

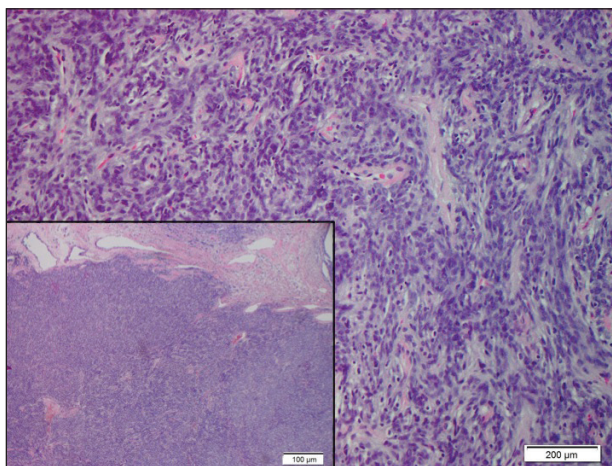
Solitary Tender Nodule on the Back



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H&E, original magnification $\times 200$ (inset, original magnification $\times 40$).

A 73-year-old man presented with a tender nodule on the back that had recently increased in size. On physical examination, a solitary 4-cm nodule was noted in the right trapezius region. The patient denied any personal or family history of similar lesions or a penchant for cysts. Due to the symptomatic nature of the lesion, surgical excision was performed.

THE BEST DIAGNOSIS IS:

- cellular dermatofibroma
- dermatofibrosarcoma protuberans
- nodular fasciitis
- solitary fibrous tumor
- spindle cell lipoma

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THE DIAGNOSIS: Solitary Fibrous Tumor

Solitary fibrous tumors (SFTs), as first described by Klemperer and Rabin¹ in 1931, are relatively uncommon mesenchymal neoplasms that occur primarily in the pleura. This lesion is now known to affect many other extrathoracic sites, such as the liver, kidney, adrenal glands, thyroid, central nervous system, and soft tissue, with rare examples originating from the skin.² Okamura et al³ reported the first known case of cutaneous SFT in 1997, with most of the literature limited to case reports. Erdag et al² described one of the largest case series of primary cutaneous SFTs. These lesions can occur across a wide age range but tend to primarily affect middle-aged adults. Solitary fibrous tumors have been known to have no sex predilection; however, Erdag et al² found a male predominance with a male to female ratio of 4 to 1.

Histopathologically, a cutaneous SFT is known to appear as a well-circumscribed nodular spindle cell proliferation arranged in interlacing fascicles with an abundant hyalinized collagen stroma (quiz image). Alternating hypocellular and hypercellular areas can be seen. Supporting vasculature often is relatively prominent, represented by angulated and branching staghorn blood vessels (Figure 1).² A common histopathologic finding of SFTs is a patternless pattern, which suggests that the tumor can have a variety of morphologic appearances (eg, storiform, fascicular, neural, herringbone growth patterns), making histologic diagnosis difficult (quiz image).⁴ Therefore, immunohistochemistry plays a large role in the diagnosis of this tumor. The most important positive markers include CD34, CD99, B-cell lymphoma 2 (BCL-2), and signal transducer and activator of transcription 6 (STAT6).⁵ Nuclear STAT6 staining is an immunomarker for NGFI-A binding protein 2 (NAB2)-STAT6 gene fusion, which is specific for SFT.^{5,6} Vivero et al⁷ also reported glutamate receptor, inotropic, AMPA 2 (GRIA2) as a useful immunostain in SFT, though it is also expressed in dermatofibrosarcoma

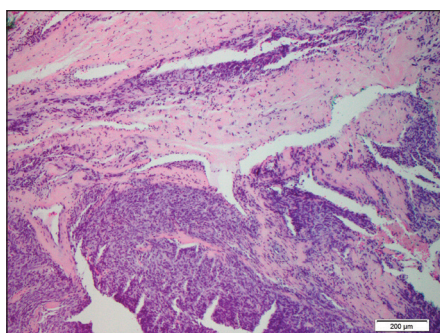


FIGURE 1. Angulated and branching staghorn vessels in a solitary fibrous tumor (H&E, original magnification $\times 100$).

protuberans (DFSP). In this case, the clinical and histopathologic findings best supported a diagnosis of SFT. Some consider hemangiopericytomas to be examples of SFTs; however, true hemangiopericytomas lack the thick hyalinized collagen and hypercellular areas seen in SFT.

A cellular dermatofibroma generally presents as a single round, reddish brown papule or nodule approximately 0.5 to 1 cm in diameter that is firm to palpation with a central depression or dimple created over the lesion from the lateral pressure. Cellular dermatofibromas mostly occur in middle-aged adults, with the most common locations on the legs and on the sides of the trunk. They are thought to arise after injuries to the skin. On histopathologic examination, cellular dermatofibromas typically exhibit a proliferation of fibrohistiocytic cells with collagen trapping, often at the periphery of the tumor (Figure 2). Although cellular dermatofibromas appear clinically different than SFTs, they often mimic SFTs histopathologically. Immunostaining also can be helpful in differentiating

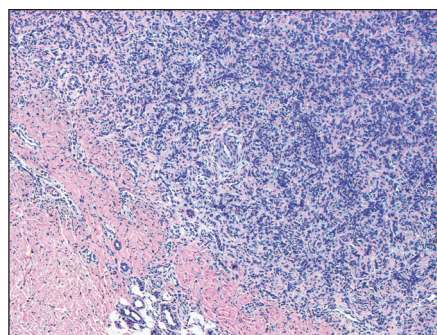


FIGURE 2. Cellular dermatofibroma demonstrating a proliferation of fibrohistiocytic cells with collagen trapping at the periphery of the tumor (H&E, original magnification $\times 100$).

