Reddish Nodule on the Left Shoulder

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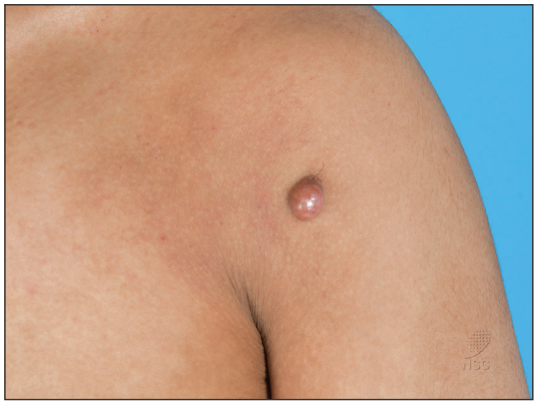

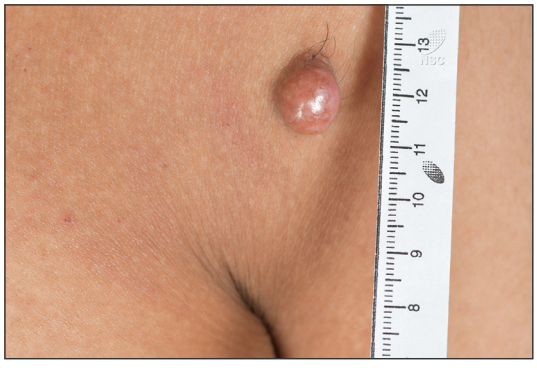

A 20-year-old man presented to the dermatology clinic for evaluation of a slow-growing nodule on the left shoulder of 1 year's duration. The patient reported a history of eczema since childhood, which had been treated by an external physician with cyclosporine and methotrexate; however, exact treatment records were unavailable as the patient had been treated at another institution. The eczema had been well controlled over the past year on topical steroids alone. The nodule was asymptomatic, and the patient denied any history of trauma or acne at the affected site. He also denied any family history of similar nodules or other notable skin findings. Physical examination revealed a well-circumscribed, 15×12-mm, firm, flesh-colored to reddish nodule on the left shoulder with a slightly whitish center. An excisional biopsy was performed.

WHAT'S YOUR **DIAGNOSIS?**

a. amelanotic melanoma
b. atypical fibroxanthoma
c. dermatofibrosarcoma protuberans
d. keloid scar
e. pilomatricoma

PLEASE TURN TO PAGE 74 FOR THE DIAGNOSIS

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THE **DIAGNOSIS:** Atypical Fibroxanthoma

Given the appearance of the nodule and the absence of features of a keloid scar, a soft-tissue or adnexal tumor was suspected. Histology revealed a thin epidermis with loss of rete ridges and a Grenz zone. There was a nodular uncircumscribed dermal proliferation of spindle cells forming interweaving fascicles with elongated ovoid nuclei and prominent nucleoli (Figure). There was moderate cellular and nuclear atypia, and no necrosis was observed. The spindle cells stained positive for CD10 and negative for AE1/AE3, cytokeratin 5/6, S100, melanoma triple marker, Factor XIII 1, ERG, CD31, CD34, desmin, and smooth muscle actin; ERG, CD31, CD34, and SMA highlighted small vessels within the tumor. The histologic diagnosis was an atypical spindle cell tumor favoring atypical fibroxanthoma (AFX). The excisional biopsy margins were clear.

The patient was referred to surgical oncology to consider re-excision of margins after the diagnosis was made. A chest radiograph was clear, and magnetic resonance imaging showed mild skin thickening and image enhancement at the left shoulder—possibly a postsurgical change—with no nodularity suggesting a residual or recurrent tumor. Surgical oncology determined that the patient did not require further excision and placed him on regular follow-up every 2 to 3 months for the next 2 years.

Atypical fibroxanthoma is a primary dermal neoplasm of uncertain origin that is considered to be on a spectrum with the more aggressive pleomorphic dermal sarcoma (PDS); it can be distinguished from PDS by histologic features such as nerve or vessel invasion.[1] Both entities share oncogenes (eg, tumor protein 53 gene mutations) and are histologically and immunohistochemically similar. Atypical fibroxanthoma largely is viewed as an intermediate-risk tumor that is locally aggressive but rarely metastasizes, with a reported local recurrence rate of 5% to 11% and metastasis risk of 1% to 2%. Conversely, PDS is a more aggressive diagnosis with a high risk for local recurrence and metastasis (7%-69% and 4%-20%, respectively).[1]

Atypical fibroxanthomas may mimic other entities, both clinically and histologically. It commonly manifests as a flesh-colored to erythematous, sometimes ulcerated nodule on sun-exposed skin in elderly patients, leading to a broad range of clinical differential diagnoses, including other primary cutaneous malignancies (eg, squamous cell carcinoma, amelanotic melanoma), cutaneous sarcomas (eg, dermatofibrosarcoma protuberans), adnexal and other tumors (eg, pleomorphic fibroma, pilomatricoma), cutaneous metastases, and even keloid scars. As the differentials can look clinically similar, a skin biopsy may be necessary to confirm the diagnosis.

Histologically, AFX tends to show an undifferentiated pleomorphic spindle cell morphology. Notably, histology can be highly variable, with other reported histologic patterns including keloidlike, pleomorphic, epithelioid, rhabdoid, clear-cell, foamy cell, granular cell, bizarre cell, pseudoangiomatous, inflammatory, and osteoclast-rich patterns.[2] Thus, the histologic differential diagnosis also is broad, and AFX primarily is a diagnosis of exclusion without specific immunohistochemical markers that serve to exclude other diagnoses. For example, AFX tends to stain positive for CD10 and CD68, though these are not specific markers for AFX. Furthermore, although certain histologic markers may commonly be more positive in AFX than PDS (eg, CD74 stains positive in 20% of AFXs and only 1% of PDSs), this is not reliable enough to be diagnostic.[3] As such, AFX is distinguished from PDS primarily by histologic features such as subcutaneous tissue invasion, vascular or perineural invasion, necrosis, or local invasion/metastases.[1] Given the rarity of both tumors, no established management guidelines exist, although excision (wide local excision or Mohs micrographic surgery) usually is recommended, with some authors suggesting margins of 1 cm for AFX and 2 cm to 3 cm for PDS.[1]

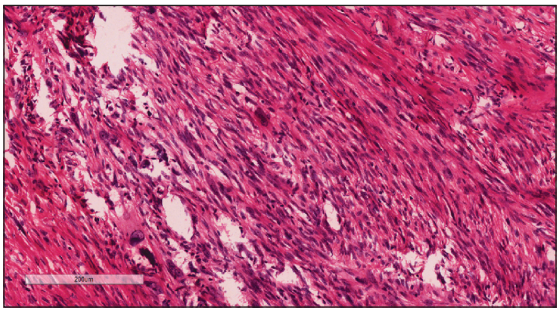

FIGURE. Atypical fibroxanthoma. A nodular uncircumscribed dermal proliferation of spindle cells forming interweaving fascicles with elongated ovoid nuclei and prominent nucleoli. Reference bar indicates 200 μm.

This atypical case of AFX arising in non–sun-exposed skin in a young man raises questions about whether unknown genetic factors or possibly prior immunosuppression could have contributed to the development of the tumor. A thorough history and physical examination can provide valuable clues for biopsy, including ongoing growth, absence of known prior trauma or acne at the site, and clinical appearance, such as the reddish, solitary, dome-shaped lesion in our patient.

REFERENCES

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