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Case Presentation

Myxofibrosarcoma presenting as an exophytic, multi-lobed nodule on the leg

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Abstract

Myxofibrosarcoma may present as a dermal or subcutaneous nodule, often on the extremity of an elderly patient. We present a case of myxofibrosarcoma on the lower leg of a 77-year-old man, which illustrates the deeply infiltrative growth pattern of these tumors, as well as the potential for superficial biopsies to show lower grade histopathologic features than subsequent excision specimens.

Keywords: myxofibrosarcoma, myxoid malignant fibrous histiocytoma,

Introduction

Myxofibrosarcoma (MFS) (formerly myxoid malignant fibrous histiocytoma) is among the most common sarcomas of the elderly, occurring most often on the lower extremities [1]. It is notable for its high rate of local recurrence and occasional metastasis. Myxofibrosarcoma often grows in an infiltrative pattern within the subcutis and fascia and may extend into the overlying dermis to present as a cutaneous lesion. These tumors represent a clinical and histopathological diagnostic challenge, as the malignant nature of the lesion may not be initially apparent [2-4]. We present a case in which the initial shave biopsy showed a superficial, low grade lesion, but subsequent excision revealed a higher grade tumor with extensive infiltration of the subcutis.

Case synopsis

A 77-year-old otherwise healthy man presented with a five month history of a progressively enlarging, asymptomatic mass of the right lower leg. Physical examination showed a pink to red 2.5cm x1.5cm compressible, exophytic, lobulated nodule of the right medial leg with an adjoining 4 cm erythematous plaque with several superimposed 0.5-1cm pink to translucent soft papules (Figure 1). The entire tumor was approximately 7 cm x 4cm.

