

Case Report

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Malignant Melanoma of Soft Tissue: An Unusual Presentation of Clear Cell Sarcoma of the Heel-A Case Report

Saravana Santhosh A^{1*}, Aarth Kumaraguru², Agniban Chakraborty³ and Anurag Roy⁴

¹Department of General Surgery, Military Hospital Ramgarh Cantt, Jharkhand, India

²Department of Anaesthesiology, Military Hospital Ramgarh Cantt, Jharkhand, India

³Senior Resident, IPGMER & SSKM, Kolkata, India

⁴Department of General Surgery, Military Hospital Gaya, Bihar, India

ABSTRACT

Clear cell sarcoma (CCS) is a rare malignant soft tissue tumor first described by Enzinger in 1965. It demonstrates melanocytic differentiation and shares histological and immunohistochemical similarities with malignant melanoma, which often makes diagnosis challenging. CCS commonly arises from tendons and aponeuroses of the distal extremities, particularly around the foot and ankle. It typically presents as a slow-growing painless mass but may occasionally present atypically, resulting in delayed diagnosis.

We report a case of a 62-year-old female presenting with a chronic non-healing ulcer over the left heel, initially treated as a plantar corn. Incision biopsy revealed clear cell sarcoma confirmed by immunohistochemistry showing positivity for S-100, HMB-45, and vimentin. Imaging studies demonstrated a localized soft tissue lesion without bone involvement or distant metastasis. The patient underwent wide local excision with sentinel lymph node biopsy followed by dermal reconstruction. Histopathology confirmed clear cell sarcoma with tumor-free margins and negative sentinel lymph nodes.

This case highlights the diagnostic challenges associated with atypical presentations of CCS and emphasizes the importance of early biopsy and surgical management to improve outcomes.

*Corresponding author

Saravana Santhosh A, Department of General Surgery, Military Hospital Ramgarh Cantt, Jharkhand, India.

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Introduction

Clear cell sarcoma (CCS) is a rare malignant tumor of soft tissue that originates from melanocytic cells derived from the neural crest. It was first described by Enzinger in 1965 as a tumor arising from tendons and aponeuroses of the extremities [1]. Because of its morphologic and immunohistochemical similarities with malignant melanoma, it has historically been referred to as “malignant melanoma of soft parts” [2].

CCS accounts for approximately **1% of all soft tissue sarcomas** and most frequently occurs in the distal extremities, particularly around the foot and ankle [3]. The tumor usually affects young to middle-aged adults and commonly presents as a slow-growing painless mass [4]. However, atypical presentations may occur, leading to delayed diagnosis.

At the molecular level, CCS is characterized by a specific chromosomal translocation **t(12;22)(q13;q12)** resulting in the **EWSR1-ATF1 fusion gene**, which helps distinguish it from malignant melanoma [5].

Due to its rarity and overlapping features with other melanocytic tumors, diagnosis can be challenging [6]. We present a case of clear cell sarcoma of the heel presenting as a chronic non-healing ulcer which initially mimicked a benign plantar lesion.

Case Presentation

A 62 years old female with a history of hypothyroidism, type II diabetes mellitus, hypertension, and dyslipidemia presented with a non-healing ulcer over the left heel for two years. The lesion became painful in September 2025.

Initially, the lesion was treated as a plantar corn, following which the patient developed a plantar abscess. She underwent incision and drainage, and an incisional biopsy was performed. Histopathological examination revealed clear cell sarcoma, which was positive for S-100, HMB-45, and vimentin with a Ki-67

proliferation index of 15-20%.

On examination, a 04 × 04 cm indurated ulcerated lesion was present over the left heel (Figure 1) with no palpable regional lymphadenopathy.



Figure 1: Non-Healing Ulcer on presentation



Figure 2: Wide Local Excision of lesion with 1cm margin



Figure 3: Excised Sarcoma



Figure 4: Integra Dermal Application at Defect

MRI of the left foot revealed an enhancing ulcerated soft tissue lesion measuring 34 × 23 × 35 mm in the subcutaneous plane abutting the plantar fascia without bone involvement. **MRI plays an important role in evaluating the local extent of soft tissue tumors and guiding surgical planning** [7].

Whole-body PET-CT demonstrated an FDG-avid lesion measuring 41 × 31 × 35 mm with SUVmax of 9.4 and no evidence of distant metastasis. **FDG PET-CT is useful in evaluating metabolic activity and detecting distant metastasis in oncology patients** [8].

The patient underwent sentinel lymph node biopsy of the left groin along with wide local excision of the tumor with a 1 cm margin (Figures 2, 3). Dermal integration using collagen (Figure 4) was applied to reconstruct the defect. Frozen section showed tumor-free margins and negative sentinel lymph nodes.

Histopathological examination revealed nests and sheets of polygonal to spindle-shaped cells separated by fibrovascular septa with abundant eosinophilic to clear cytoplasm and prominent nucleoli.

Immunohistochemistry showed positivity for HMB-45, S-100, and vimentin, confirming clear cell sarcoma with Ki-67 proliferation index of 40-45%. No lymphovascular invasion was identified and sentinel lymph nodes were free of metastasis.

Discussion

Clear cell sarcoma is a rare malignant tumor with melanocytic differentiation that predominantly arises in the distal extremities, particularly around the foot and ankle [3,4]. Because of its slow-growing nature, diagnosis is often delayed. In the present case, the tumor mimicked a corn and presented as a chronic non-healing ulcer, illustrating the diagnostic challenge associated with plantar lesions.

Histologically, CCS is characterized by nests or fascicles of uniform polygonal cells with clear cytoplasm and vesicular nuclei [4]. Immunohistochemical positivity for **S-100 and HMB-45** reflects melanocytic differentiation but also makes differentiation from malignant melanoma difficult [4].

Molecular identification of EWSR1 gene rearrangement can help establish a definitive diagnosis [5].

MRI plays an important role in determining local tumor extent and surgical planning, while PET-CT imaging can help detect distant metastasis and evaluate tumor metabolic activity [7,8].

The mainstay of treatment for CCS is wide local excision with adequate margins [3,9]. Unlike most soft tissue sarcomas, CCS demonstrates a relatively higher incidence of lymph node metastasis [9]. Sentinel lymph node biopsy has therefore been proposed as a useful staging tool for detecting occult nodal metastasis [10].

CCS is associated with a high risk of local recurrence and late metastasis, particularly to the lungs and lymph nodes [3,9]. Tumour size greater than 5 cm, necrosis, and presence of metastasis are considered poor prognostic factors [3]. Long-term surveillance is essential because recurrence may occur several years after treatment.

Conclusion

Clear cell sarcoma is a rare malignant tumor that may present with atypical clinical features such as chronic non-healing plantar ulcers. Early biopsy of suspicious lesions is essential for timely diagnosis. Wide local excision with adequate margins remains the cornerstone of treatment, and sentinel lymph node biopsy may be useful for staging. Long-term follow-up is necessary due to the risk of late recurrence and metastasis.

Consent

Written informed consent obtained.

Ethical Approval

Not required as per institutional policy.

Conflict of Interest

None.

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