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Inflammatory Myofibroblastic Tumor

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Inflammatory myofibroblastic tumor (IMT) is a distinctive, rarely metastasizing, (myo)fibroblastic neoplasm composed of spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and/or eosinophils. Genetically, most IMTs harbor tyrosine kinase fusions, usually the *ALK* gene with an ever-expanding list of partner genes (*TIMP3-ALK* is most common), although these rearrangements are rare in adults. More common in abdominal soft tissue and lung sites, head and neck tumors comprise about 20% of all IMTs. Children and young adults are usually affected, with a slight female predominance. Symptoms are nonspecific and generally related to obstructive symptoms due to mass effect. Patients are generally managed surgically, although molecular-targeted therapy (eg, crizotinib) shows good response. While metastatic disease is exceptional, local recurrences are seen in about 25% of patients.

Tumors are usually polypoid, fleshy or myxoid lesions, usually small, especially due to laryngeal confines. The spindled myofibroblastic cells are slightly plump to ganglionic in appearance (Figure 1), set within a myxoid, loose background stroma, rich with inflammatory cells. Cells have vesicular, open nuclear chromatin, with nuclei that have up to several small nucleoli. Sometimes, the tumors are more cellular with compact fascicles of spindled cells with a more collagenized stroma. A hypocellular proliferation is less common. Mitoses are present but generally not increased. Dystrophic calcifications may be present. The majority of cases show a strong ALK immunoreactivity, with the pattern of reactivity dependent on the fusion partner (diffuse cytoplasmic [Figure 2], perinuclear, granular cytoplasmic, nuclear membranous), while various muscle markers usually show a patchy, weak to stronger reaction; focal keratin reactivity may be seen in about 30% of cases.

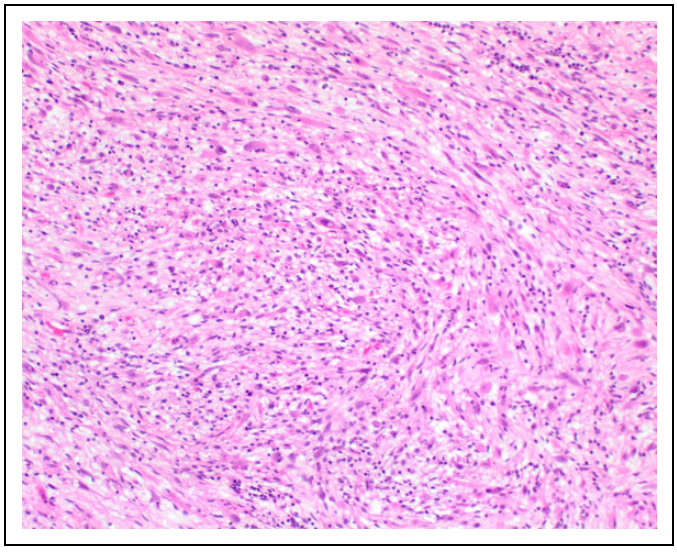

Figure 1. There is a storiform to fascicular spindled cell proliferation, focally with ganglion-like cells, set within an inflammatory cell background.

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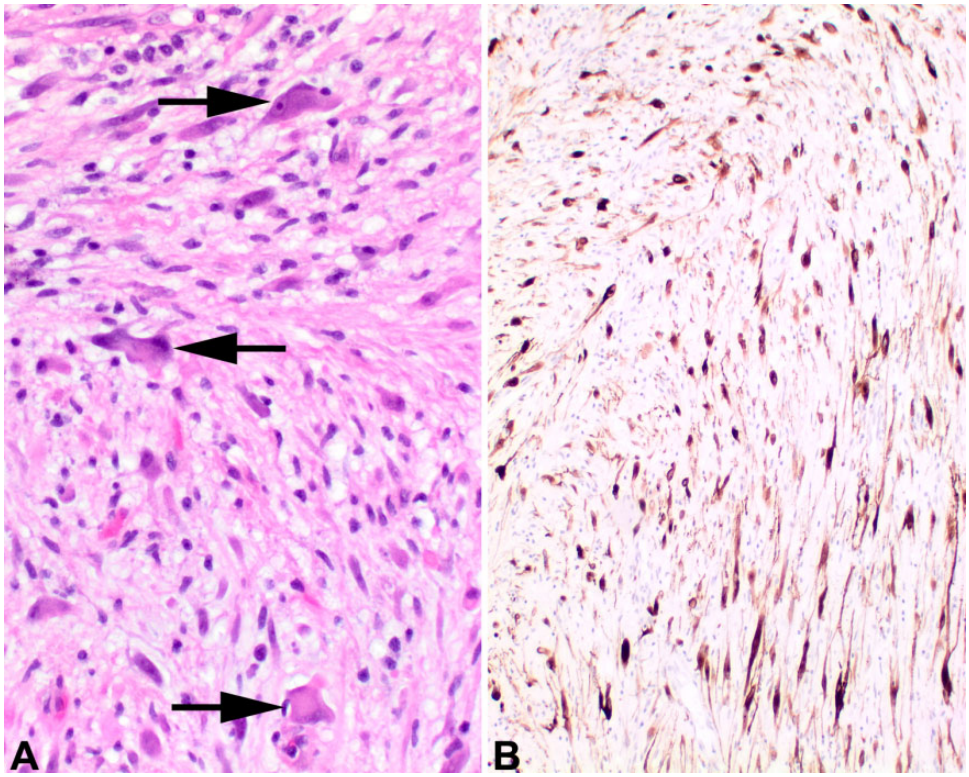

Figure 2. A, Well-developed ganglion-like cells (black arrows) are seen in this inflammatory myofibroblastic tumor. B, Nearly all of the neoplastic cells show a strong cytoplasmic reaction with ALK immunohistochemistry.

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Suggested Reading

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