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Case Report

# Low Grade Myxofibrosarcoma: A Case of Young Women with an Unusual Localization

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## Abstract

Low grade myxofibrosarcoma is an uncommon sarcoma with a significant potential for late recurrence and metastatic spread. Here we describe the case of young women with a low grade myxofibrosarcoma occurring in an unusual localization.

**Keywords: Low grade myxofibrosarcoma; Lower limb; Slender spindle cells; Immunochemistry**

## Introduction

Low-Grade Myxofibrosarcoma (LGMFS) is an uncommon sarcoma that was first described by Evans in 1987 [1]. It occurs mostly in a subfascial location as deep-seated intramuscular mass in the proximal extremities or trunk of young adults, although a wide age range has been noted including some pediatric cases.

This report provides clinical, dermoscopic and pathology findings in young women with a lower limb low grade myxofibrosarcoma.

## Case Presentation

A 27 years-old women was referred to our department for an exophytic tumor of the left ankle evolving a year prior to her referral. The physical examination revealed a rounded, exophytic, ulcero-budding tumor, with a fibrinous surface and an indurated base, measuring 15 cm × 13 cm, next to the medial malleolus of her left ankle (Figure 1). Dermoscopic analysis showed white yellowish areas with some glomerular vessels (Figure 2). The perilesional skin was hard and verrucous. Lymph node examination found an ipsilateral inguinal lymphadenopathy. Diagnoses of verrucous squamous cell carcinoma, sarcoma or atypical mycobacterial infection were suspected. Histopathology of a biopsy specimen showed a dermal myxoid infiltration of slender spindle cells with a low mitotic index, a myxoid stroma rich in capillary vessels and some fibrous foci organized in bands. The immunochemical data evoked a low grade myxofibrosarcoma with 5% of tumor cells strongly expressing anti Ki67, while immunostaining was negative for CD34, EMA, PS100 and MUC4. Molecular biology is not available in our context. A magnetic resonance imaging of the concerned limb showed a large locally advanced tumor infiltration in the inner side of the ankle, strongly vascularized (Figure 3). No metastatic localizations were found after performing a computed tomography scan. Due to its local extension, leg amputation with lymph node dissection associated to adjuvant radiation therapy was decided.

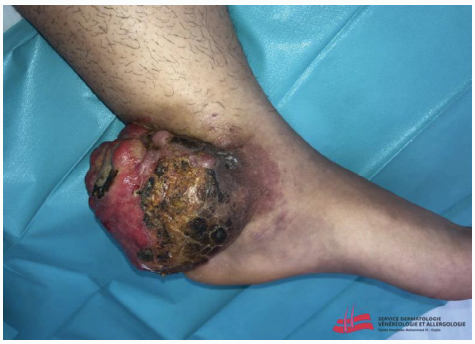

**Figure 1**: A rounded exophytic ulcero-budding tumor of the left ankle.

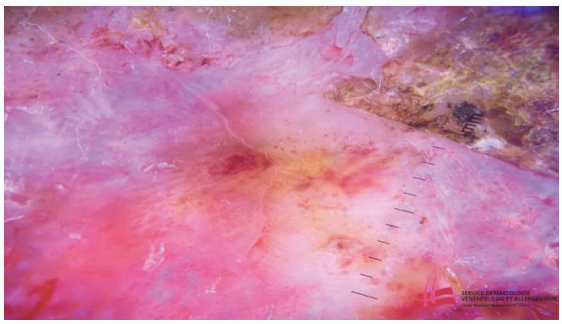

**Figure 2**: Dermoscopic findings: white yellowish areas with some glomerular vessels.

## Discussion

Myxofibrosarcoma (MFS) represents a tumor comprised of malignant spindle cells in a myxoid stroma. The entity was originally considered a myxoid variant of malignant fibrous histiocytoma. As currently classified, MFS constitutes a distinctive entity with reproducible histomorphologic and clinical features [2]. It can be graded under the FNCLCC (Federation Nationale des Centres de Lutte Contre le Cancer) system. Low grade-cases are characterized histopathologically by low cellularity, predominantly myxoid stroma,

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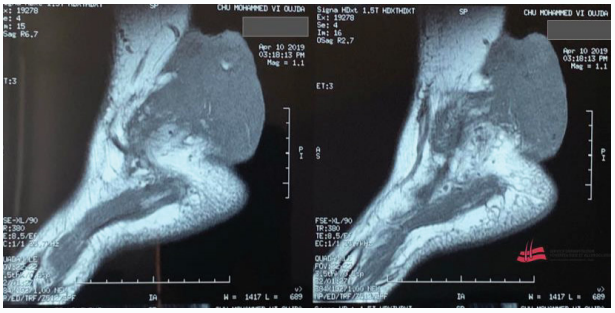

**Figure 3**: Magnetic resonance imaging: a large locally advanced tumoral infiltration in the inner side of the ankle strongly vascularized.

malignant fibroblastic cells demonstrating spindle cell morphology and cellular condensation around characteristic curvilinear blood vessels. The incidence of LGMFS has been reported as 0.18 per million [3]. It can affect patients of all ages but has a peak incidence in young adults, with a mean age of 33 years and a median of 32.5 years (age range 10 to 69 years). The male to female ratio was reported to be either equal or 3:1 [4]. The majority of LGMFS cases present as painless slow growing and deep-seated masses within the proximal extremities and trunk. Patients presenting with metastatic disease may have a decade's long history of a primary mass in the extremity or trunk [5]. Histologically, this tumor consists of slender spindle cells with long, narrow, delicate and mostly non-branching cell processes, embedded in a variable amount of collagenous stroma. Immunohistochemically, LGMFS characteristically shows strong and diffuse granular cytoplasmic immunoreactivity with MUC4 which is a highly sensitive marker in its diagnosis, labeling up to 100% of LGMFS and has been shown to be absent in most other soft tissue neoplasms. They are characterized in the majority of cases by a balanced translocation, t(7;16)(q34;p11), resulting in fusion of the FUS and CREB3L2 genes, with a small minority of cases showing a variant FUS-CREB3L1 fusion resulting from t(11;16)(p11;p11). LGMFS is not expected to be very chemo- or radiosensitive due to its low nuclear grade and infrequent mitotic activity. The best response to chemotherapy was short-term stabilization of disease progression with Trabectedin. Another study by le Cesne et al. [6], suggested that Trabectedin could offer some benefit in translocation-related soft tissue sarcomas such as LGMFS. However, surgical excision with clear resection margins remains the first line treatment option. In case of local recurrence or metastasis (especially lung metastasis) Proton Beam Therapy (PBT) and pazopanib can be effective [7].

## Conclusion

The diagnosis of LGMFS may be challenging. However, correct recognition is crucial as this tumor has significant potential for late recurrence and metastatic spread. Appropriate morphologic and immunohistochemical findings with cytogenetic and molecular analysis (FISH and RT-PCR) are optimal to retain the diagnosis.

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