Case Report

Epithelioid sarcoma: A rare cause of non-healing ulcer

Paudel V[1], Das AK[2]

[1]Assistant Professor, Department of Dermatology, Venereology and Leprosy, National Medical College, Parsa, Birgunj, Nepal, [2]Associate Professor, Department of Dermatology, Venereology and Leprosy, Maharajgunj Medical Campus, Institute of Medicine, Kathmandu, Nepal

Corresponding Author: Dr. Vikash Paudel, MBBS, MD

Email: luck_vik@hotmail.com, vikashpoudel@iom.edu.np

Abstract

Epithelioid sarcoma, first described by Enzinger in 1970, is a rare slow-growing soft tissue malignancy, associated with a high incidence of regional recurrence and distant metastases. It is characterized by a proliferation of epithelium-like cells and the absence of a granuloma-like pattern with positivity for vimentin, cytokeratin. The recommended treatment is a wide resection with adequate margins, lymph node dissection. We report a 37-year-old male with chronic,non healing ulcer in lower leg of seven years duration who was treated conservatively leading to recurrence of the lesion with metastasis up to left inguinal region resulting lymphedema of the penis.

Keywords: Epithelioidsarcoma, Soft tissue sarcoma, Vimentin, Cytokeratin

Introduction

Soft-tissue sarcomas are rare tumors, accounting for 1 % of all cancers. Epithelioid sarcoma is a rare histological subtype occurring exclusively in the distal extremity.[1,10]It is remarkable for its clinical diversities and histological diagnostic difficulties, resulting in initial misdiagnosis and late onset of treatment. We report a case of epithelioid sarcoma of lower limb presented with non healingulcerwith metastasis leading deformity of penis.

Case Summary

A 37 years married male, presented with non healing ulcer over the left ankle of 7 years duration with swelling of the left leg and and deformity of penis for 1 year. Initially, it had started as an asymptomatic small nodular pea-sized swelling situated above the lateral aspect of left heel which gradually increase in size and started ulcerating of its own with sero- sanguineous discharge within 1 year. There were eruptions of similar nodule up-to the left calf region. He had consulted various centers without improvement and surgical procedure was also performed. But, there was re-growth of nodule and ulceration along with swelling, thickening of lower limb and bending of his peniswith in a year. There is no history of fever, cough, loss of weight, night sweats, discharge from urethra, and sores in the genitals but had difficulty in sexual contact.

On clinical examination, there was an erythematous ulcer above left ankle of 3cm x 3cm size [Figure-1] with rolled pink border, raised edges with presence of few satellite lesions and hyperpigmentation. The base was firm and the floor was covered with granulation tissue. Firm, indurated and non-pitting swellings of left upper thigh and the penis, with firm to hard non matted lymph nodes were present to left inguinal region.[Figure-2]. Histopathological diagnosis showed the dermis showing central area of necrosis surrounded by epithelioid cells in sheets and nests which were polygonal with moderate amount of dense cytoplasm containing vesicular nuclei with small nucleoli. The tumor cells were positive for vimentin and cytokeratin. With the clinical and histological diagnosis of epithelioid sarcoma with metastasis, the patient was referred to the oncology center for managements.