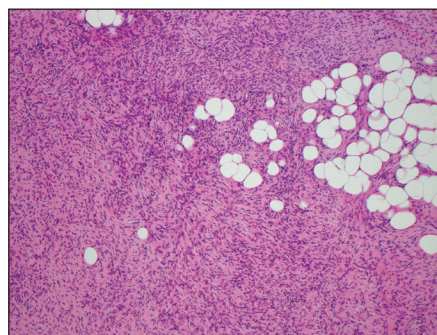


FIGURE 3. Dermatofibrosarcoma protuberans demonstrating a dense, hypercellular, spindle cell proliferation in a storiform pattern with adipocyte entrapment (H&E, original magnification $\times 100$).

cellular dermatofibromas in which cells stain positive for factor XIIIa. CD34 staining is negative.

Dermatofibrosarcoma protuberans usually appears as one or multiple firm, red to violaceous nodules or plaques. They most often occur on the trunk in middle-aged adults. Histopathologically, DFSP presents with a dense, hypercellular, spindle cell proliferation that demonstrates a typical storiform pattern. The tumor generally infiltrates into the deep dermis and subcutaneous adipose layer with characteristic adipocyte entrapment (Figure 3). Positive CD34 and negative factor XIIIa staining helps to differentiate DFSP from a cellular dermatofibroma. Immunohistochemically, it is more difficult to distinguish DFSP from SFT, as both are CD34⁺ spindle cell neoplasms that also stain positive for CD99 and BCL-2.² GRIA2 positivity also is seen in both SFT and DFSP.⁷ However, differentiation can be made on morphologic grounds alone, as DFSP has ill-defined tumor borders with adnexal and fat entrapment and SFT tends to be more circumscribed with prominent arborizing hyalinized vessels.⁸

Spindle cell lipoma (SCL) is an asymptomatic subcutaneous tumor commonly located on the back, neck, and shoulders in older patients, typically men. It often presents as a solitary lesion, though multiple lesions may occur. It is a well-circumscribed tumor of mature adipose tissue with areas of spindle cell proliferation and ropey collagen bundles (Figure 4). In early lesions, the spindle cell areas are myxoid with the presence of many mast cells.⁹ The spindle cells stain positive for CD34. Although spindle cell lipoma would be included in both the clinical and histopathologic differential diagnosis for SFT, its histopathologic features often are enough to differentiate SCL, which is highlighted by the aforementioned features as well as a relatively low cellularity and lack of ectatic vessels.⁸ However, discerning tumor variants, such as low-fat pseudoangiomatous SCL and lipomatous or myxoid SFT, might prove more challenging.

Nodular fasciitis typically presents as a rapidly growing subcutaneous nodule that may be tender. It is a benign reactive process usually affecting the arms and trunk of young to middle-aged adults, though it commonly involves the head and neck region in children.¹⁰ The tumor histopathologically appears as a well-circumscribed subcutaneous or

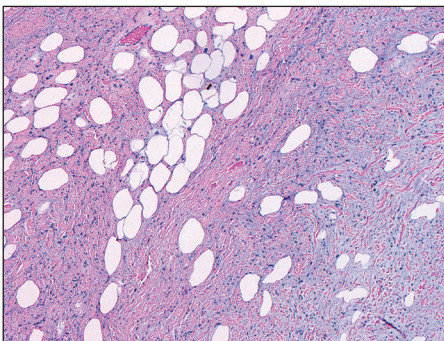


FIGURE 4. Spindle cell lipoma showing a spindle cell proliferation and ropey collagen bundles in a myxoid stroma (H&E, original magnification $\times 100$).

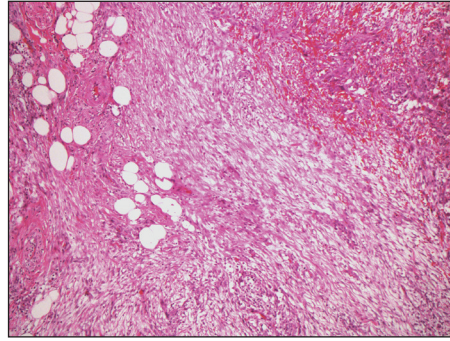


FIGURE 5. Nodular fasciitis demonstrating spindle-shaped and stellate fibroblasts loosely arranged in an edematous myxomatous stroma with the presence of extravasated erythrocytes (H&E, original magnification $\times 100$).

fascial nodule with an angulated appearance. Spindle-shaped and stellate fibroblasts are loosely arranged in an edematous myxomatous stroma with a feathered appearance (Figure 5). Extravasated erythrocytes often are present. With time, collagen bundles become thicker and hyalinized. Immunohistochemical studies demonstrate positivity for vimentin, calponin, muscle-specific actin, and smooth muscle actin. Desmin, CD34, cytokeratin, and S-100 typically are negative.¹⁰⁻¹² Therefore, CD34 staining is one of the main differentiating factors between nodular fasciitis and SFTs.

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