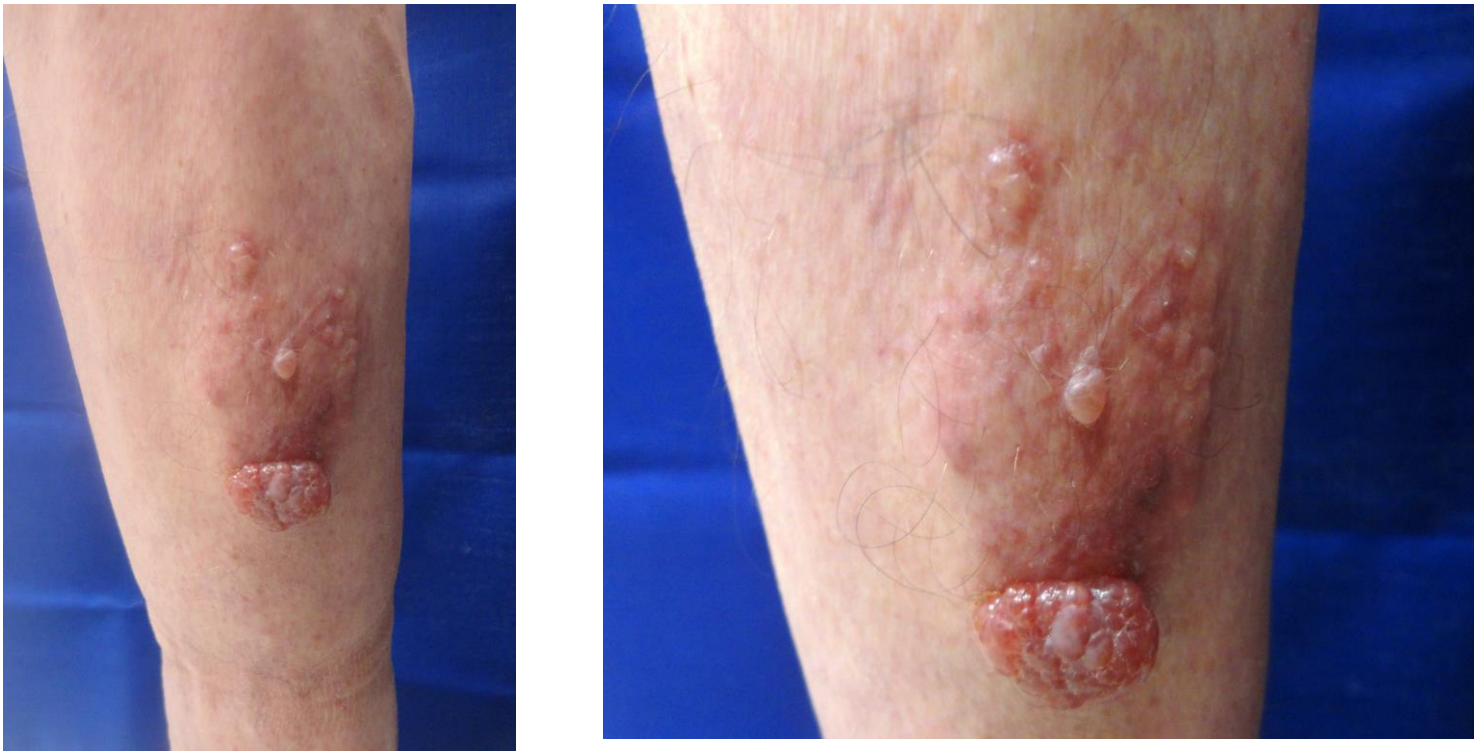

Figure 1. (A) Red exophytic, lobulated nodule of the right medial leg (B) Closer view of right medial leg lesion

A shave biopsy from the exophytic portion showed a dermal proliferation of widely spaced spindle and multinucleate cells within a myxoid stroma (Figure 2). The tumor cells showed mild to moderate pleomorphism, with rare mitotic figures. The tumor cells were negative for S100 by immunohistochemistry and the findings were interpreted as low grade myxofibrosarcoma.

