

Epithelioid sarcoma, masquerading as sporotrichosis

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Abstract

Epithelioid sarcoma (ES) is a rare soft tissue neoplasm appearing as multiple slow growing skin nodules. It is a locally invasive tumor, but may metastasize to regional lymph nodes and lungs.

A 19 year old male presented with multiple painful skin nodules, with linear distribution on right upper limb for 2 years. Initial histology mimicked fungal infection, but response to antifungal treatment was poor. His last skin biopsy showed nodular tumor in the dermis with central necrosis and cytological atypia. Immunohistochemistry confirmed the diagnosis of ES. Since he had radiological evidence of lung metastases, chemo radiotherapy was given. Out of the two phenotypes of ES, distal type may show a sporotrichoid pattern. Histology may mimic non neoplastic granulomatous dermatoses. Cells are positive for cytokeratins, epithelial membrane antigen, vimentin and CD34.

Diagnosis would be difficult as ES mimics other common non neoplastic granulomatous dermatoses including sporotrichosis.

Key words: epithelioid sarcoma, sporotrichoid distribution, sporotrichosis

Introduction

Epithelioid sarcoma (ES) is a rare soft tissue neoplasm of dermis and subcutaneous tissue. It clinically appears as multiple slow growing skin nodules, with or without ulcers. It is locally invasive, but may metastasize to regional lymph nodes and lungs.

Sometimes skin lesions show sporotrichoid distribution, which is the arrangement of skin papules, nodules along lymphatic vessels. Sporotrichoid arrangement often associates with lymphocutaneous infections. But rarely neoplasms may present in a similar manner.

Histology of ES has a granulomatous morphology with necrosis, simulating fungal infections and other non-neoplastic granulomatous dermatoses.

Diagnosis is confirmed with immunohistochemistry.

Here we present a case of an ES of a young male whose diagnosis was challenging due to its clinical and histological mimicry of sporotrichosis.

Case

A 19 year old male presented with multiple skin nodules on right upper limb for 2 years. Lesions appeared on palm and gradually progressed upwards. Some spontaneously ulcerated. Skin of the right palm was thickened. He did not have constitutional or systemic symptoms. He denied previous history of trauma to the affected area. His past medical history was insignificant.

On examination he had multiple blackish firm skin papules and nodules with sporotrichoid arrangement on right upper limb. Some of them showed central ulceration. Superficial veins of the same limb had thrombotic nodules (Figure 1, 2). Right axillary and supraclavicular lymph nodes were significantly enlarged. No similar lesions were noted anywhere else including the other arm. System examination was normal.

Initial skin biopsy which was performed 2 years back was reported as a benign cyst. Afterwards the skin nodules were repeatedly excised and some were surgically drained. His second skin biopsy revealed necrobiotic granuloma suggestive of fungal infection. Fungal culture was negative.

Possibility of deep fungal infections including sporotrichosis was considered initially and the patient was treated with a combination of oral antifungals.

As the response to treatment was poor, repeat biopsy was arranged to exclude other lymphocutaneous infections as well as non infective causes including neoplasms.

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Figure 1.



Figure 2.

Histology showed a nodular infiltrative tumor in the dermis with central necrosis, pleomorphic cells of epithelioid nature with eosinophilic cytoplasm and mitoses (Figure 3).

Tumor cells showed cytoplasmic positivity for AE1/AE3 and CK 19 (Figure 4a) and membranous positivity for CD34. (Figure 4b).

Special stains and cultures were negative for fungi, tuberculosis and atypical mycobacteria.

Right upperlimb venous duplex showed superficial thrombophlebitis.

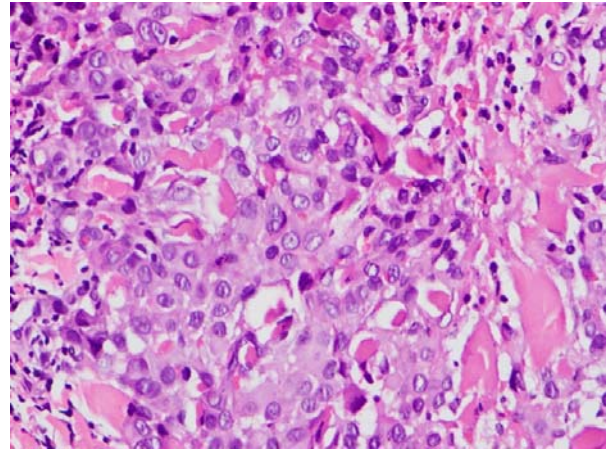


Figure 3.

