Erythematous and Ulcerated Plaque on the Left Temple

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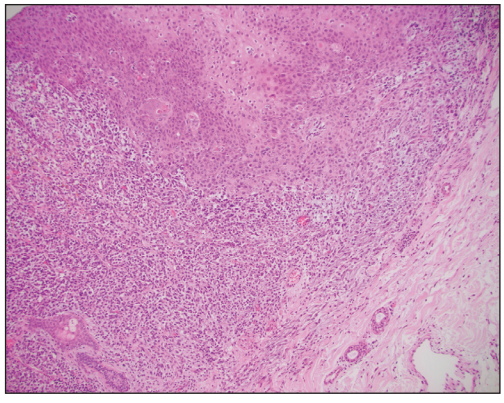

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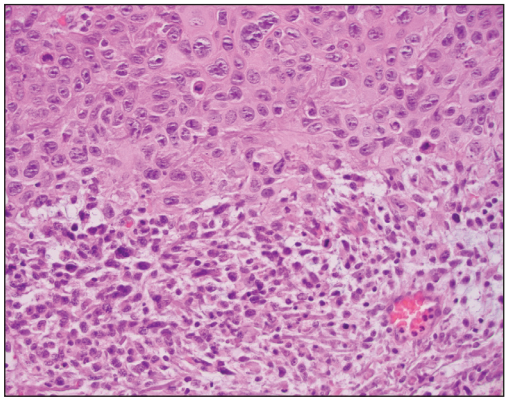

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A 72-year-old man with a history of nonmelanoma skin cancer and lung transplant maintained on stable doses of prednisone and tacrolimus presented with a 1.3×1.8-cm, slow-growing, well-demarcated, ulcerated, erythematous plaque with overlying serous crust on the left temple of 6 months' duration. No cervical or axillary lymphadenopathy was appreciated on physical examination. A biopsy was performed followed by Mohs micrographic surgery. Microscopic examination of the debulking specimen revealed atypical spindle cells in the papillary and reticular dermis radiating from a central focus of a moderately differentiated squamous cell carcinoma. The squamous cells stained positive for cytokeratin 5/6, pankeratin, and p40, while the spindle cells stained positive only for vimentin.

THE BEST **DIAGNOSIS IS:**

a. atypical fibroxanthoma
b. pleomorphic leiomyosarcoma
c. primary cutaneous carcinosarcoma
d. primary cutaneous myoepithelial carcinoma
e. spindle cell squamous cell carcinoma

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THE **DIAGNOSIS:** Primary Cutaneous Carcinosarcoma

The immunohistochemical findings supported an epithelial component consistent with moderately differentiated squamous cell carcinoma (SCC) and a mesenchymal component with features consistent with a sarcoma. Consequently, the lesion was diagnosed as a primary cutaneous carcinosarcoma (PCCS).

Primary cutaneous carcinosarcoma is a rare biphasic neoplasm consisting of malignant epithelial (carcinoma) and mesenchymal (sarcoma) components.[1] Primary cutaneous carcinosarcomas are uncommon, poorly understood, primary cutaneous tumors.[2,3] Characteristic of this tumor, cytokeratins highlight the epithelial component while vimentin highlights the mesenchymal component.[4] Histologically, the sarcomatous components of PCCS often are highly variable, with an absence of transitional areas within the epithelial component, which frequently resembles basal cell carcinoma and/or SCC.[5-7] Primary cutaneous carcinosarcoma favors areas of chronic UV radiation exposure, particularly on the head and neck. Most tumors present with a slowly growing, polypoid, flesh-colored to erythematous nodule due to the infiltrative mesenchymal component.[7] Primary cutaneous carcinosarcoma primarily is diagnosed in elderly patients, with the majority of cases diagnosed in the eighth or ninth decades of life (range, 32–98 years).[1,8] Men appear to be twice as likely to be diagnosed with a PCCS compared to women.[1] Primary cutaneous carcinosarcomas are recognized as aggressive tumors with a high propensity to metastasize and recur locally, necessitating early diagnosis and treatment.[4] Accurate diagnosis of PCCSs can be challenging due to the biphasic nature of the neoplasm as well as poor differentiation or unequal proportions of the epithelial and mesenchymal components.[5] Additionally, overlapping diagnostic criteria coupled with vague demarcation between soft-tissue sarcomas and distinct carcinomas also may contribute to a delay in diagnosis.[9] Treatment is achieved surgically by complete wide resection, with no evidence to support the use of adjuvant or neoadjuvant external beam radiation therapy. Due to the small number of reported cases, no treatment recommendations currently exist.[1]

