinophilic nucleoli and eosinophilic to clear cytoplasm.

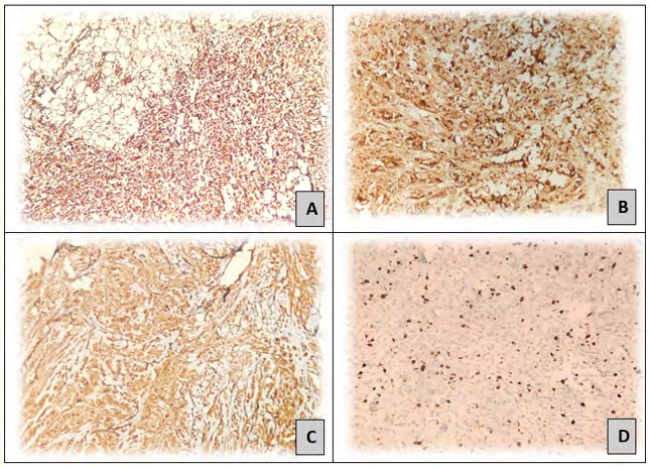

Figure 3: Immunohistochemistry staining of the sections from the lesion: A- SOX10: The neoplastic cells show nuclear positivity; B- S100: The neoplastic cells show nuclear and cytoplasmic positivity; C- MelanA: The neoplastic cells show cytoplasmic positivity; D- Ki67 proliferation index is 40-45%.

Conclusion

Clear cell sarcoma of soft tissue is an uncommon soft tissue malignancy and usually presents in young adults. Our case, though presented in an elderly lady, presented with typical clinical presentation of mass lesion with associated pain and tenderness. Clear cell sarcoma of soft tissue is associated with high incidence of local recurrence and metastasis [2,5]. The case report highlights the importance of evaluating a non-healing ulcer with malignancy as one of the differentials. An early diagnosis with initial radical surgery is the key to the favourable outcome. The survival rate is only 47% despite surgery in many patients underlining the importance of clinical suspicion, early detection, wide surgery and possible use of adjuvant therapy [5].

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