A 61-year-old woman presented with a 2.5-cm hyperpigmented exophytic nodule on the anterior aspect of the left shin of approximately 2 years’ duration. The patient initially noticed a small lesion following a bee sting, but it subsequently grew over the ensuing 2 years. A shave biopsy was obtained.

**THE BEST DIAGNOSIS IS:**

a. angiosarcoma  
b. blue nevus  
c. dermatofibroma  
d. dermatofibrosarcoma protuberans  
e. sclerotic fibroma  

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Dermatofibroma (DF) is a commonly encountered lesion. Although usually a straightforward clinical diagnosis, histopathological diagnosis is sometimes required. Conventional histologic findings of DF are hyperkeratosis, induction of the epidermis with acanthosis, and basal layer hyperpigmentation. Within the dermis there usually is proliferation of fibroblasts, histiocytes, and blood vessels that sometimes spares the overlying papillary dermis. Nomenclature of specific variants may be assigned based on the predominant component (eg, nodular subepidermal fibrosis, histiocytoma, sclerosing hemangioma) or histologic findings (eg, fibrocollagenous, sclerotic, cellular, histiocytic, lipidized, angiomatous, aneurysmal, clear cell, monster cell, myxoid, keloidal, palisading, osteoclastic, epithelioid). Of the histologic variants, fibrocollagenous is most common, but knowledge of other variants is important for accurate diagnosis, especially to exclude malignancy.

The sclerosing hemangioma variant of DF may present a diagnostic dilemma. In addition to typical features of DF, pseudovascular spaces, abundant hemosiderin, and reactive-appearing spindled cells are histologically demonstrated. The marked sclerosis and pigment deposition may mimic a blue nevus, and the dilated pseudovascular spaces may be reminiscent of a vascular neoplasm such as angiosarcoma or Kaposi sarcoma. However, the presence of characteristic features such as peripheral collagen trapping and overlying epidermal hyperplasia provide important clues for correct diagnosis.

Angiosarcomas (Figure 1) are malignant neoplasms with vascular differentiation. Cutaneous angiosarcomas present as purple plaques or nodules on the head and/or neck in elderly individuals as well as in patients with chronic lymphedema or prior radiation exposure. They are aggressive neoplasms with high rates of recurrence and metastases. Microscopically, the tumor is composed of Anastomosing vascular channels lined by atypical endothelial cells with a multilayered appearance. There is frequent red blood cell extravasation, and substantial hemosiderin deposition may be noted in long-standing lesions. Neoplastic cells are positive for vascular markers (CD34, CD31, ETS-related gene transcription factor). Notably, cases associated with radiation exposure and chronic lymphedema are positive for MYC.

Blue nevi (Figure 2) are benign melanocytic tumors that occur most frequently in children but may present in any age group. Clinical presentation is a blue to black, slightly raised papule that may be found on any site of the body. Biopsy typically shows a wedge-shaped infiltrate of spindled melanocytes with elongated dendritic processes in a sclerotic collagenous stroma. There frequently is a striking population of heavily pigmented melanophages. The melanocytes are positive for melanoma antigen recognized by T cells (MART-1)/melan-A, S-100, and transcription factor SOX-10. In contrast to other benign nevi, human melanoma black-45 will be positive in the dermal component.

Dermatofibrosarcoma protuberans (Figure 3) is a dermal-based tumor of intermediate malignant potential with a high rate of local recurrence and potential for sarcomatous transformation. Dermatofibrosarcoma protuberans most
The differential diagnosis for DF expands once atypical clinical and histopathological findings are present. In this case, the nodule was much larger and darker than the usual appearance of DF (3–10 mm). Given the lesion’s nodularity, the clinical dimple sign on lateral compression could not be seen. On biopsy, the predominance of blood vessels and sclerosis further complicated the diagnostic picture. In unusual cases such as this one, correlation of clinical history, histology, and immunophenotype is ever important.

REFERENCES