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(CASE STUDY)

Fatal outcome in a high-grade myxofibrosarcoma: A case report

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Abstract

High-grade myxofibrosarcoma is an uncommon and aggressive soft tissue sarcoma, most frequently occurring in elderly patients and typically arising in the limbs. It demonstrates infiltrative growth, a myxoid stroma, and a high potential for metastasis. We present the case of a 74-year-old woman with a rapidly enlarging, ulcerated tumor on her left lower leg. Histopathological and immunohistochemical studies confirmed high-grade myxofibrosarcoma. Despite planned staging and therapeutic management, the patient died one month after diagnosis. This case underscores the aggressive nature of high-grade myxofibrosarcoma, even when superficially located, and highlights the importance of early detection and intervention.

Keywords: Myxofibrosarcoma; Cutaneous sarcoma; Ulcerated tumor; Aggressive tumor

1. Introduction

Soft tissue sarcomas are uncommon malignant tumors derived from mesenchymal structures and encompass a wide spectrum of histological subtypes [1]. Myxofibrosarcoma (MFS) represents one of the most frequently encountered sarcomas in older adults and shows a clear predilection for the extremities [1]. Despite its often superficial presentation, MFS may follow an aggressive clinical course. We report a case of rapidly evolving myxofibrosarcoma of the lower limb in a 74-year-old woman, resulting in early death, to highlight the potential severity of this neoplasm.

2. Case Report

A 74-year-old woman with no significant past medical or surgical history presented with a progressively enlarging lesion on the left lower leg that had been evolving for approximately eight months. Clinical examination revealed a large, ulcerated, exophytic tumor measuring about 10 cm in diameter, with an indurated base, purulent exudate, and bleeding upon contact. The mass was located on the anterolateral surface of the leg and was partially covered with hemorrhagic crusts and fibrinous material (Figure 1). Dermoscopic evaluation was not contributory due to active bleeding and abundant exudation. Palpation of regional lymph nodes revealed small, mobile, painless bilateral inguinal nodes without fixation to underlying tissues.

Given the suspicion of a malignant soft tissue tumor, an incisional biopsy was performed. Histopathological examination demonstrated a diffuse proliferation of spindle-shaped cells with elongated, hyperchromatic nuclei and eosinophilic

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cytoplasm. The tumor displayed a characteristic branching vascular pattern, accompanied by a polymorphic inflammatory infiltrate and areas of suppurative necrosis (Figure 2).

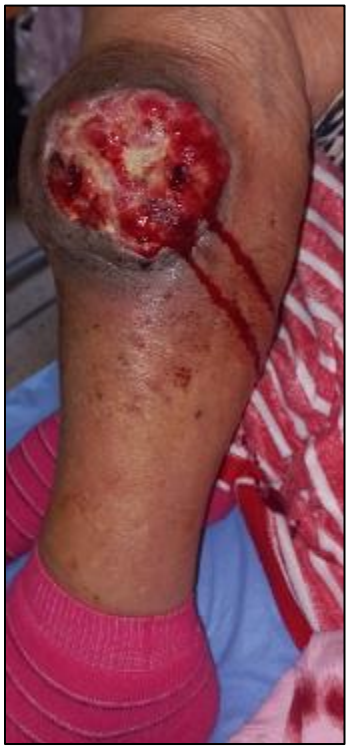

Figure 1 Dermatological examination reveals a large (approximately 10 cm), ulcerated, exophytic mass with an infiltrated base on the anterolateral aspect of the left leg, partially covered with hemorrhagic crusts and fibrinous deposits, with purulent discharge and bleeding on contact

