

Multiple Low-Grade Fibromyxoid Sarcoma on the Upper Arms with Atypical Histological Presentation

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Key Words

Low-grade fibromyxoid sarcoma · Multiple recurrence · Palisaded granuloma-like bodies (rosettes)

Abstract

Low-grade fibromyxoid sarcoma (LGFMS) is a rare variant of spindle cell tumor that is composed of collagen-rich and myxoid parts. We describe the case of a 61-year-old Japanese patient with multiple, recurrent LGFMS on the upper arms with atypical histological presentation. In the present case, we resected the tumor several times with a minimal surgical margin, as in Moh's microsurgery. However, this can frequently lead to local recurrence of the tumor. Our case suggested that, regarding mesenchymal tumors with potential of malignancy in the skin, an initial wide excision is indispensable for complete remission of the tumor, even for low-grade malignancy such as LGFMS.

Introduction

Fibrosarcomas are mesenchymal neoplasms with fibrous differentiation that have a low-grade potential for malignancy. Various types of low-grade fibrosarcoma were previously reported: low-grade myxofibrosarcoma [1–3], low-grade fibromyxoid sarcoma (LGFMS), hyalinizing spindle cell tumor with giant rosettes [4, 5], sclerosing epithelioid fibrosarcoma, low-grade fibrosarcoma with palisaded granuloma-like bodies [6], and low-grade fibrosarcomas not otherwise specified [7]. Among them, LGFMS is a rare variant of spindle cell tumor that is composed of collagen-rich and myxoid parts [1, 2]. LGFMS proliferates mainly in subcutaneous or superficial soft tissue on the proximal extremities in

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young to middle-aged adults. Clinically, the tumor sometimes recurs locally but rarely has a metastatic potential [6]. In this report, we present a case of multiple LGFMS on the upper arms with atypical histological presentation.

Case Report

A 61-year-old Japanese female presented to our clinic with an 8-month history of asymptomatic subcutaneous nodules on her arm. On her first visit to our outpatient clinic, physical examination revealed an elastic, hard subcutaneous nodule on her bilateral upper arms (fig. 1). The nodules were approximately 25 mm in diameter. We excised two nodules and histological findings revealed degenerated collagen fibers and necrosis, surrounded by histiocytes and partially palisaded granuloma-like bodies (collagen rosettes) (fig. 2a, b). We observed spindle-shaped cells that had eosinophilic cytoplasm, with no nuclear atypia or mitotic activity (fig. 2b). Mucin deposition was prominent at the centers of the granuloma-like bodies (fig. 2c). We diagnosed this tumor as a subcutaneous type of granuloma annulare or myxofibroma. One year later, the patient noticed a subcutaneous nodule on her left arm and we excised the nodule again. Histological findings were similar to those of the previous lesion. A half year later, a subcutaneous nodule, 20 mm in diameter, appeared on her left arm near the operative area. Then, we screened for a possible internal malignancy with positron emission tomography but found none. We excised the nodule again below the muscle fascia, and the histopathological findings revealed myxoedematous proliferation, degenerated collagen fibers and necrosis, surrounded by lymphocytes, histiocytes and multinucleated giant cells in the dermal to subcutaneous area (fig. 2d). Alcian-blue PAS staining revealed mucin deposition at the center of the tumor. From the above data, we diagnosed this tumor as LGFMS. Two years after the last excision, there was no sign of recurrence or metastasis.

Discussion

In this report, we describe a case of multiple, recurrent low-grade fibromyxoid sarcoma on the upper arms. The histological findings of the nodules revealed degenerated collagen fibers and necrosis, surrounded by histiocytes and partially palisaded granuloma-like bodies (collagen rosettes), mimicking a subcutaneous type of generalized granuloma annulare. Unexpectedly, the nodules recurred several times at both local sites and sites remote from the resected area. Concerning the most recent nodule, histological findings revealed prominent mucin deposition at the center of the tumor. From the clinical course and histological findings, we finally diagnosed this patient with multiple, recurrent LGFMS on the arm.