Surgical management with wide local excision has been disappointing, with recurrence rates reported as high as 33%.[6] Primary cutaneous carcinosarcoma has an estimated overall recurrence rate of 19% and a 5-year disease-free rate of 50%.[10] Risk factors associated with poorer prognosis include tumors with adnexal subtype, age less than 65 years, rapid tumor growth, a tumor greater than 20 mm at presentation, and a long-standing tumor lasting up to 30 years.[2,4] Although wide local excision and Mohs micrographic surgery (MMS) both have been utilized successfully, MMS has been shown to result in a cure rate of greater than 98%.[6]

Atypical fibroxanthoma (AFX) is a cutaneous tumor of fibrohistiocytic mesenchymal origin that typically manifests on sun-damaged skin in elderly individuals. Clinically, it presents as a rapidly growing neoplasm that often ulcerates and bleeds. These heterogenous neoplasms have several distinct characteristics, including dense cellularity with disorganized, large, pleomorphic, and atypical-appearing spindle-shaped cells arising in the upper layers of the dermis, often disseminating into the reticular dermis and occasionally into the subcutaneous fat (Figure 1). The neoplastic cells often exhibit hyperchromic and irregular nuclei, multinucleated giant cells, and atypical mitotic figures. In most cases, negative immunohistochemical staining with SOX-10, S-100, cytokeratins, desmin, and caldesmon will allow pathologists to differentiate between AFX and other common tumors on the differential diagnosis, such as SCC, melanoma, and leiomyosarcoma. CD10 and procollagen type 1 are positive antigenic markers in AFX, but they are not specific. The standard treatment of AFX includes wide local excision or MMS for superior margin control.[11]

Spindle cell SCC presents as a raised or exophytic nodule, often with spontaneous bleeding and central ulceration. It usually presents on sun-damaged skin or in individuals with a history of ionizing radiation. Histologically, it is characterized by atypical spindle-shaped keratinocytes in the dermis existing as single cells

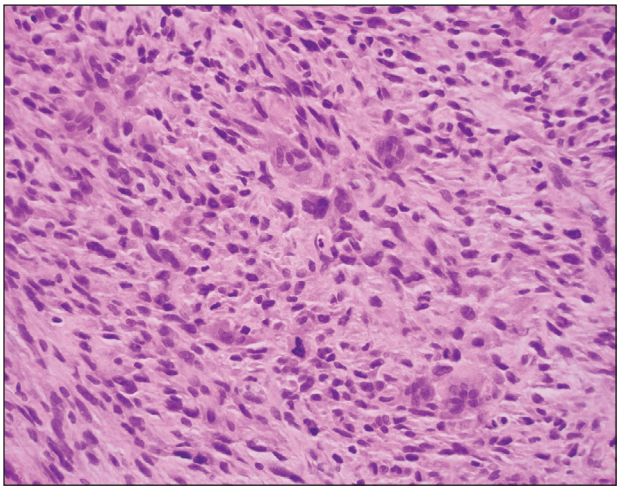

FIGURE 1. Atypical fibroxanthoma. The cells are arranged in a haphazard pattern and are accompanied by giant cells. The tumor cells display nuclear hyperchromasia with prominent atypical mitoses (H&E, original magnification ×40).

or cohesive nests along with keratin pearls (Figure 2). The atypical spindle cells may comprise the entire tumor or only a small portion. The use of immunohistochemical markers often is required to establish a definitive diagnosis. Spindle cell SCC stains positively, albeit frequently focally, for p63, p40, and high-molecular-weight cytokeratins such as cytokeratin 5/6, while S-100 protein, SOX-10, MART-1/Melan-A, and muscle-specific actin stains typically are negative. Wide local excision or MMS is recommended for treatment of these lesions.[12]

Primary cutaneous myoepithelial carcinomas are uncommon neoplasms of myoepithelial differentiation. Clinically, they often arise as soft nodular lesions on the head, neck, and lower extremities with a bimodal age distribution (<21 years and >50 years). Histologically, cutaneous myoepithelial tumors are well-differentiated, dermal-based nodules without connection to the overlying epidermis (Figure 3). The myoepithelial cells can exhibit spindled, epithelioid, plasmacytoid, or clear cell morphologic features and show variability in cell growth patterns. One of the most common growth patterns is oval to round cells forming cords and chains in a chondromyxoid stroma. Most cases display an immunophenotyped co-expression of an epithelial cytokeratin and S-100 protein. Myoepithelial markers also may be present, including keratins, smooth muscle actin, calponin, glial fibrillary acidic protein, p63, and desmin. Surgical removal with wide local excision or MMS is essential.[13]

