Painful, Purpuric, Nodular Lesion With an Irregular Surface on the Shoulder

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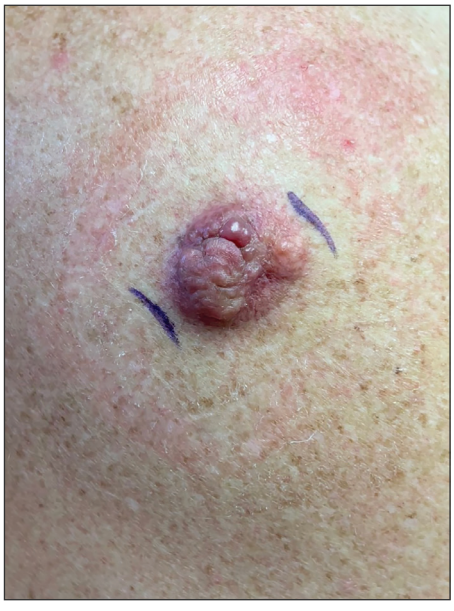

A 53-year-old man presented to the dermatology clinic for evaluation of a painful, purpuric, nodular lesion on the left shoulder of 3 months' duration. The lesion had an irregular surface that was surrounded by an erythematous ring. Biopsy revealed fascicles of eosinophilic cells within the dermis. The nuclei were heterogeneous in size and shape and had blunted ends. Frequent atypia and mitotic figures were observed, and the lesion extended into the subcutis. Immunostaining was positive for desmin and smooth muscle actin and negative for SOX10, Melan-A, PRAME (preferentially expressed antigen in melanoma), CD34, and Factor XIIIa.

WHAT'S YOUR **DIAGNOSIS?**

a. atypical fibroxanthoma
b. basal cell carcinoma
c. cutaneous leiomyosarcoma
d. dermatofibrosarcoma protuberans
e. nodular melanoma

PLEASE TURN TO PAGE 122 FOR THE DIAGNOSIS

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The authors have no relevant financial disclosures to report.

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Cutis. 2026 April;117(4):108, 122. doi:10.12788/cutis.1370

THE **DIAGNOSIS:** Cutaneous Leiomyosarcoma

Based on the clinical and histopathologic findings, our patient was diagnosed with primary cutaneous leiomyosarcoma (LMS), a rare soft-tissue neoplasm that arises from smooth muscle and typically manifests as a firm pink nodule.[1] The neoplasm may occur in the area of a prior traumatic injury or develop spontaneously without an identifiable cause.[1-3] Cutaneous LMS represents 2% to 3% of all soft-tissue sarcomas worldwide, with an estimated incidence of 1 in 500,000 annually.[1,4] Men who are in their fifth to seventh decades of life are at the highest risk for LMS.[1]

Histologically, cutaneous LMS can be subclassified as dermal, which has a low metastatic risk and excellent prognosis, or subcutaneous, which is associated with poorer outcomes and vascular muscle origin.[1] In our case, hematoxylin and eosin staining revealed fascicles of smooth muscle fibers with hypercellularity, atypia, and mitotic figures (Figure). The neoplasm stained positive for desmin, vimentin, and smooth muscle actin and negative for SOX10, Melan-A, PRAME (preferentially expressed antigen in melanoma), CD34, and Factor XIIIa.[1]

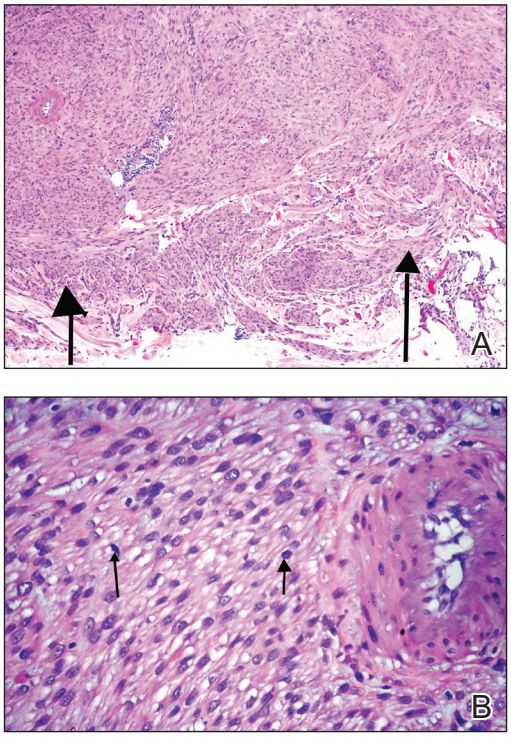

FIGURE. Cutaneous leiomyosarcoma. A, On histopathology, low-power view showed a dermal spindle-cell neoplasm with interlacing fascicles of fusiform leiomyocytes (arrows)(H&E, original magnification ×4). B, High-power view showed striking nuclear atypia in spindle cells with a high mitotic index, including numerous atypical mitotic figures (arrows)(H&E, original magnification ×40).

Standard treatment for LMS is surgical excision.[5] Poor prognostic factors include lesions with a diameter of 5 cm or larger, deep subcutaneous tumor invasion, and distant metastases.[2,5]

The differential diagnosis may include dermatofibrosarcoma protuberans, which can have a similar pink nodular appearance and also may manifest after injury[6]; however, this lesion would stain positive for CD34 on histopathology.[1] Nodular melanoma also can manifest as a solitary red, raised lesion, but it would stain positive for SOX10, PRAME, and Melan-A on histopathology.[7] Basal cell carcinoma, which also may have a similar clinical appearance, is associated with nests of basaloid cells and palisading nuclei histologically.[8] Lastly, atypical fibroxanthoma also manifests as a red nodule or plaque and is associated with atypical mitotic figures on histology; however, it notably stains negative for desmin.[9]

In summary, cutaneous LMS should be included in the differential diagnosis for raised, pink nodules. Given its nonspecific clinical presentation, this rare and malignant neoplasm requires biopsy and immunohistochemical staining for accurate diagnosis.

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