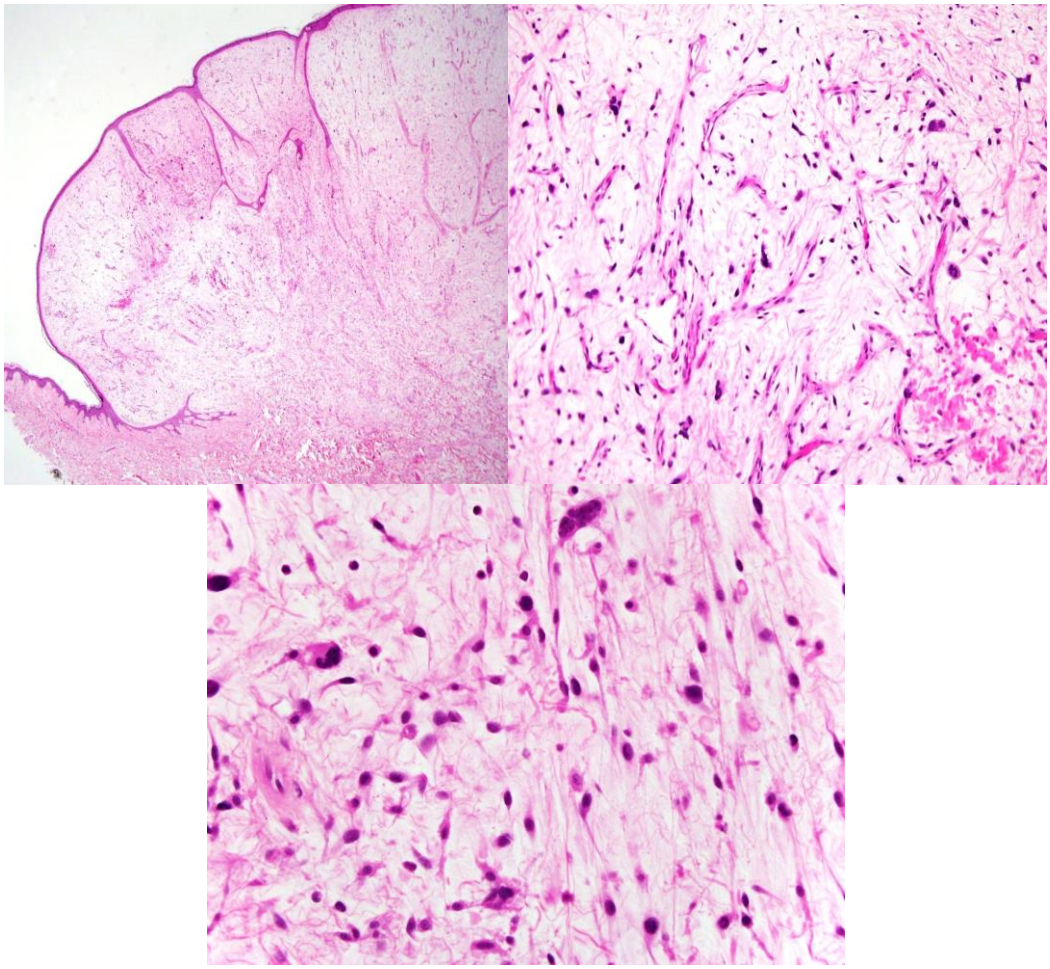

Figure 2. (A) Initial biopsy showing an exophytic dermal neoplasm (hematoxylin and eosin, x20) (B) Widely spaced spindle and multinucleate cells in a myxoid stroma (H&E, x100) (C) Higher magnification of pleomorphic cells (H&E, x400)

The patient was referred to the plastic surgery department and excision with 2 cm margins was performed. Histologically, the excision specimen showed some areas similar in appearance to the biopsy specimen. However, there were also broad areas of subcutaneous extension, with increased cellularity, frequent mitotic figures, and prominent nuclear atypia (Figure 3), consistent with an intermediate to high grade myxofibrosarcoma. Although wide surgical margins were taken, the lesion extended to within 1 mm of the margin histologically. He completed adjuvant radiation and has been followed for 1 year with clinical examinations and imaging studies, without evidence of local recurrence or metastasis.