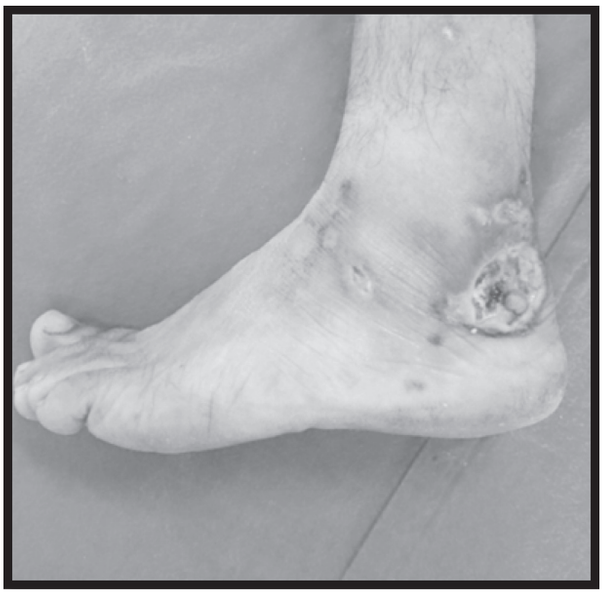

Figure 1 Ulcer in the foot

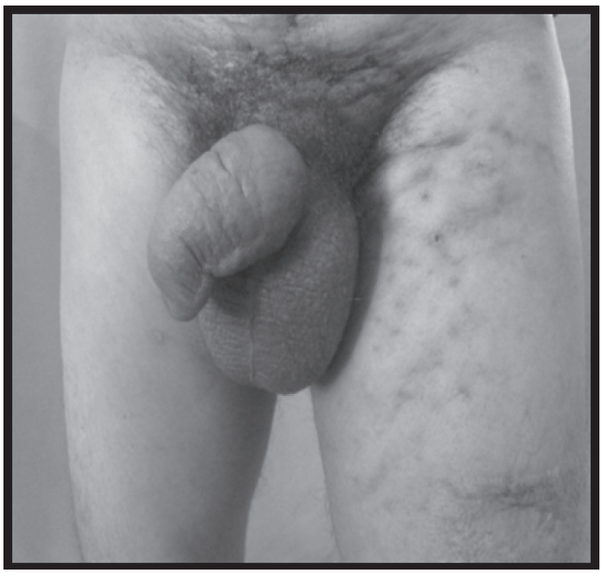

Figure 2 Swelling of thigh and penis

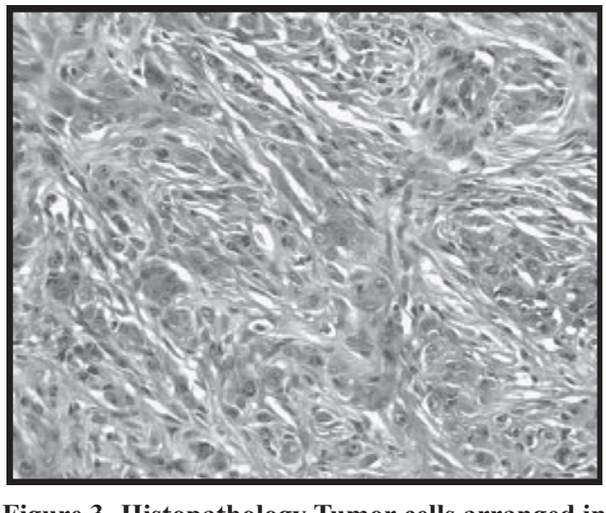

Figure 3- Histopathology Tumor cells arranged in sheets and nests (H&E x100)

Discussion

Epithelioid sarcoma is a rare subtype of soft tissue sarcoma.[1] It was first described by Enzinger in 1970[1], after being described as aponeurotic sarcoma in 1961 by Laskowski [2]. It is a slow-growing tumor presenting as a subcutaneous or deep dermal mass in distal portions of the extremities of adolescents and young adults.[3] It is remarkable for the diagnostic dilemmas both clinically and histologically, often results in misdiagnosis on first encounter and the loss of crucial treatment time[3-4]. Our case had non healing ulcer for a long duration of five years before the correct diagnosis was made. It is a mesenchymal neoplasm with multidirectional differentiation showing reactivity for the epithelial and the mesenchymal markers, cytokeratin, vimentin, epithelial membrane antigen, CD3 with loss of *INI1* expression[5,6]. It has strong characteristic propensity for multi-focal presentation, loco-regional recurrence, ultimately metastasis via lymphatic unlike most soft tissue sarcomas. Local recurrence mainly developed within 1 year after primary treatment but can recur after many years[1]. Distant metastatic disease has been reported in up to 45% of patients with epithelioid sarcoma. The metastasis is usually to the lung and pleura.[1,8] Our patient had local recurrence after 2 years with local metastasis up-to inguinal regional but no distant metastasis were detected. The recommended treatment is a wide resection with adequate margins, lymph node dissection, and optional adjuvant radiotherapy.[9]Chemotherapy is recommended for metastatic disease along with palliative therapy.

Conclusion

Though the tumor is rare, it is remarkable for the diagnostic difficulties, both clinically and histologically, resulting in a high frequency of initial misdiagnosis. So, a suspicious eye should always be present to the benign looking tumors and ulcer for early diagnosis. Optimally, collaborations involving multidisciplinary approach determines effective management strategies.

Conflict of interest: None declared.

References

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