Fibrohistiocytic Tumors

Ryan C. Romano, DO[a], Karen J. Fritchie, MD[b,*]

KEYWORDS

• Fibrohistiocytic • Histiocytes • Fibrous histiocytoma • Xanthogranuloma
• Dermatofibrosarcoma protuberans

KEY POINTS

- Fibrohistiocytic tumors, some of which are common and others of which are rarely encountered, present significant challenges to the dermatopathologist owing to clinical and histopathologic overlap.
- The "fibrohistiocytic" designation does not necessarily denote lineage differentiation in all cases but rather may be used for lesions in which tumor cells resemble histiocytes and/or fibroblasts.
- Most fibrohistiocytic lesions are best diagnosed by careful histologic examination; immunohistochemistry and molecular studies play a limited role in the workup of this family of tumors.

OVERVIEW

Fibrohistiocytic tumors represent a diverse group of mesenchymal neoplasms with widely variable presentations. The "fibrohistiocytic" designation does not necessarily denote lineage differentiation in all cases, but rather may also be used for lesions in which tumor cells resemble histiocytes and/or fibroblasts. These lesions exhibit a range of clinical behavior ranging from reactive (xanthoma) to benign (fibrous histiocytoma and its variants) to tumors with significant local recurrence potential (dermatofibrosarcoma protuberans). This review highlights key morphologic features of these entities, as well as potential diagnostic pitfalls and histologic mimics. Although immunohistochemistry may be helpful in excluding other diagnoses, immunostains play a relatively limited role in the workup of the majority of lesions in this family. A subset of fibrohistiocytic neoplasms, including dermatofibrosarcoma protuberans and angiomatoid fibrous histiocytoma, have unique genetic aberrations, and the use of cytogenetic/molecular studies in these tumors as an adjunct to microscopic examination is discussed.

The authors have nothing to disclose.
[a] Department of Dermatology, Mayo Clinic, Gonda 16, 200 First Street Southwest, Rochester, MN 55905, USA; [b] Division of Anatomic Pathology, Department of Laboratory Medicine and Pathology, Mayo Clinic College of Medicine, Mayo Clinic, Hilton 11, 200 First Street Southwest, Rochester, MN 55905, USA
* Corresponding author.
E-mail address: fritchie.karen@mayo.edu

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FIBROUS HISTIOCYTOMA AND VARIANTS

Epidemiology and Clinical Features

Fibrous histiocytoma (dermatofibroma) is typically diagnosed in young to middle-aged adults with a female predominance and shows a strong predilection for involving the extremities.[1,2] Most patients present with a single, asymptomatic, long-standing lesion, although multiplicity has been associated with immune suppression.[2,3] The aneurysmal variant may exhibit the clinical appearance of rapid growth secondary to intratumoral hemorrhage.[4] There has been debate as to whether fibrous histiocytoma represents a reactive, inflammatory process or a clonal neoplasm, but the prevailing opinion favors a neoplastic process.[5–7]

Gross Features

The majority of lesions measure less than 2.0 cm and are well-circumscribed (**Fig. 1**A). Cystic change or hemorrhage may be seen.

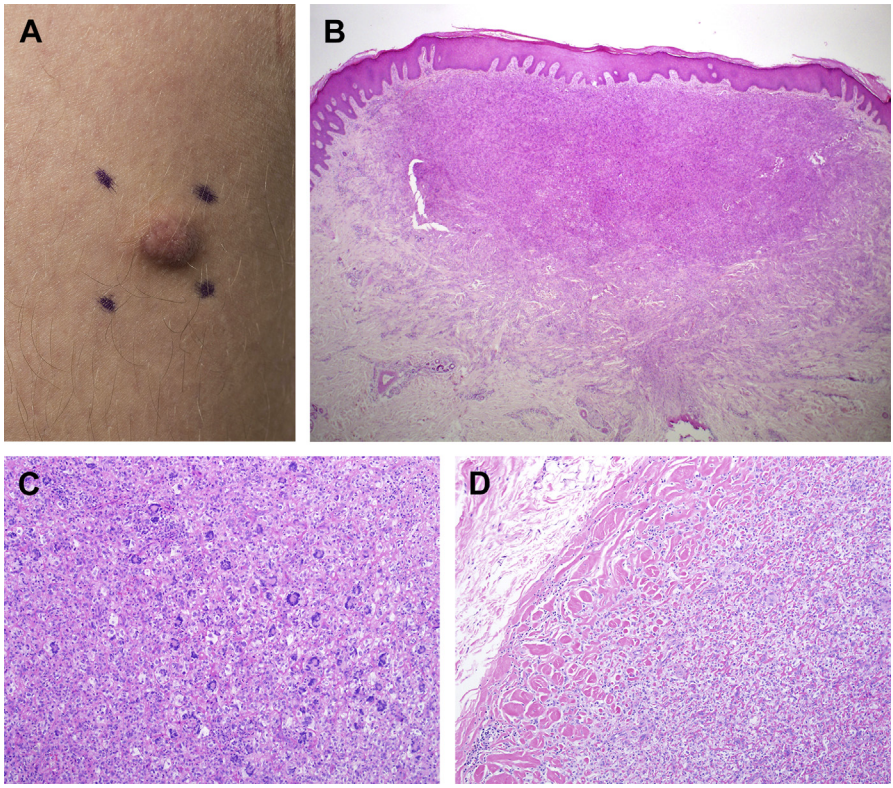

Fig. 1. (*A*) Clinically, fibrous histiocytoma appears as a well-demarcated, slightly raised flesh-colored to tan-pink papule. Low-power examination (*B*, H&E, original magnification ×40) shows a circumscribed, dermal-based lesion with a pushing border and lack of significant extension into the subcutis. Closer inspection (*C*, H&E, original magnification ×100) reveals a polymorphous proliferation of multinucleated giant cells, foamy histocytes, and bland mononuclear cells. The peripheral border shows prominent collagen entrapment (*D*, H&E, original magnification ×100).