LGFMS is a distinctive variant of fibrosarcoma that was first described by Evans [8]. It is characterized by spindle-shaped cells arranged in a whorled pattern with alternating fibrous and myxoid stroma. About 30% of these tumors also develop collagen rosettes [9]. Local recurrence is seen in 9.3% of patients and metastasis occurs in 5.6% of patients [9]. In Japan, on the other hand, local recurrence is seen in 12.9% of patients and usually no metastasis occurs [10]. This discrepancy might be caused by differences in the types of LGFMS, since the concept of LGFMS has been developing during recent decades. For instance, Mansoor and White [11] reported another variant of subcutaneous malignancies similar to LMGFS, myxofibrosarcoma presenting in the skin, which is characterized by low-grade myxoid tumors with dermal involvement but no collagen rosettes. LGFMS and myxofibrosarcoma

might be misdiagnosed as benign dermal myxoid neoplasm, including superficial angiomyxoma, myxoid neurofibroma, nerve sheath myxoma, and myxoid dermatofibrosarcoma protuberans [12]. Interestingly, Folpe et al. [9] reported that in 15% of patients with LGFMS, a histologic diagnosis was delayed by over 5 years, which suggested difficulties in diagnosing LGFMS. Our case was also confusing because from the beginning, the tumors were observed bilaterally on the upper arms, which strongly suggested an inflammatory disorder, such as a granulomatous disorder. After several excisional biopsies, these tumors revealed bilateral primary lesions of LGFMS. At the final excision, we excised the tumor below the muscle fascia, and there has been no sign of local or remote recurrence for 2 years. Our case suggested that for mesenchymal tumors with potential of malignancy in the skin, initial wide excision is indispensable for complete remission of the tumor, even for low-grade malignancy such as LGFMS.

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Disclosure Statement

There are no conflicts of interest.

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Fig. 1. An elastic, hard subcutaneous nodule on the bilateral upper arms is shown.

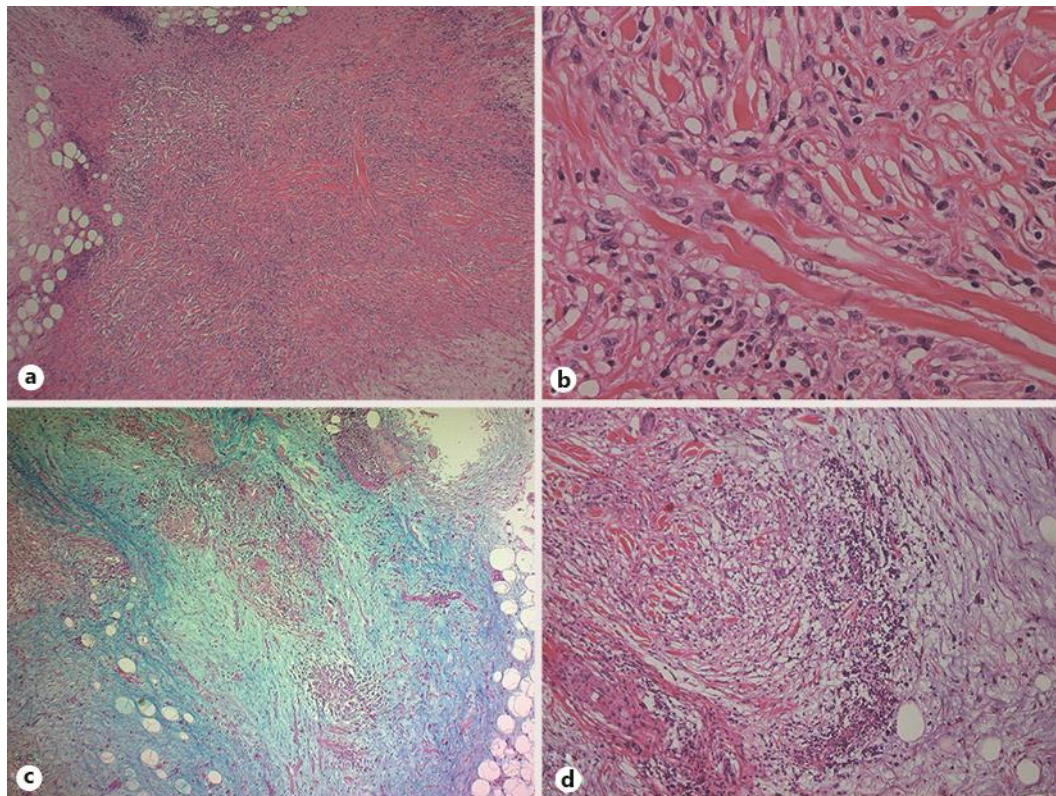


Fig. 2. Degenerated collagen fibers and necrosis surrounded by histiocytes and partially palisaded granuloma-like bodies (rosettes, **a**). Spindle-shaped cells with eosinophilic cytoplasm, showing no nuclear atypia or mitotic activity (**b**). Mucin deposition is prominent at the centers of the granuloma-like bodies (**c**). Myxoedematous proliferation, degenerated collagen fibers and necrosis surrounded by lymphocytes, histiocytes and multinucleated giant cells in the dermal to subcutaneous area, partially resembling a palisading granuloma (**d**).