Fibromyxoid sarcoma of the leg

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ABSTRACT

A 48-year-old female with an atypical plaque-like lesion of the lower leg is presented in this article. Histologic investigation revealed a rare low-grade fibromyxoid sarcoma (pT1a cN0 cM0; stage Ia) of suprafascial localization. Staging of the patient did not reveal metastatic spread. The tumor was surgically removed with wide safety margins. The defect was closed using a mesh graft transplant and vacuum-assisted closure. Healing was complete. Regular follow-up for at least 5 years is recommended. Besides the rareness of this tumor, this case is also remarkable because of the localization on the lower leg and the suprafascial soft tissue.

Key words: Fibromyxoid sarcoma, histology, surgery

INTRODUCTION

Sarcomas are mesenchymal neoplasms with various lines of differentiation, i.e., fibrocytic, myogenic, neurogenic, vascular, chondro-osseus, or undefined. The low-grade fibromyxoid sarcoma (LGFS) is a very rare entity. It is a spindle-cell tumor composed of collagen-rich and myxoid parts.[1,2] About 40% of these tumors also develop collagen rosettes.[3]

LGFS prefers subfascial soft tissue layers on the trunk and proximal extremities in younger adults but can also develop in internal organs. Pediatric cases have also been reported.[1-3]

Herein, we present a 48-year-old female patient who developed a LGFS on the lower leg. The clinical presentation, histopathology, surgical treatment, and follow-up are discussed.

CASE REPORT

A 48-year-old woman was referred to our department because she had developed a slow-growing plaque below the left knee for 2 years. A diagnostic biopsy had been taken by the referring dermatologist that suggested an LGFS.

On examination we found an otherwise healthy, slim, woman with a symptomless firm subcutaneous plaque of about 2 cm size on the anterior aspect of the left lower leg [Figure 1]. There was some bluish discoloration and circumscribed ulceration but no erythema or warmth.

Routine laboratory tests were unremarkable. Thoracic dual-energy x-ray and abdominal and lymph node ultrasound excluded a metastatic spread. Histology revealed a spindle-cell tumor, with mild atypia and fibroblast-like morphology [Figure 2]. The eosinophilic cells did not show increased mitotic activity. Cells were arranged in a whorled or plexiform pattern, with alternating collagenous stroma and myxoid zones. The cells stained positively for vimentin, but were negative for CD34 and S100. The covering epidermis was partially ulcerated. A final diagnosis of Low GradeF ibromyxoid Sarcoma was reached. Under general anesthesia, the tumor was surgically removed with a wide safety margin (>3 cm). The defect was covered by a mesh graft transplant covered by vacuum-assisted closure (VAC™; KCI International) [Figure 3]. Compression stockings were prescribed to protect the transplant and prevent leg edema. A regular follow-up for at least 5 years was recommended after consultation with the referring dermatologist.

DISCUSSION

LGFS is a very rare and distinctive type of fibrosarcoma that was first described by Evans in 1987.[1] There is a discrepancy between the bland histologic features with sparse mitotic figures and absent or mild nuclear and cellular pleomorphism

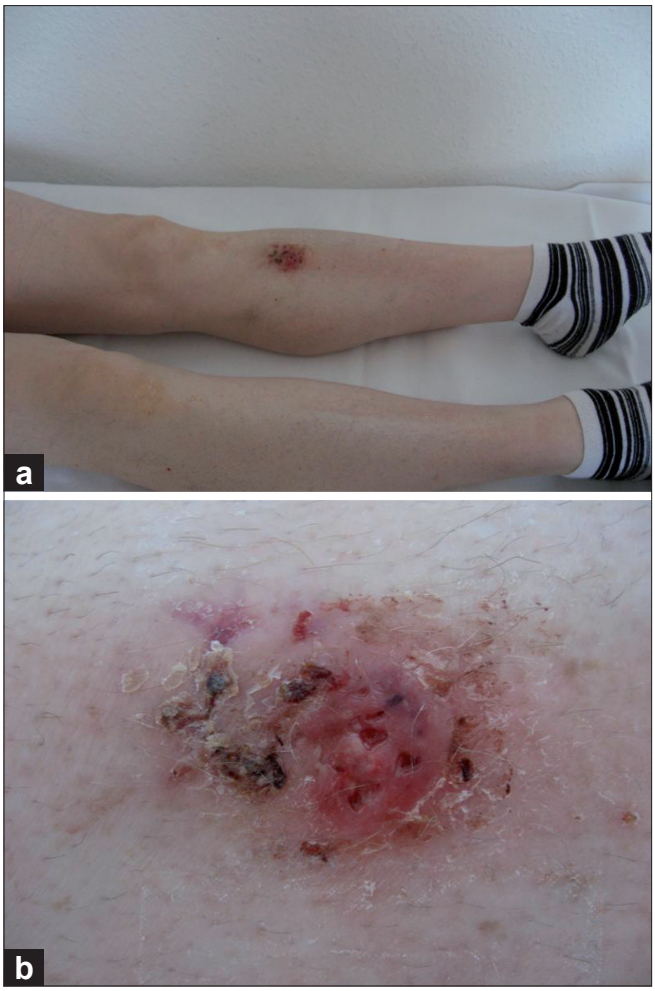

Figure 1: Low-grade fibromyxoid sarcoma of the left lower leg. (a) Overview and (b) the ulcerated honeycomb-like plaque

