

Case Letter

Epithelioid Sarcoma: Report of 3 Cases

To the Editor:

Epithelioid sarcoma (ES) is a rare aggressive sarcoma of uncertain origin that was first described by Enzinger¹ in 1970. It most commonly presents as a painless nodule on the upper extremities of young adults. We report 3 cases of ES (2 in children) that demonstrate the difficulties encountered in the diagnosis and treatment of this entity.

A 14-year-old adolescent girl presented with an ulcerated nodule on the dorsal surface of the right foot of 3 years' duration (Figure 1). With a clinical diagnosis of dermatofibroma, histologic analysis revealed proliferation of epithelioid cells, showing a focal storiform pattern with marked nuclear pleomorphism and mitotic figures. Cellular proliferations were positive for vimentin, cytokeratin, and epithelial membrane antigen (EMA), and were negative for S-100 and CD31 stains. Two local recurrences occurred over the subsequent 4 years and radical amputation was performed. Follow-up for another 10 years showed no further recurrence.

A 10-year-old girl presented with a small nodule on the first finger of the right hand that had slowly grown in the last year. Histopathology showed a cellular fusiform tumor with focal myxoid and necrotic areas. Cellular areas were positive with cytokeratin, vimentin, and CD34, with a Ki-67 proliferation index greater than 10%. Limited surgery was performed, but follow-up revealed multiple local recurrences. After 7 years, amputation of the forearm was performed. Computed tomography 10 years after initial presentation revealed multiple pulmonary nodules that were histologically confirmed as metastasis. She currently is being treated with chemotherapy with partial response.

A 38-year-old man who was a professional cyclist presented with a painless ulcerlike growth on the back

of his left hand that had been progressing for 4 years (Figure 2). Prior biopsies did not reveal any abnormalities that could lead to a diagnosis. The tumor was composed of several nodules with centrally necrotic zones of a granulomatous appearance (Figure 3). At high magnification, spindle and epithelioid cells with pleomorphic nuclei and clear or eosinophilic cytoplasm were observed (Figure 4), and the biopsy was positive for cytokeratin, vimentin, CD34, and EMA. The patient was treated by surgical amputation and postoperative adjuvant radiotherapy. Two years later he developed multiple lung metastases and was treated with chemotherapy. The patient presented again with a complicated course and died 1 year later.

Epithelioid sarcoma is an uncommon malignancy, representing less than 1% of soft tissue sarcomas.¹ It usually presents as a nondiagnostic papule or a



Figure 1. A solitary ulcerated nodule on the dorsal surface of the right foot.

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The authors report no conflict of interest.

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Figure 2. A painless ulcer on the left hand involved and destroyed the soft tissue and bone.

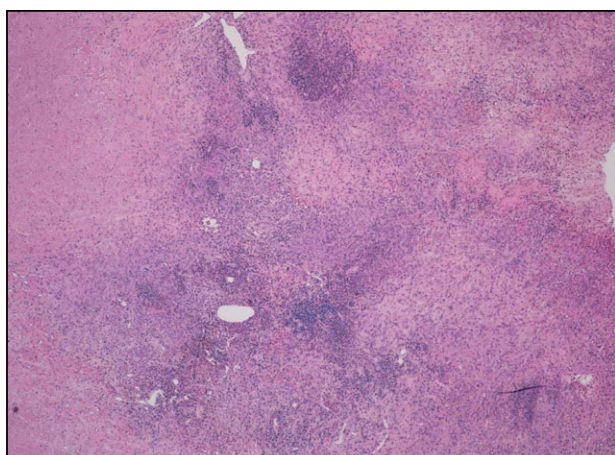


Figure 3. A nodular proliferation with a granulomatous appearance on histopathology (H&E, original magnification ×40).

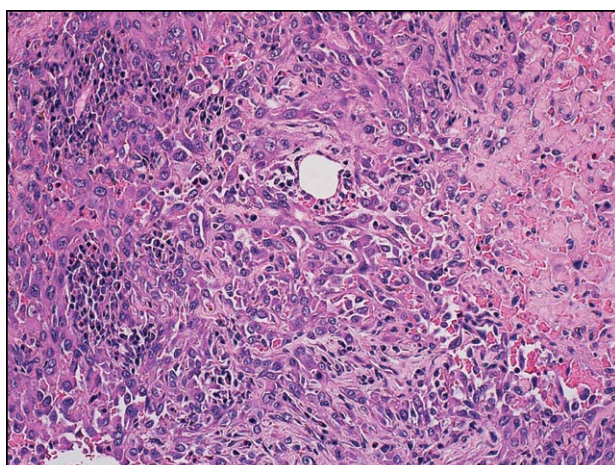


Figure 4. Histology showed a hyperplastic epidermis with hyperkeratosis and parakeratosis. In the upper and mid dermis, a dense lymphocytic infiltrate was present (H&E, original magnification ×100).

solitary, sometimes painful hard nodule. Ulceration occurs in up to 12% of cases.² Diagnosis relies on histopathological and immunohistochemical findings, with a tumor consisting of a nodular proliferation of spindle cells and epithelioid to pleomorphic nuclei, abundant eosinophilic cytoplasm, and a necrotic core with a granulomatous appearance. Neoplastic cells are positive for vimentin, cytokeratin, EMA, and CD34. The coexpression of vimentin and cytokeratin is characteristic of this tumor.³⁻⁵ Local recurrence occurs in 63% to 77% of cases, generally proximal to the area of excision. Metastases are fairly common (45%), with the most common sites being the lymph nodes and lungs.⁶

The key principles that govern the management and prognosis of ES include early suspicion; prompt diagnosis; and aggressive radical excision, which by itself is the most effective treatment in lowering the recurrence rate. Postoperative adjuvant radiotherapy is recommended for large tumors (>5 cm), though chemotherapy is reserved for metastatic disease.

Prophylactic regional lymph node dissection is not recommended.⁷ Although ES is particularly rare in children, clinicians and pathologists should be aware of this possibility and may include ES in the differential diagnosis of chronic lesions of the distal extremities, not only in young adults but also in children.

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