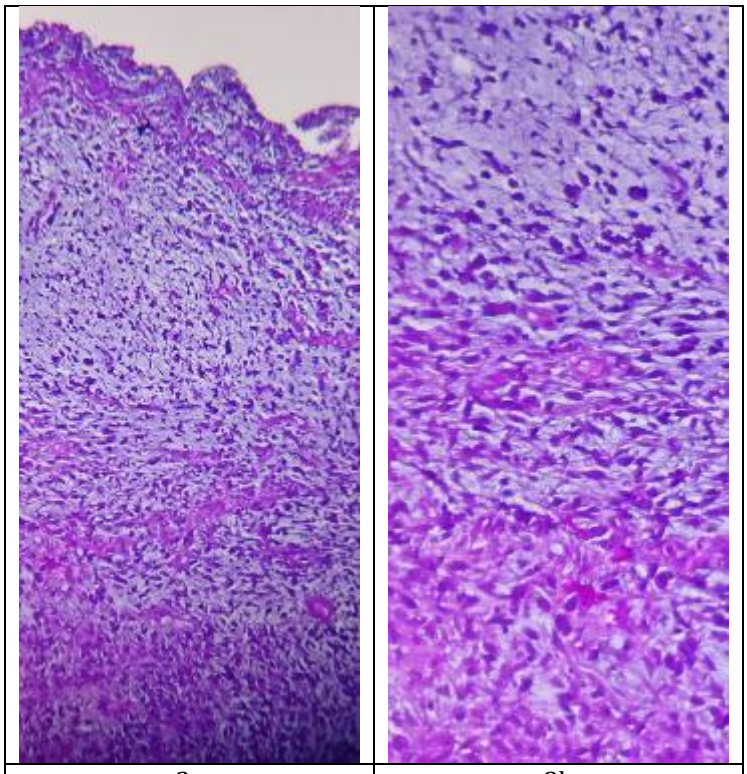

Figure 2 Myxofibrosarcoma: the tumor shows features of a high-grade pleomorphic sarcoma with areas of myxoid stroma

Immunohistochemical analysis showed focal expression of Factor XIIIa, CD163, and smooth muscle actin (SMA), while markers including HMB45, Melan-A, CD31, CD34, S100 protein, STAT6, and MUC4 were negative. The Ki-67 proliferation index was estimated at approximately 60%. These findings supported the diagnosis of high-grade myxofibrosarcoma.

Although further staging investigations and surgical evaluation were planned, they could not be completed, as the patient died one month after the diagnosis.

3. Discussion

Myxofibrosarcoma, formerly known as malignant myxoid fibrous histiocytoma, is a malignant fibroblastic tumor characterized by varying degrees of myxoid stroma, cytological atypia, and distinctive curvilinear vasculature [1]. It most frequently arises in the dermal and subcutaneous tissues, accounting for nearly two-thirds of reported cases. Initially classified as a subtype of malignant fibrous histiocytoma, MFS was recognized as a distinct pathological entity by the World Health Organization in 2002 and represents approximately 5% of all soft tissue sarcomas [2–4].

This tumor predominantly affects elderly individuals, with peak incidence in the sixth and seventh decades of life, and most commonly involves the lower extremities [5]. Clinically, MFS typically appears as slowly enlarging, painless nodules with a gelatinous or gray-white appearance; however, advanced presentations with ulceration may occur, as illustrated by our case. Histologically, MFS is composed of loosely arranged spindle or stellate cells embedded in a myxoid matrix rich in hyaluronic acid [6]. The diagnosis is based on the presence of pleomorphic spindle cells, thin-walled curvilinear blood vessels, and a myxoid extracellular background [7,8].

Tumor grade plays a crucial role in prognosis. Low-grade lesions are characterized by low cellularity and abundant myxoid stroma, whereas high-grade tumors demonstrate increased cellular density, reduced myxoid areas, multinucleated giant cells, elevated mitotic activity, and extensive necrosis [8,9]. High-grade and deeply seated tumors are associated with a significantly higher risk of local recurrence, distant metastasis, and disease-related mortality.

A hallmark feature of MFS is its infiltrative growth pattern, which can be visualized on magnetic resonance imaging as elongated extensions along fascial planes, commonly referred to as the “tail sign” [10]. Standard treatment for MFS of the trunk and limbs involves wide surgical excision, often preceded by external beam radiotherapy to improve local control and margin delineation [11]. Misinterpretation of infiltrative margins as reactive edema may lead to incomplete resection and increased recurrence rates. Chemotherapy is generally reserved for unresectable, locally advanced, or metastatic disease and typically includes anthracycline- or gemcitabine-based regimens [12]. Local recurrence rates range from 50% to 60%, and metastatic spread occurs in approximately 20–35% of intermediate- and high-grade tumors [1].

4. Conclusion

High-grade myxofibrosarcoma may exhibit rapid progression and aggressive behavior, even when confined to superficial tissues. Its infiltrative nature and significant metastatic potential underscore the importance of early diagnosis and prompt multidisciplinary management to improve patient outcomes.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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