

## A Young man with an Ulcerated Lesion on the Right Ankle

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**Abstract-** We present a case of epitheloid sarcoma that was referred with a 2×3cm ulcerated lesion on the right ankle and edema of the lower leg. Foot drop of the right side was present that had caused walking difficulty. After a few months, he developed several sporthricoidal nodular lesions on the medial aspect of right thigh, inguinal lymphadenopathy, weight loss, anorexia and respiratory symptoms. Chest x-ray and HRCT showed pulmonary metastasis. Histopathological evaluation and immunohistochemical profile of both skin lesion and involved inguinal lymph node were consistent with epitheloid sarcoma. The case is interesting because as far as we know there has been no report of epitheloid sarcoma in literature presenting with foot drop and edema prior to obvious skin involvement.

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**Key words:** Sarcoma; epithelium; ankle

### Introduction

Soft tissue tumors arise in the connective tissue. Epitheloid sarcoma is a relatively rare form of these tumors that occurs more commonly in the extremities. It affects males three times more often than females. We present a case of epitheloid sarcoma that was present with foot drop and edema of the distal extremity prior to obvious skin involvement (1).

### Case Report

A 26-year-old male who referred to our clinic with an ulcerated lesion on the right ankle. He first noted edema of the lower leg a few months prior to his admission. He was otherwise well. In physical examination, there was a 2×3cm ulcer around which there were few satellite lesions. The edema was woody hard on palpation. There was foot drop of the right side that had caused walking difficulty (Figure 1).

Seven months later, he developed several sporthricoidal nodular lesions on the medial aspect of right thigh accompanied by inguinal lymphadenopathy (Figure 2), weight loss, anorexia and respiratory symptoms. Routine laboratory tests were normal. Chest x-ray and HRCT showed bilateral multiple fine pulmonary infiltrations.



**Figure 1.** An ulcerated lesion on the right ankle with edema and foot drop



**Figure 2.** Seven months later: several sporthricoidal nodular lesions on the medial aspect of right thigh accompanied by inguinal lymphadenopathy

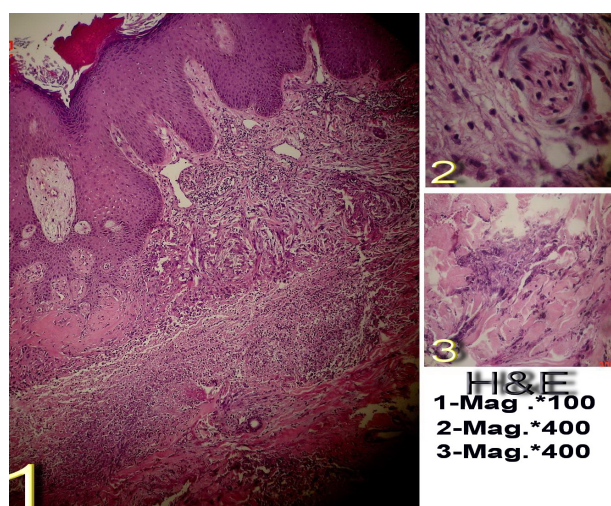
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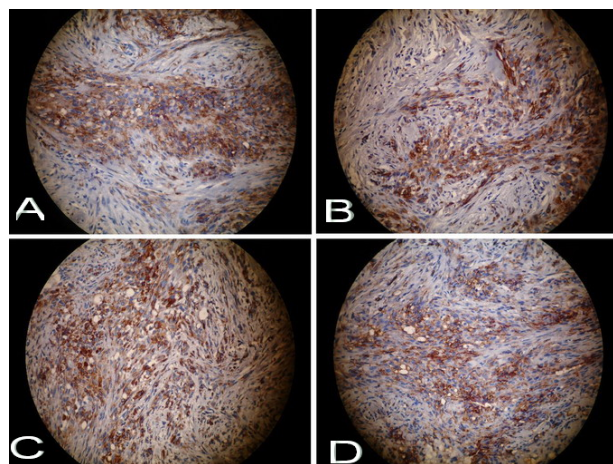
In the leg X-ray, fine calcification of the lesion was seen. The smear and culture of the lesions for mycobacterium and fungi were negative.

Histopathological evaluation showed epidermal acanthosis with ill-defined aggregates of spindle shaped and epithelioid cells that were distributed in upper and deep reticular dermis. Epithelial cells had surrounded a homogenous necrotic zone with associated hyalinized stroma. There were many lymphocytes between collagen bundles; some perineural invasions and calcification were seen (Figure 3).