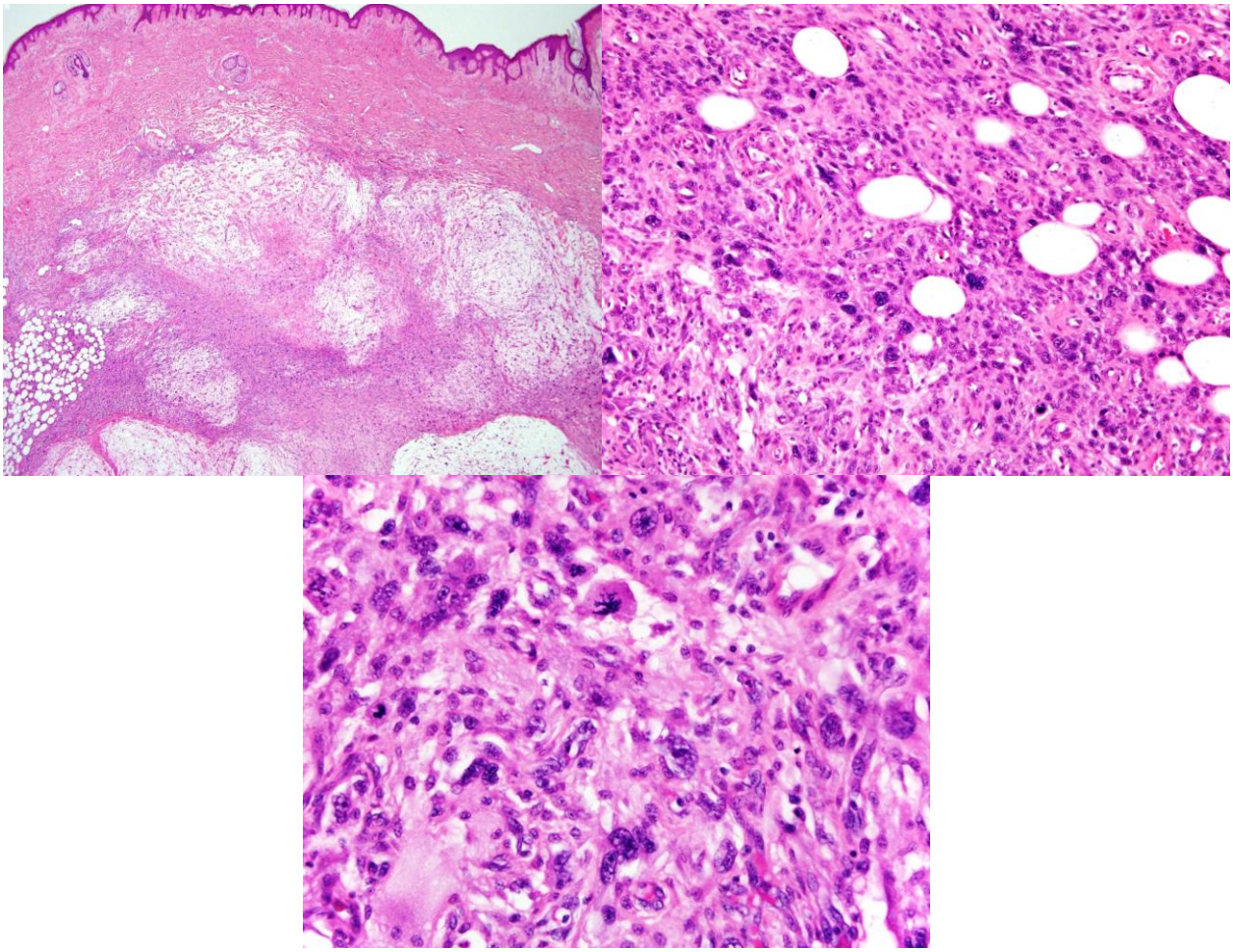

Figure 3. (A) Excision specimen showing extension of tumor into the subcutis (H&E, x20) (B) Areas of solid tumor within the subcutis (H&E, x100) (C) Prominent nuclear pleomorphism and mitotic activity (H&E, x400)

Discussion

Myxofibrosarcoma typically presents in adults over 50 years of age and most commonly arises on the lower extremity, followed by the upper extremity, trunk, and head and neck [1, 5]. The clinical appearance is nonspecific, and given its rarity, it may be clinically mistaken for more common processes such as lipoma, cyst, or panniculitis [4]. Potential diagnostic clues include a multilobular architecture and large size, with reported median diameters of 5-9.9 cm [1, 6]. Although some lesions presenting in the skin may appear to be relatively well-circumscribed, there is often wide subclinical extension within the underlying soft tissue [7]. The 'tail sign' of MFSs on magnetic resonance imaging studies has recently been shown to be a moderately specific (79-90%) and sensitive (64-77%) sign that can be used to help differentiate myxofibrosarcoma from other myxoid tumors [8].

Histologically, low grade MFSs are hypocellular neoplasms with spindled and stellate cells separated by an abundant myxoid stroma, with many elongated, curvilinear, thin-walled blood vessels. Mitotic figures are infrequent; however, enlarged, hyperchromatic nuclei may be observed. High grade lesions contain sheets and fascicles of variably spindled cells with prominent nuclear pleomorphism, frequent mitotic figures, and areas of necrosis [4,9]. As observed in our patient, many tumors have a higher grade histomorphology within the subcutaneous soft tissue and predominantly low grade areas within the dermis [4]. As a result, the histologic findings in a superficial biopsy may not be representative of the overall process and may even be misinterpreted as a benign process such as angiomyxoma, cutaneous mucinosis, myxoid neurofibroma, or granulomatous dermatosis [3-4,7,10], emphasizing the need for biopsies of sufficient size and depth, along with correlation of clinical and histologic findings.

Current treatment recommendations for myxofibrosarcoma include wide excision and adjuvant radiation [5]. Owing to infiltrative growth extending beyond the clinically evident tumor borders, positive histologic margins are common, occurring in 13.3-34% of cases [1, 5, 6, 11]. Local recurrence is likewise common, with reported rates of 15-54% [1, 5, 6, 12]. Metastases are found in 3% of patients at the time of diagnosis and an additional 20% thereafter [6], with 5-year overall survival rates of 63-77% [1, 6]. Increasing tumor size, histologic grade, and tumor attachment to bone have been suggested as negative prognostic indicators, although studies are not uniform in this regard [1, 6, 11].

Conclusions

In conclusion, myxofibrosarcoma may present as a cutaneous or subcutaneous nodule, often on the extremity of an elderly patient. Dermatologists and dermatopathologists should be familiar with the presentation of myxofibrosarcoma, including its propensity to extend beyond the clinically evident margin and to appear histopathologically less aggressive in superficial biopsies than in deeper excisional specimens.

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