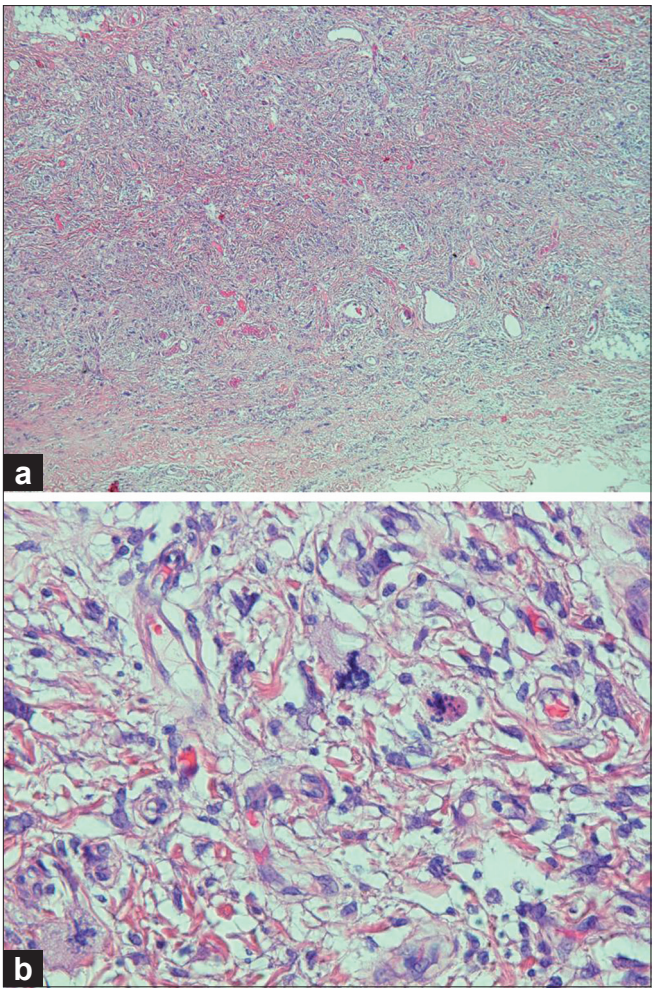

Figure 2: Histopathology. (a) Overview (H&E, ×4) and (b) detail (H&E, ×40)

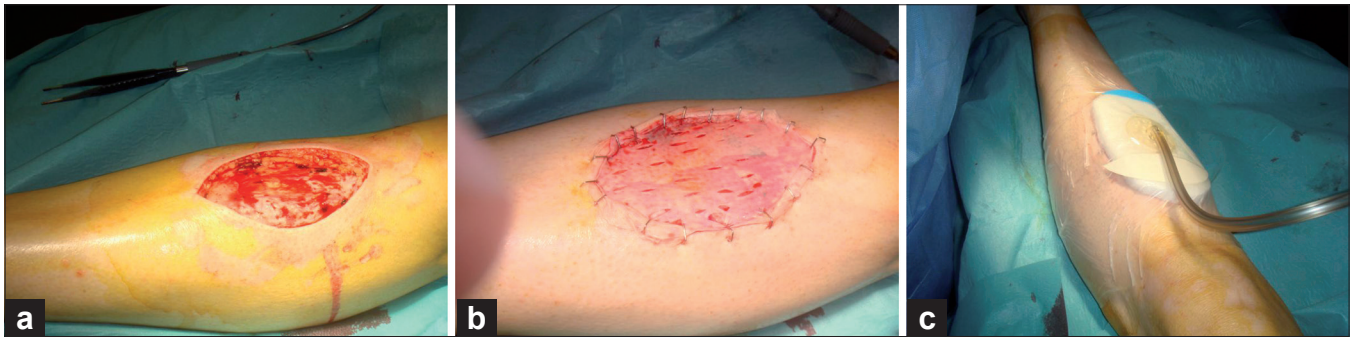

Figure 3: Surgical procedure. (a) Wide excision of suprafascial soft tissue; (b) mesh graft transplantation; and (c) placement of microporous white sponge for vacuum-assisted closure above the transplant

and the anaplasia. The hyalinizing spindle-cell tumor with giant rosettes is considered a subtype of LGFS.[1-3] The tumor is further characterized by t(7;16)(q34;p11) translocation and fusion of *FUS* and *CREB3L1* genes.[4,5] Tumor cell phenotype is positive for vimentin, EMA, CD99, and bcl-2, but negative for CD34, SMA, S-100, desmin, keratins, neuron-specific enolase, and CD177.[5] The differential diagnosis includes other types of sarcomas, myxoma, neurofibroma, peripheral sheath tumor, histocytoma, desmoid tumor, and others.[2,3,5]

LGFS usually presents as a painless, slow-growing, soft tissue malignancy. The diagnosis often is delayed – mainly because

the patients do not seek treatment early. Folpe *et al*. (2000) reported that in 15% of patients a histologic diagnosis was delayed by >5 years. In our case the delay was >2 years.[3]

Although the histopathologic features suggest a low-grade malignancy, local recurrence is seen in more than 50% of patients and metastasis occurs in 6% of patients.[3] Tumor cell dormancy is responsible for very late metastasis in some patients, with 45 years being the longest period observed between primary surgery and metastasis.[6,7] Therefore, patients should be encouraged to have regular follow-up.

The present case did not show any subfascial involvement. Complete surgery with wide margins (>3 cm) is the most important procedure. We used a combination of mesh graft transplantation and vacuum-assisted closure to cover the large defect. It has been demonstrated recently that such a combination, with microporous sponge for vacuum-assisted closure, results in a significantly improved take rate.[8] The prognosis for superficial LGFS seems to be better than that for deep LGFS.[9]

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