The New Kids on the Block
Recently Characterized Soft Tissue Tumors

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KEYWORDS
• Soft tissue pathology • Sarcoma • Molecular • Liposarcoma • ALK

ABSTRACT

Soft tissue pathology is a rapidly changing subspecialty. New entities are described relatively often, and new molecular findings for soft tissue tumors are reported in the literature almost every month. This article summarizes the major features and diagnostic approach to several recently characterized entities: superficial CD34-positive fibroblastic tumor, fibrosarcoma-like lipomatous neoplasm, angiofibroma of soft tissue, low-grade sinonasal sarcoma with neural and myogenic features, malignant gastrointestinal neuroectodermal tumor, hemosiderotic fibrolipomatous tumor, and epithelioid inflammatory myofibroblastic sarcoma. Additionally, the article also provides a summary table of recent molecular findings in soft tissue tumors.

OVERVIEW

Soft tissue pathology is a rapidly changing subspecialty. Novel molecular findings are published monthly, rivaling even hematopathology in the speed with which the genetic underpinnings of neoplasia are probed. Tumors that were previously known under one rubric are reclassified with relative frequency. Lesions that for decades were thought to be reactive now are discovered to possess gene rearrangements (e.g. – nodular fasciitis), and the features of previously unknown or incompletely described tumors are coalesced and synthesized into new entities. The relative rarity of soft tissue tumors only adds to the challenge of keeping abreast of all of these advances. In this article, we attempt to highlight some of the more pertinent soft tissue tumors that have been recently characterized and to make them more accessible to the general surgical pathologist. We also provide a brief summary of significant molecular findings, not only in these newly described tumors, but also in older entities for which molecular abnormalities have only recently been elucidated.

SUPERFICIAL CD34-POSITIVE FIBROBLASTIC TUMOR

Carter and colleagues[1] characterized superficial CD34-positive fibroblastic tumor in 2013. They described 18 cases of a tumor with unique characteristics: striking nuclear pleomorphism, paradoxically rare mitotic activity, indolent behavior, and diffuse CD34 expression. All cases occurred in adults and all presented as a slow-growing supra-fascial mass of 10 cm or less (mean: 4.1 cm), most commonly in the lower extremity. Awareness of this entity is important, as there is a tendency for overdiagnosis as a pleomorphic sarcoma in light of the nuclear atypia, even though most cases do not recur or metastasize. Of 13 patients with follow-up (median: 24 months), there were no local recurrences, no distant metastases, and no death from disease; 1 patient had a regional lymph node metastasis 7 years after presentation. In light of this, Carter and colleagues[1] suggested this entity be considered as a lesion of borderline malignancy.

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PATHOLOGIC FEATURES

Superficial CD34-positive fibroblastic tumor is a subcutaneous mass with little or no involvement of the deep musculature. It is often relatively circumscribed, although it may show peripheral infiltrative growth. Most tumor cells display marked nuclear pleomorphism with hyperchromasia and multiple large inclusion-like nucleoli as well as cytoplasmic nuclear pseudoinclusions. The cells are spindled to epithelioid with abundant eosinophilic often granular cytoplasm and are arranged into hypercellular sheets or fascicles (**Fig. 1**A, B). Xanthomatous foamy tumor cells are commonly seen, and mixed inflammation is often present. Small vessels are arranged in a plexiform pattern in the background of the tumor, but large ectatic hyalinized vessels, like those of pleomorphic hyalinizing angiectactic tumor (PHAT), are not seen. Despite the extreme degree of nuclear atypia, mitotic figures

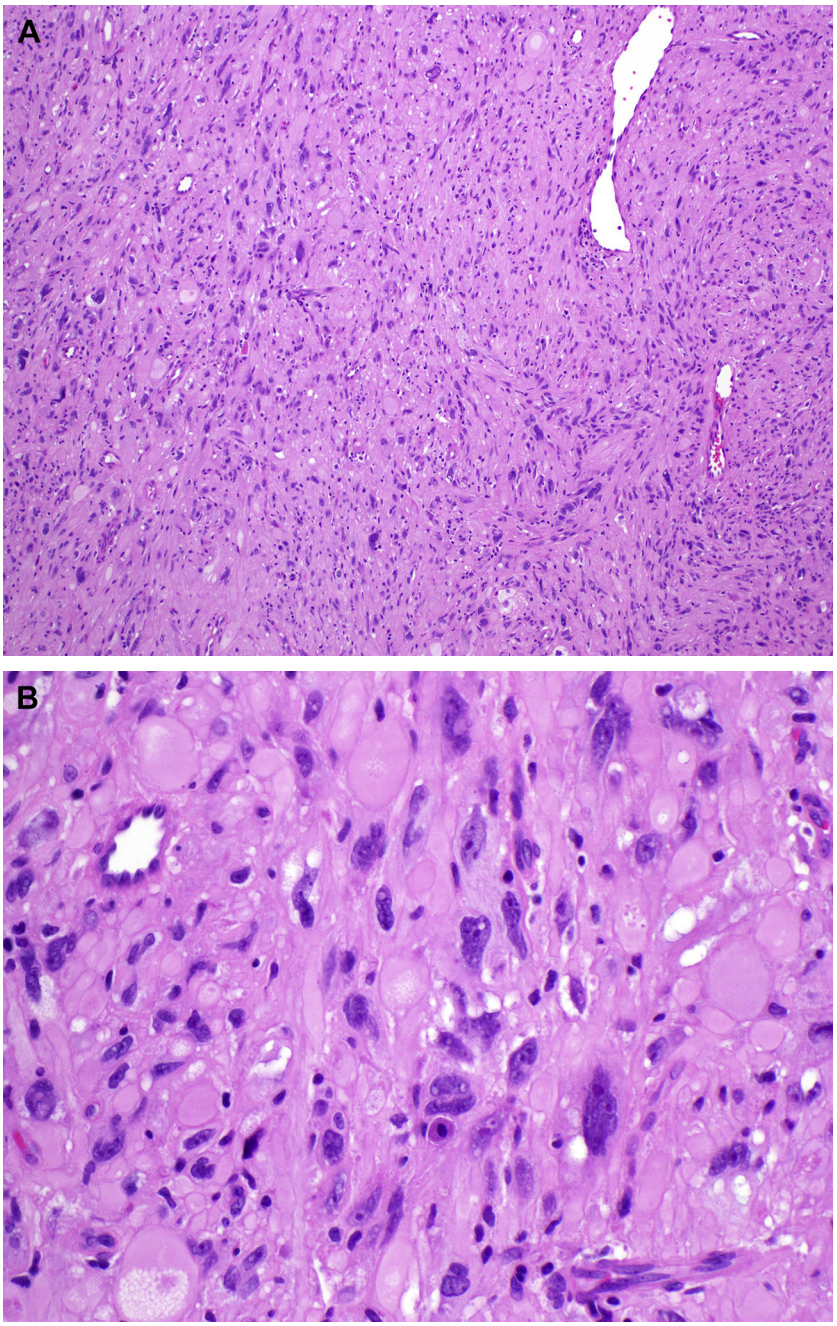

Fig. 1. (*A*) Superficial CD34-positive fibroblastic tumor is composed of sheets of pleomorphic spindle cells. (*B*) Superficial CD34-positive fibroblastic tumor: there is marked nuclear pleomorphism but mitoses are rare. Tumor cells display abundant eosinophilic cytoplasm.