Myxofibrosarcoma - Case report*

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Abstract: Myxofibrosarcoma is recognized as a malignant neoplasm of fibroblastic origin with increased prevalence in the elderly, presenting as nodules or tumors that may extend to the dermis and skeletal muscle, preferably in the lower limbs. Histologically it is characterized by a proliferation of spindle cells in a myxoid stroma. Myxofibrosarcoma has a high potential for local recurrence and metastasis, mainly when it presents a high or intermediate histological grade. We report the case of an eighty-four year old patient with a difficult diagnosis of a highly aggressive tumor.
Keywords: Aged; Myxosarcoma; Neoplasm metastasis; Neoplasm recurrence, local; Skin neoplasms

INTRODUCTION

Myxofibrosarcoma, previously known as myxoid malignant fibrous histiocytoma, is considered one of the most common fibroblastic sarcomas of the elderly, predominantly affecting patients between sixty and eighty years of age. [1,2,3]

We report the case of an intermediate grade myxofibrosarcoma surgically resected, but with later recurrence and metastasis to local lymph nodes.

CASE REPORT

An 84-year-old white man presented to our division with a three month-history of an erythematous, painful, rapidly growing tumor in the right lower limb.

Dermatological examination revealed a 3.2 cm x 2.8 cm hyperkeratotic ulcerated tumor in the posterolateral view of the right lower limb (Figure 1). A skin biopsy was performed and the histopathology showed proliferation of atypical spindle cells with irregular and hyperchromatic nuclei

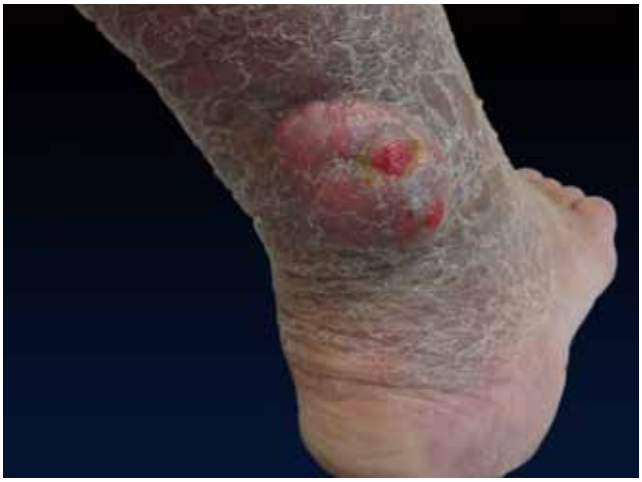

Figure 1: Hyperkeratotic erythematous tumor with areas of ulceration in right lower limb

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arranged in bundles located in the papillary and reticular dermis. The immunohistochemistry showed negative S-100, cytokeratins, ERG, p-63 and desmin markers. Only CD34 was positive. Based on the immunohistochemistry and on the undifferentiated histopathological pattern, the lesion was classified as an undifferentiated pleomorphic sarcoma.

The patient was referred to the oncologist for tumor staging. Imaging tests showed that the tumor was located above the muscle fascia; no lymph node or distant metastasis were detected (Figure 2). The surgical resection was performed with a 2 cm margin and the histopathology showed proliferation of ovoid or spindle cells arranged in bundles, with irregular, hyperchromatic nuclei and mitotic figures, combined with myxoid stroma (Figure 3). The presence of elongated, curvilinear, thin-walled blood vessels was also noticed (Figure 4). It was then reclassified as myxofibrosarcoma.

The initially proposed treatment was radiotherapy. However, two months later the patient presented local recurrence of the tumor and a right inguinal nodal metastasis. The treatment was then changed to palliative chemotherapy with doxorubicin.

DISCUSSION

Myxofibrosarcoma is considered the most common malignant mesenchymal neoplasm in elderly patients, with slight male predominance. It usually presents as painless, slow growing, skin colored or erythematous nodules or tumors. Most lesions are located in the lower limbs and rarely on the trunk, head and neck.[4] About two-thirds of the cases develop in the dermal and subcutaneous tissues, and one third in the fascia and skeletal muscle.[1, 2]

The diagnosis of myxofibrosarcoma is hystopathological and tumors are classified as low, intermediate and high grade.[2] The first is characterized by hypocellularity, cells with hyperchromatic and pleomorphic nuclei associated with extensive myxoid areas. Intermediate grade tumors are more cellular with frequent nuclear atypia. Finally, high-grade tumors are densely cellular, pleomorphic, with numerous mitotic figures, areas of hemorrhage and necrosis. Few areas of myxoid stroma are found with a predominantly solid component. A characteristic histological finding is the presence of elongated, curvilinear, thin-walled blood vessels.

It is important to note that skin biopsy, when taken superficially, may show misleading features, since the shallow portions of the tumor show benign

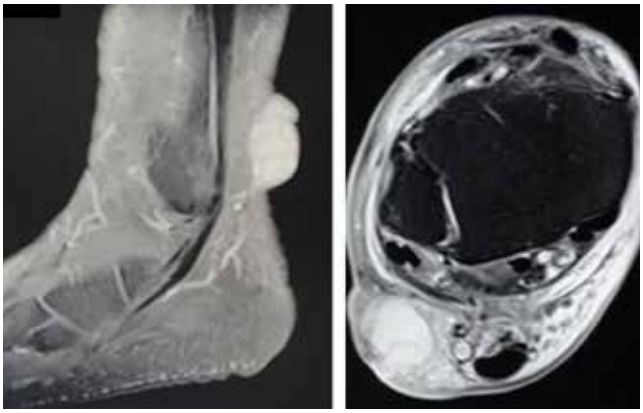

Figure 2: Magnetic resonance imaging of the ankle showing expansive lesion with defined boundaries centered in the subcutaneous plane

