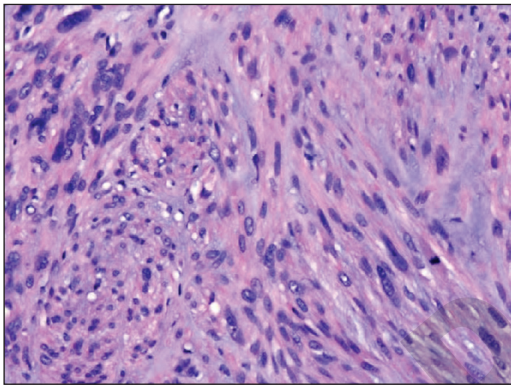


Firm Exophytic Tumor on the Shin

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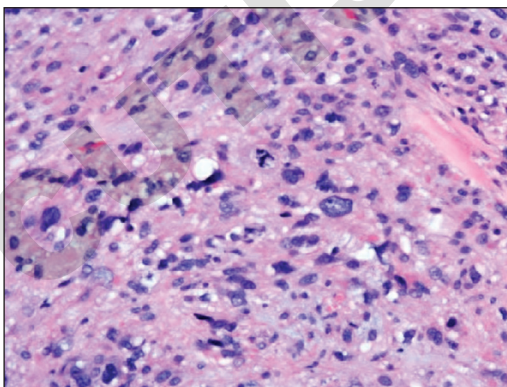


H&E, original magnification $\times 200$.

A 62-year-old man presented with a firm, exophytic, 2.8 \times 1.5-cm tumor on the left shin of 6 to 7 years' duration. An excisional biopsy was obtained for histopathologic evaluation.

THE BEST DIAGNOSIS IS:

- atypical fibroxanthoma
- leiomyosarcoma
- melanoma
- spindle cell squamous cell carcinoma
- undifferentiated pleomorphic sarcoma



H&E, original magnification $\times 200$.

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THE DIAGNOSIS: Leiomyosarcoma

Cutaneous leiomyosarcomas are relatively rare neoplasms that favor the head, neck, and extremities of older adults.¹ Dermal leiomyosarcomas originate from arrector pili and are locally aggressive, whereas subcutaneous leiomyosarcomas arise from vascular smooth muscle and metastasize in 30% to 60% of cases.² Clinically, leiomyosarcomas present as solitary, firm, well-circumscribed nodules with possible ulceration and crusting.³ Histopathology of leiomyosarcoma shows fascicles of atypical spindle cells with blunt-ended nuclei and perinuclear glycogen vacuoles, variable atypia, and mitotic figures (quiz images). Definitive diagnosis is based on positive immunohistochemical staining for desmin and smooth muscle actin.⁴ Treatment entails complete removal via wide local excision or Mohs micrographic surgery.⁵

Atypical fibroxanthoma (AFX) is a malignant fibrohistiocytic neoplasm that arises in the dermis and preferentially affects the head and neck in older individuals.³ Atypical fibroxanthoma presents as a nonspecific, pink-red, sometimes ulcerated papule on sun-damaged skin that may clinically resemble a squamous cell carcinoma (SCC) or basal cell carcinoma.⁶ Histopathology shows pleomorphic spindle cells with hyperchromatic nuclei and abundant cytoplasm mixed with multinucleated giant cells and scattered mitotic figures (Figure 1). Immunohistochemistry is essential for distinguishing AFX from other spindle cell neoplasms. Atypical fibroxanthoma stains positively for vimentin, procollagen-1, CD10, and CD68 but is negative for S-100, human melanoma black 45, Melan-A, desmin, cytokeratin, p40, and p63.⁶ Treatment includes wide local excision or Mohs micrographic surgery.

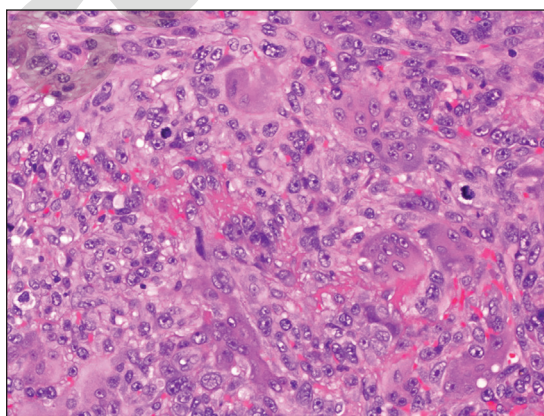


FIGURE 1. Atypical fibroxanthoma. Markedly atypical cells, giant cells, and scattered mitotic figures (H&E, original magnification $\times 200$).

