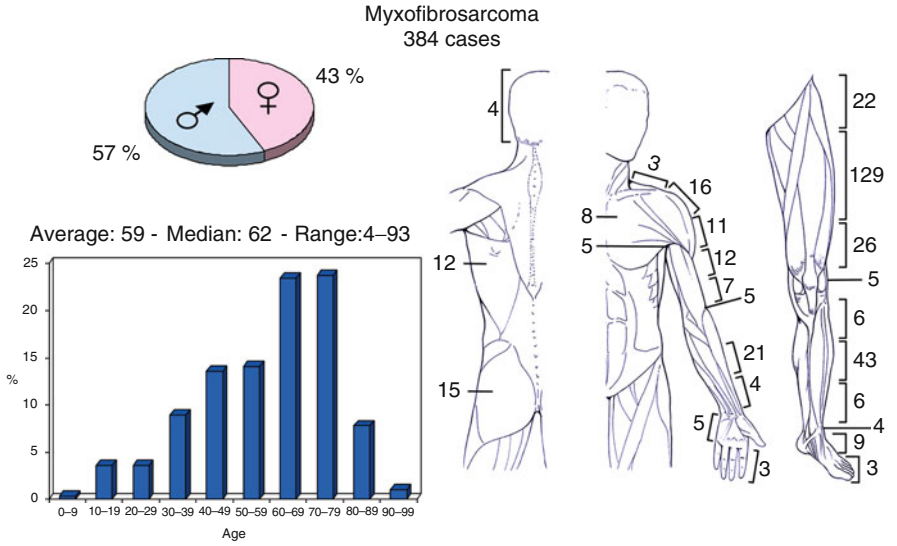
Chapter 75
Myxofibrosarcoma

Marco Gambarotti

Definition: Malignant fibroblastic lesions with myxoid stroma, pleomorphism, and a distinctive curvilinear vascular pattern. The designation of myxofibrosarcomas and myxoid malignant fibrous histiocytoma has been considered almost synonymous in the 2013 WHO classification of soft tissue tumors. This classification does not mention a minimal amount of myxoid matrix for the definition of myxofibrosarcoma.
Epidemiology: Among the most common sarcomas in elderly patients. Overall age range is wide, but they are more frequent in the sixth to eighth decade, whereas they are exceptional under 20 years of age. There is a slight male predominance.



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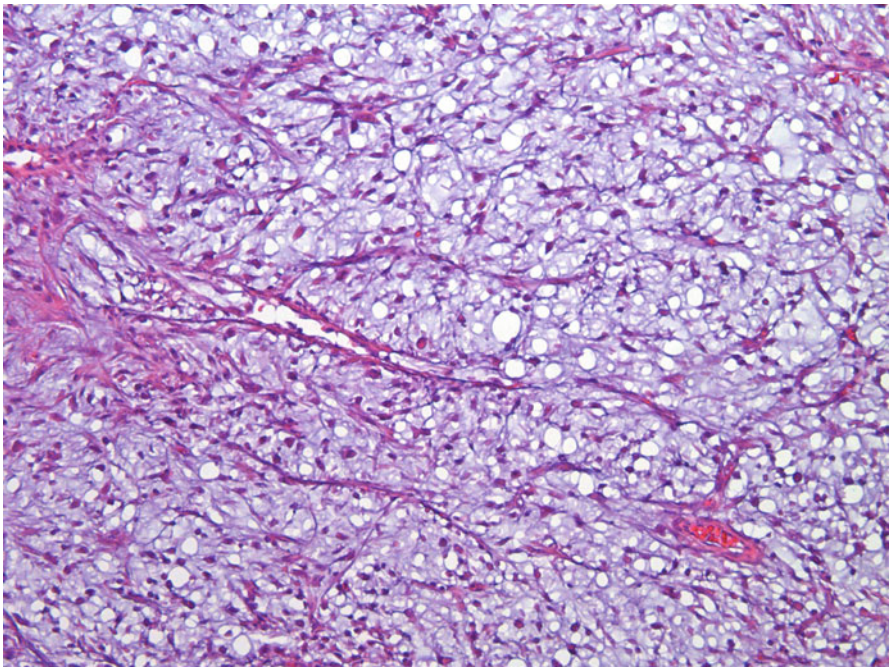
Location: The most frequent sites are the lower limbs, followed by upper limbs and limb girdles. Rare in trunk, head and neck, and hands and feet. Most cases described in the retroperitoneum and in the abdominal cavity represent dedifferentiated liposarcomas. About half of cases arise in dermal/subcutaneous tissue, with the remaining arising in the underlying fascia and skeletal muscle.
Clinical and Imaging: Enlarging painless mass, with infiltrative margins. Very often heterogeneous at MRI.
Histopathology: Grossly, they appear as multiple gelatinous to firm nodules with infiltrative margins. Histologically, all cases share distinct morphological features: multinodular growth with incomplete fibrous septa, a myxoid stroma composed of hyaluronic acid, and prominent elongated, curvilinear, thin-walled vessels with a perivascular condensation of tumor cells. Frequently, the so-called pseudolipoblasts (vacuolated neoplastic fibroblastic cells with cytoplasmic acid mucin) are seen. Low-grade lesions are hypocellular, composed of few non-cohesive, plump spindled or stellate cells with ill-defined cytoplasm and hyperchromatic nuclei; mitosis is infrequent. High-grade lesions are composed in large part of solid sheets and cellular fascicles of spindle and pleomorphic tumor cells with numerous, often atypical, mitoses and areas of necrosis. Immunohistochemically, tumor cells stain positive for vimentin.
Course and Staging: In up to 50–60 % of cases, local recurrences unrelated to histological grade repeatedly occur. In contrast, metastases and death from tumor are closely related to tumor grade: low-grade tumors do not metastasize, while metastases develop in 20–35 % of intermediate- and high-grade neoplasms. Metastases occur in the lung, bone, and lymph nodes. Low-grade lesions that recur may subsequently increase in grade. The depth of the lesion does not influence the rate of local recurrence, while deep-seated neoplasms have a higher percentage of metastases and tumor-associated mortality. Overall 5-year survival rate is 60–70 %.
Treatment: Excision with wide margins and adjuvant radiation therapy and/or systemic chemotherapy.

Immunohistochemical Panel

VIM	+



Bland-appearing spindle cells in a loose myxoid extracellular matrix. Cells are arranged in lobules, with elongated curvilinear blood vessels

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