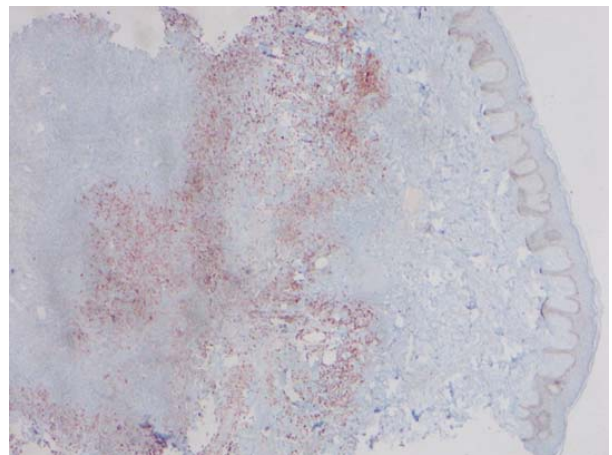


Figure 4a. CK19.

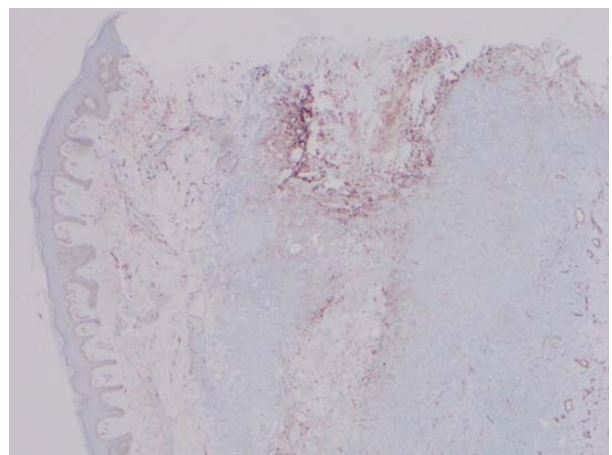


Figure 4b. CD34.

Mantoux test was negative and chest X-ray was normal.

A diagnosis of ES was made after clinicopathological correlation.

Patient underwent contrast enhanced CT of the chest, abdomen and the pelvis, which revealed multiple solid and thin walled cystic lesions scattered throughout both lung fields, consistent with pulmonary metastases.

He was referred to the oncologist, where he was commenced on chemotherapy followed by radiotherapy. Some of his skin nodules healed while some ulcerated. His repeat CT scan after completion of chemotherapy showed persisting lung metastases.

Discussion

Epithelioid sarcoma is first described by Enzinger in 1970¹.

It is a rare tumor which has two distinct clinical phenotypes. Commonly seen classic or distal type affects upper limbs of young individuals. It may appear as multiple skin nodules starting from palms and may show a sporotrichoid pattern along upper limb lymphatics upwards. Sometimes tumor cells invade adjacent veins producing venous thrombosis. Proximal type consists of centrally located nodules involving perineal, pubic, genital and truncal areas^{1,2}.

Sporotrichosis is a fungal infection due to *Sporothrix schenckii*, which presents with papules and ulcerated nodules due to distal inoculation of the organism producing proximal lymphangitis. Other lymphocutaneous infections with similar presentation include nocardia, mycobacterium marinum, leishmaniasis and staphylococci³.

Neoplasms which may present with sporotrichoid skin nodules include keratoacanthoma, squamous cell carcinoma, lymphoma, ES, langerhans cell histiocytosis, melanoma, and peripheral nerve sheath tumors⁴.

Histology of the classic type of ES's has granulomata composed of epithelioid cells, which are polygonal cells with deeply eosinophilic cytoplasm surrounding a necrotic area¹. It mimics fungal infections or necrobiotic granulomatous dermatoses like subcutaneous granuloma annulare, necrobiotic xanthogranuloma and rheumatoid nodule. Presence of mitotic figures helps to differentiate ES from others.

Proximal type lacks the granulomatous appearance and shows cytological pleomorphism and nuclear atypia¹.

Clinical appearance, slow growth and the histological features may confuse the clinician leading to misdiagnosis.

Our patient was initially treated with repeated drainage and antibiotics as for pyogenic infection. Since the histology was of necrobiotic granuloma, later he was managed with antifungals.

Due to poor response, histology was reassessed and combined with immunohistochemistry.

In ES cells are positive for cytokeratins, epithelial membrane antigen, vimentin and CD34¹. Our patient showed positivity for cytokeratins AE1/AE3 and CK19 and for CD34.

Diagnosis and the management of the patient need multidisciplinary approach with the participation of dermatologist, pathologist, radiologist, oncosurgeon and oncologist.

This neoplasm is considered as locally invasive, hence surgical excision with radiotherapy may cure the early stage tumor. But it may recur. If the tumor is inoperable or metastasized, chemo radiotherapy is offered.

Prognosis of epithelioid sarcoma depends on type, size, vascular invasion, resectability and metastasis. Five year survival is 50-70%. Proximal type is more aggressive⁵.

Conclusion

Epithelioid sarcoma mimics non neoplastic dermatoses both clinically and histologically. Therefore, patients with sporotrichoid skin lesion who do not respond to conventional treatment, need to be reassessed to exclude the possibility of this rare tumor.

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