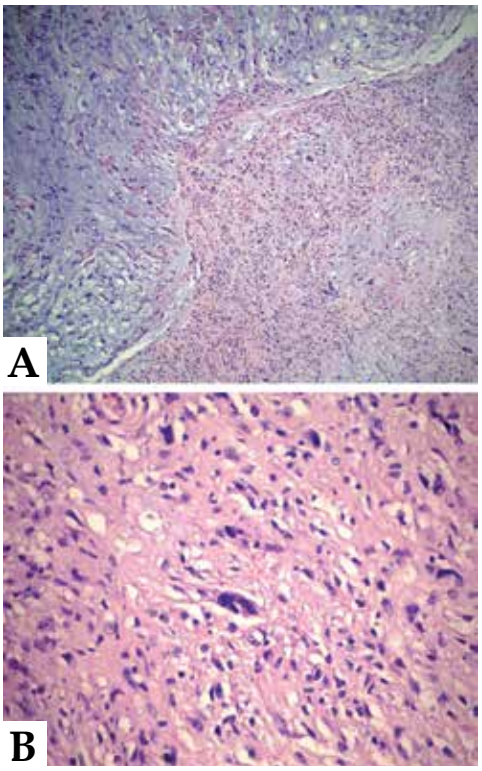

Figure 3: Intermediate grade myxofibrosarcoma. **(A)** Transition between myxoid area and cellular area. (HE, 100X). **(B)** Atypical cells with scant cytoplasm (HE, 400X).

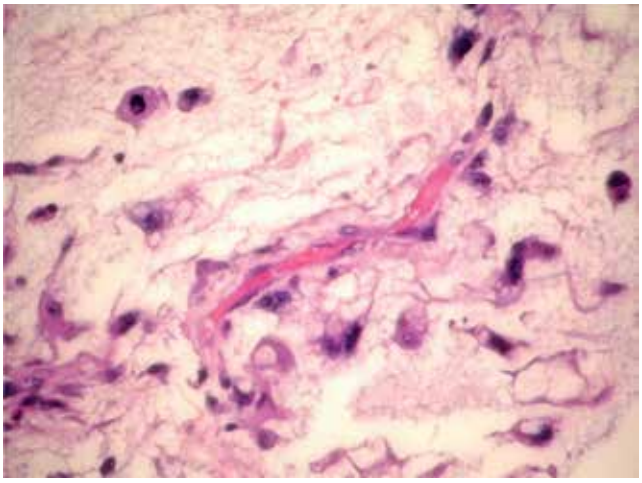

Figure 4: Myxofibrosarcoma revealing elongated, curvilinear, thin-walled blood vessel (HE, 400x)

characteristics, whereas deeper samples show histomorphological features of malignancy.[5] Therefore superficial biopsies may underclassify a high degree tumor or even classify it as a benign neoplasm. This peculiar characteristic makes it very important to represent the lesion adequately for histopathological examination.[2,6]

Due to the low specificity of immunohistochemistry in myxofibrosarcoma, the results should always be interpreted with caution. The neoplastic cells are often positive for vimentin and CD34, and rarely positive for smooth muscle actin. Desmin, cytokeratins, S100 and HMB-45; CD68 are usually negative.[1,3]

Therapy consists of surgical resection with a safety margin of at least 2 cm.[5] High local recurrence rate (50-60%), regardless of the grade, results in the indication of adjuvant radiotherapy.[1] Metastasis occur in 20-35% of high and intermediate grade tumors, especially in bones and lungs.[1,7,8] As it is a rare disease, more studies are needed to establish the benefit of chemotherapy. ❑

REFERENCES

1. Mentzel T, van den Berg E, Molenaar W. Myxofibrosarcoma. In: Fletcher C, Unni K, Mertens F, editors. WHO classification of tumors - pathology and genetics, tumors of soft tissue and bone. Lyon: IARC Press; 2002. p.102-1031.
2. Clarke LE, Zhang PJ, Crawford GH, Elenitsas R. Myxofibrosarcoma in the skin. J Cutan Pathol. 2008;35:935-40.
3. Mentzel T. Sarcomas of the skin in the elderly. Clin Dermatol. 2011;29:80-90.
4. Mansoor A, White CR Jr. Myxofibrosarcoma presenting in the skin: clinicopathological features and differential diagnosis with cutaneous myxoid neoplasms. Am J Dermatopathol. 2003;25:281-6.
5. Clarke LE. Fibrous and fibrohistiocytic neoplasms: an update. Dermatol Clin. 2012;30:643-56, vi
6. Tupinambá WL, Schettini RA, Souza Júnior Jd, Schettini AP, Rodrigues CA, Oliveira Fda S. Mixofibrossarcoma--case report. An Bras Dermatol. 2011;86:S110-3.
7. Haglund KE, Raut CP, Nascimento AF, Wang Q, George S, Baldini EH. Recurrence patterns and survival for patients with intermediate- and high-grade myxofibrosarcoma. Int J Radiat Oncol Biol Phys. 2012;82:361-7.
8. Willems SM, Debiec-Rychter M, Szuhai K, Hogendoorn PC, Sciot R. Local recurrence of myxofibrosarcoma is associated with increase in tumour grade and cytogenetic aberrations, suggesting a multistep tumour progression model. Mod Pathol. 2006;19:407-16.

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