

## What Is Your Diagnosis?

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A 9-year-old girl presented with a 6×6-mm asymptomatic flat papule on her left medial forearm of 2 years' duration.

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## The Diagnosis: Desmoplastic Spitz Nevus

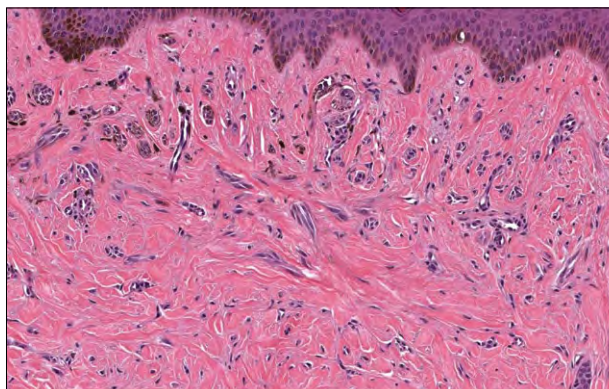
**D**esmoplastic Spitz nevus is a variant of the benign melanocytic Spitz nevus and also is called spindle cell and epithelioid cell nevus. It contains a prominent fibrotic stroma composed of thick collagen bundles.<sup>1</sup> The term *desmoplastic nevus* has been used synonymously by some authors<sup>2</sup>; however, others maintain it as a distinct entity.<sup>3</sup> Hyalinizing Spitz nevus, composed of epithelioid and spindle cells embedded in a paucicellular hyalinized stroma, and angiomatoid Spitz nevus, which contains melanocytes in a fibrotic stroma with numerous small blood vessels, are believed to be variants of desmoplastic Spitz nevus by some authors.<sup>4,5</sup>

Desmoplastic Spitz nevus clinically presents as a red or red-brown, smooth, firm, flat or dome-shaped papule, often on the trunk or extremities, that is present for months to years (Figure 1). It has been reported in patients as young as 8 years but is more common in patients older than 20 years.<sup>1,3,6</sup> It may be mistaken for dermatofibroma, Spitz nevus, atypical nevus, melanoma, mastocytoma, and keloid.

Histologically, desmoplastic Spitz nevus is composed of large spindle and epithelioid cells distributed singly or as small clusters in an abundant fibrotic dermal stroma of thick collagen bundles (Figure 2). Some nuclear atypia may be present, but mitotic figures are rare and never abnormal. Intranuclear invaginations of cytoplasm as well as giant cells may be seen. There may be a lymphocytic inflammatory infiltrate.



**Figure 1.** A 6×6-mm burgundy-colored, flat papule with darker macules scattered within its boundaries on the left medial forearm.



**Figure 2.** Spindle and epithelioid cells distributed singly and as small clusters in an abundant fibrotic dermal stroma (H&E, original magnification ×200).

Epidermal hyperplasia and occasionally hyperpigmentation may be present.<sup>1,3,6</sup> Desmoplastic Spitz nevus stains positively with S-100 protein. It also often is positive for HMB-45 (human melanoma black), but its staining decreases with depth, indicating maturation of the melanocytes. Factor XIIIa also may be positive. Ki-67 may be expressed in a small percentage of cells.<sup>3,6</sup> The histologic differential diagnosis includes desmoplastic malignant melanoma, dermatofibroma, hypopigmented blue nevus, neurofibroma, and schwannoma.

Desmoplastic Spitz nevus follows a benign clinical course and does not recur after complete excision.<sup>1</sup>

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