In immunohistochemical assessment, the majority of dermal infiltrating cells were positive for Vimentin and CD34; the squamous epithelium and dermal infiltrating cells were also positive for epithelial membrane antigen (EMA) and Cytokeratin (Figure 4).



**Figure 3.** 1-Epidermal acanthosis with ill-defined aggregates of spindle shaped and epithelioid cells surrounded a homogenous necrotic zone. 2- Perineural invasion. 3- calcification



**Figure 4.** IHC profile showed relative positivity for vimentin (A), CD34 (B), EMA (C) and CK (D)

Immunohistochemical profile of the involved lymph nodes had also similar pattern. Therefore, the diagnosis was consistent with epithelioid sarcoma with lymph node and pulmonary metastasis. The patient was first referred to a surgeon for radical surgery but he refused it. In the next admission, he had developed pulmonary and lymph node metastases; thus chemotherapy was introduced after which some skin and lung lesions regressed and edema decreased.

Unfortunately, six months later, the skin lesions reappeared more severely, the patient became very cachexic, and few weeks later, he died.

## Discussion

As described by Enzinger in 1970, epithelioid sarcoma is a rare type of soft tissue sarcoma. It affects principally young adult men (median age 23 years).

The tumor usually involves the extremities, particularly the hand, fingers, forearm, pretibial region and foot. It usually presents as firm, flesh colored, indolent nodules which can be ulcerated and involve subcutis and deeper soft tissues particularly fascial planes, aponeurosis and tendon sheaths. Enzinger reviewed the clinicopathological features of 62 cases. Pain or tenderness was usually mild; severe pain, associated with numbness and other sensory changes, was present in three patients in whom the tumor had grown along a large nerve trunk; in two patients, nerve involvement caused severe muscular atrophy; (1) unlike Enzinger's review, our patient had presented with edema and foot drop.

This tumor originates from primitive mesenchymal cells with the capacity for epithelial differentiation (2). Epithelioid sarcoma is often misinterpreted on histopathology and misdiagnosed as granulomatous disease, wart, synovial carcinoma, ulcerating squamous cell carcinoma, amelanotic melanoma, clear cell sarcoma and epithelioid hemangioendothelioma (1,3,4). In histopathology, the principle characteristics are nodular arrangement of tumor cells, their tendency to undergo degeneration and necrosis, and deep acidophilia of the tumor tissue (1). In immunohistochemical profile, epithelioid sarcomas usually express vimentin, cytokeratins and epithelial membrane antigen; about half are positive for CD34 but S100 protein, desmin, and FLI-1 are usually negative (5).

In the large case series of Chase and Enzinger, of 202 cases, 77% recurred after surgery, and 45% metastasized- predominantly to lungs (51%), local

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lymph nodes (34%), scalp (22%) and other skin areas, bone, brain, liver, and pleura (3).

Prognosis is dependent on the depth of the tumor in relation to the deep fascia, local recurrence and regional lymph node involvement. The size of the primary lesion is not a reliable indicator of prognosis, but smaller tumors are associated with significantly better distant metastasis-free interval (4).

Adverse prognostic factors include proximal location, amount of necrosis, vascular invasion and inadequate excision. Favorable factors are young age at the first diagnosis and female sex (3). In conclusion, this tumor is remarkable for the diagnostic difficulties it poses, both clinically and histologically, resulting in a high frequency of initial misdiagnosis. Clinicians should be aware that the initial biopsy may not be constitutive and that repeated biopsy may be necessary (3). In addition, it has a high recurrence rate, which can be reduced by proper surgery as soon as possible.

## References

1. Enzinger F. Epithelioid sarcoma. A sarcoma simulating a granuloma or carcinoma. *Cancer* 1970;26:1029-41.
2. Engers R, Gabbert HE. Mechanisms of tumor metastasis: cell biological aspects and clinical implications. *J Cancer Res Clin Oncol* 2000;126(12):682-92.
3. Chase DR, Enzinger FM. Epithelioid sarcoma. Diagnosis, prognostic indicators, and treatment. *Am J Surg Pathol* 1985;9(4):241-63.
4. Spillane AJ, Thomas JM, Fisher C. Epithelioid sarcoma: The clinicopathologic complexities of this rare soft tissue sarcoma. *Ann Surg Oncol* 2000;7:218-25.
5. Arber DA, Kandalaf PL, Mehta P, Battifora H. Vimentin-negative epithelioid sarcoma. The value of an immunohistochemical panel that includes CD34. *Am J Surg Pathol* 1993;17(3):302-7.