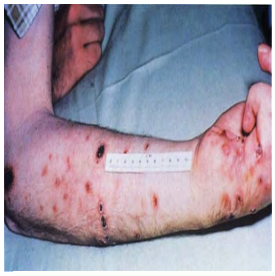

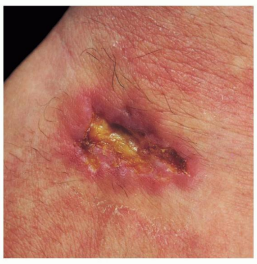

 EPITHELIOID

SARCOMA

Epidemiology

Epithelioid sarcoma (ES), an extremely rare, aggressive sarcoma, occurs most often on the hands and fingers of young males. Proximally located tumors (e.g., groin, thigh, vulva, and axilla) have a much worse prognosis.

Etiology and Pathogenesis

The etiology is obscure; possibly trauma plays some role. Inactivation of the SMARCB1/INI1 tumor suppressor gene was identified in a series of 6 of 11 ES, which may in part explain its pathogenesis or propensity for progression.

Clinical Findings

HISTORY

The patient may present with subcutaneous nodules on the hand or fingers. Often, there is a delay in diagnosis due to the rare nature of this tumor and the resemblance to more common inflammatory skin lesions, such as granuloma annulare.

CUTANEOUS LESIONS

ES usually presents as a firm dermal or subcutaneous nodule, sometimes with ulceration or sinus formation . Linear subcutaneous nodules due to progressive multifocal spread along tendons, nerves, and fascia may give a sporotrichoid appearance.

LABORATORY TESTS

Diagnosis is by biopsy, dermal lesions may be biopsied by punch or wedge biopsy. Most lesions occur in fascia, tendon, or subcutaneous septa; approximately 25 percent arise in the dermis. The tumor usually presents as a multinodular mass composed of epithelioid cells with abundant eosinophilic , often vacuolated, cytoplasm admixed with spindle cells in a collagenous and myxoid stroma . Characteristically, there is central necrosis with a surrounding palisade of tumor cells.

SPECIAL TESTS

Tumor cells express vimentin and epithelial markers. In a large series, epithelial membrane antigen was present in 96 percent of patients, and muscle-specific
actin
and CD34 were noted in 41 percent and 52 percent of the patients, respectively.

Differential Diagnosis

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Prognosis and Clinical Course

The tumor has local recurrence rates of up to 50 percent and nodal or hematogenous metastasis in 40 percent of patients. The lung and pleura are the most common sites of metastatic disease.

Because the classic location on distal extremities often discourages early deep biopsy, and because epithelioid sarcoma may closely mimic various non-neoplastic lesions histologically such as
granuloma
annulare
, it often evades diagnosis at an early, localized stage.

Treatment

Radical excision traditionally has been the treatment of choice, but recurrences occur even with amputation. Conservative (limb-sparing) surgery combined with preoperative or postoperative radiation therapy has been tried, with results comparable with those reported with radical amputation. Chemotherapeutic regimens have not been successful. A high mitotic rate, large areas of necrosis, vascular invasion, and size greater than 5 cm are associated with worse prognosis.