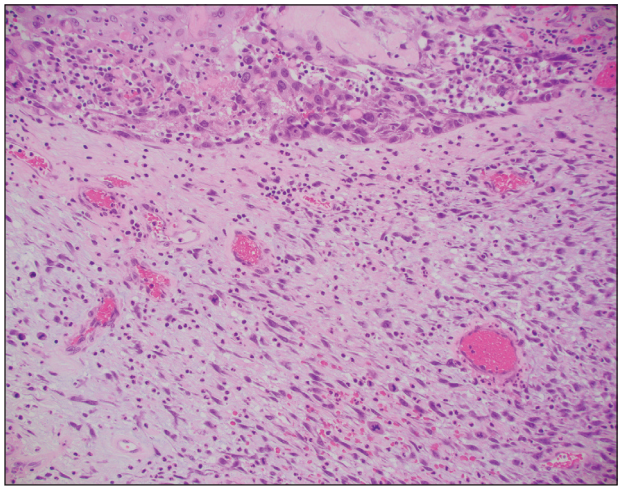

FIGURE 2. Spindle cell squamous cell carcinoma. The tumor contains spindle-shaped cells with prominent mitoses, dyskeratosis, and keratin (H&E, original magnification ×20).

Leiomyosarcoma (LMS) is a tumor that originates from smooth muscle and rarely develops in the dermis.[14] Pleomorphic LMS is a morphologic variant of LMS that has a low propensity to metastasize but commonly exhibits local recurrence.[15] Leiomyosarcoma can present in any age group but most commonly manifests in individuals aged 50 to 70 years. Clinically, LMS presents as a firm solitary nodule with a smooth pink surface or a more exophytic tumor with a reddish or brown color on the extensor surface of the lower limbs; it is less common on the scalp and face.[14] Histologically, most cases of pleomorphic LMS show small foci of fascicles consisting of smooth muscle tumor cells in addition to cellular pleomorphism (Figure 4).[15] Many of these cells demonstrate a clear perinuclear vacuole that generally is appreciated in neoplastic smooth muscle cells.[14] Pleomorphic LMS typically stains positively for at least one smooth muscle marker including desmin, h-caldesmon, muscle-specific actin, α-smooth muscle actin, or smooth muscle myosin in the leiomyosarcomatous fascicular areas.[16] Complete surgical excision is the treatment of choice, and the best results are obtained with MMS.[14]

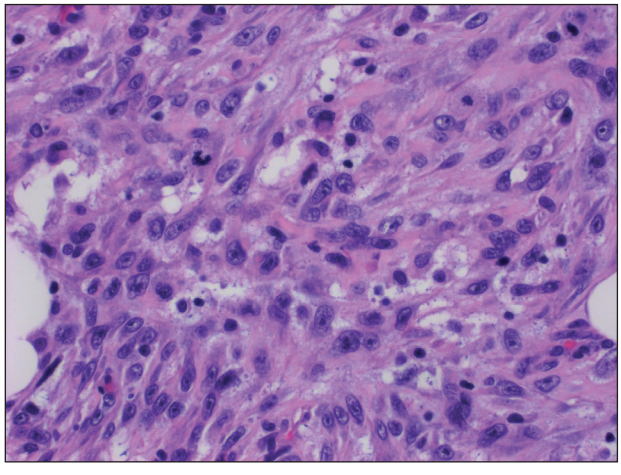

FIGURE 3. Myoepithelial carcinoma. There are cords of spindle cells in hyalinized stroma. The spindle cells have severe atypia with coarse chromatin and prominent nucleoli (H&E, original magnification ×40).

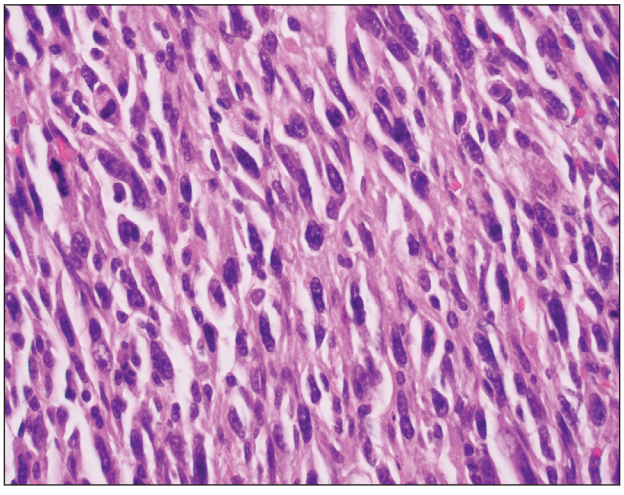

FIGURE 4. Leiomyosarcoma. The spindle cells display a fascicular growth pattern with cigar-shaped nuclei and perinuclear vacuoles (H&E, original magnification ×60).

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