Melanoma is an aggressive cancer with the propensity to metastasize. Both desmoplastic and spindle cell variants demonstrate atypical spindled melanocytes on histology, and desmoplasia is seen in the desmoplastic variant (Figure 2). In some cases, evaluation of the epidermis for melanoma in situ may aid in diagnosis.⁷ Clinical and prognostic features differ between the 2 variants. Desmoplastic melanomas usually present on the head and neck as scarlike nodules with a low rate of nodal involvement, while spindle cell melanomas can occur anywhere on the body, often are amelanotic, and are associated with widespread metastatic disease at the time of presentation.⁸ SOX10 (SRY-box transcription factor 10) and S-100 may be the only markers that are positive in desmoplastic melanoma.^{9,10} Treatment depends on the thickness of the lesion.¹¹

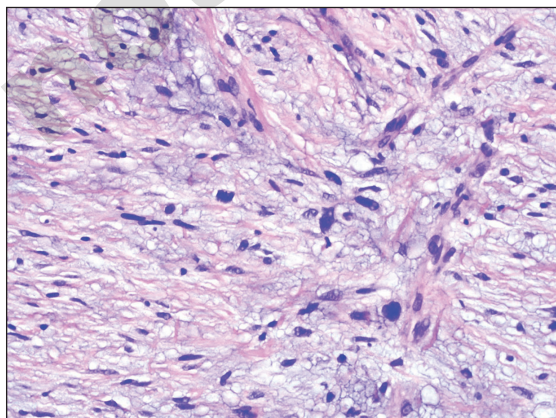


FIGURE 2. Desmoplastic melanoma. Scattered atypical spindle cells in elastotic dermis with desmoplastic reaction (H&E, original magnification $\times 200$).

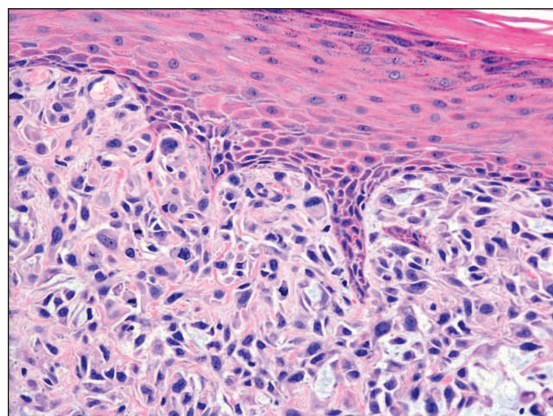


FIGURE 3. Spindle cell squamous cell carcinoma. Atypical spindle cells with eosinophilic cytoplasm (H&E, original magnification $\times 200$).

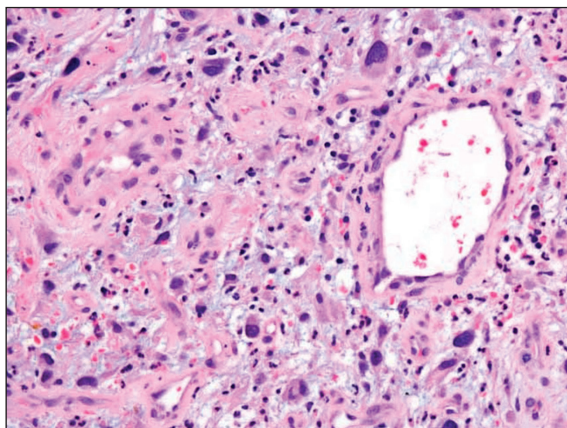


FIGURE 4. Undifferentiated pleomorphic sarcoma. Markedly atypical pleomorphic cells (H&E, original magnification $\times 200$).

Spindle cell SCC is a histologic variant of SCC characterized by spindled epithelial cells. Spindle cell SCC typically presents as an ulcerated or exophytic mass in sun-exposed areas or areas exposed to ionizing radiation, or in immunocompromised individuals. Histopathology shows spindled pleomorphic keratinocytes with elongated nuclei infiltrating the dermis and minimal keratinization (Figure 3).¹² Immunohistochemistry is necessary to distinguish spindle cell SCC from other spindle cell tumors such as spindle cell melanoma, AFX, and leiomyosarcoma. Spindle cell SCC is positive for high-molecular-weight cytokeratin, p40, and p63. Mohs micrographic surgery provides the highest cure rate, and radiation therapy may be considered when clear surgical margins cannot be obtained.⁶

Undifferentiated pleomorphic sarcoma (UPS) (formerly known as malignant fibrous histiocytoma) describes tumors that resemble AFX but are more invasive. They commonly involve the soft tissue with a higher risk for both recurrence and metastasis than AFX.¹³ Histopathology shows marked cytologic pleomorphism, bizarre cellular forms, atypical mitoses, and ulceration (Figure 4).¹⁴ Diagnosis of UPS is by exclusion and is dependent on immunohistochemical studies. In contrast to AFX, UPS is more likely to be positive for LN-2 (CD74).⁶ Undifferentiated pleomorphic sarcoma has been treated with surgical excision in combination with chemical and radiation therapy, but due to limited data, optimal management is less clear compared to AFX.¹⁵ There is a substantial risk for local recurrence and metastasis, and

the lungs are the most common sites of distant metastasis.¹³ In a study of 23 individuals with high-grade UPS, 5-year metastasis-free survival and local recurrence-free survival were 26% and 16%, respectively.¹⁰

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