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Low – Grade Fibromyxoid Sarcoma: Case Report

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Abstract

Low-grade fibromyxoid sarcoma (LGFMS) an uncommon type of soft-tissue sarcoma, with uncertain epidemiology due to the rarity of this disease, however, it is more frequent in young adults. This tumor originates from subcutaneous cellular tissue or muscle, predominantly in the lower extremities, shoulders, and inguinal region. We present a case of a 64-year-old woman with a necrotic exophytic tumor in the proximal third of the right leg with immunohistochemistry demonstrating (LGFMS).

Keywords: Low-grade fibromyxoid sarcoma; Soft-tissue sarcoma; Morphology; Prognosis

Introduction

Low-grade fibromyxoid sarcoma (LGFMS) was described for the first time by Evans, also called Evans's tumor [1,2]. This tumor is a bland spindle cell neoplasm, usually arises from deep soft tissues or subcutaneous tissue or muscle of the extremities or trunk [1-6]. This tumor presents as a slow growing mass, with an indolent clinical course, and in some cases could be metastatic [1,3-5,7].

It is a rare neoplasm, but probably subdiagnostic, because it arises in association with other entities [2,4,8,9]. The incidence of LGFMS has been reported as 0.18 per million and can affect patients of all ages, especially in young adults, in both men and women [1-4,10], The male to female ratio has been reported as 7:2 in infants and 8:5 in young adolescents [6]. Furthermore, in adult cases, the male to female ratio was reported 3:1 [3,6], with a mean age of 33 years and a median of 32.5 (Age range: 10 to 69) [3,6]. The tumor is more commonly found in the inferior extremities, the shoulder, or the inguinal region. However, there are reports of cases in the chest, armpit, buttocks and neck [4,6].

In the last century, data about recurrence, metastasis, and biological behavior were very limited [3,8,9]. After this revision, reported a potential for both recurrence (9-10%) and metastasis (5-6%). Of all the metastatic tumors that developed, they were most seen in the lungs [6] and 1 % of patients who died of LGFMS succumbed to the disease at a mean of 38 months and a median of 24 months from the initial diagnosis [5,6].

LGFMS has shown to have a translocation between chromosome 7 and 16, resulting a chimeric fusion protein derived from the fusion in the sarcoma (FUS); gene (**sarcoma gene**) of chromosome 16p11, and the cAMP-responsive element-binding protein (3-like 2) (CREB3L2) gene of 17q33. A minority of cases have been shown to display a FUS-CREB3L1 and CREB3L2 gene [2,4,5].

LGFMS has defined clinical characteristics, but not specific to the disease, that's the reason why the diagnosis is a challenge. Immunohistochemistry has a fundamental role and a huge impact, because it implicates adequate resection margins for higher survival of patients [3,6].

We will present a case report, to demonstrate the importance of the early diagnosis of this kind of disease, especially in developing countries, where accessibility to specialized medical services is very difficult and delay over time dramatically impacts to prognosis and the natural course of the disease.

Clinical Case

A 64-year-old woman from Caucasia (Antioquia-Colombia), who was admitted to emergency department at Simon Bolivar hospital in Bogotá, due to 9-month evolution of an exophytic

lesion in the proximal third of her right leg, which was rapidly growing, painful, bleeding intermittently, produced serohematic exudate and had a foul odor (Figures 1&2). She had a prior biopsy from that lesion, with pathological diagnosis of dermatofibroma. Physical examination revealed that in the anterior region of the proximal third of the right leg, there was an exophytic, friable, erythematous-violet tumor, infiltrated with multiple ulcers and necrotic crusts on the surface, with a size of 10x8 cm. Since there was not an adequate clinical and histopathological correlation we decided to take a new skin biopsy, which showed spindle cells that alternated epithelioid areas (Figure 3) with myxoid areas (Figure 4), and fibrous sarcoma with minimal fibroblastic cells and nuclear pleomorphism (Figure 5). Findings suggest a low-grade fibromyxoid sarcoma. Immunohistochemistry was negative for S100, CD34, desmin, CD68, Melan A, and ASM (Figure 6). With the definitive diagnosis, an enlarged local resection with 4 cm margins including the fascia was decided; with the result of histopathology that describes tumor-free edges of the lesion, we decided to use partial thickness skin autografts to cover the residual defect (Figure 7). Among the extension studies, although the patient was asymptomatic from the respiratory point of view, computerized axial tomography was performed with contrast of the thorax, finding a mass of 2cm in diameter at the right lung base, with spiculated edges, giving us the first diagnostic was lung metastasis, so she was referred to comprehensive management by oncology, but the patient did not agree to complete the treatment.

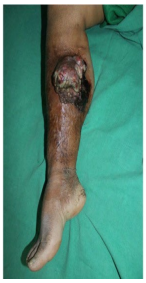

Figure 1: In this photo, we can see the lesion in the area of the right leg and calf, approximately 7-8 cm, with necrotic areas adjacent to the lesion.

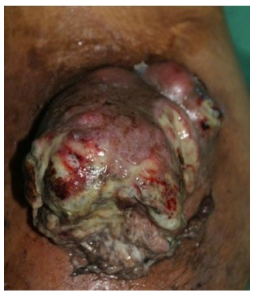

Figure 2: The central, lobulated, exophytic lesion with necrotic borders, with hematic and purulent discharge is observed.

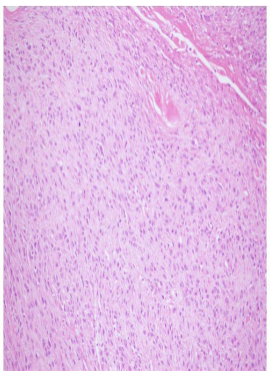

Figure 3: The histological plate showed spindle cells alternating with epithelioid areas.

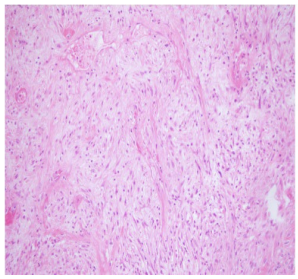

Figure 4: The histological plate shows spindle cells with myxoid areas.

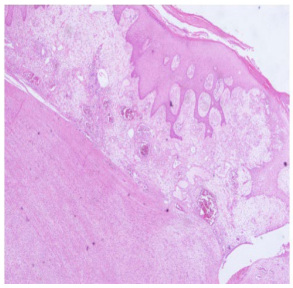

Figure 5: The histological plate shows a fibrous sarcoma with minimal fibroblastic cells and nuclear pleomorphism.

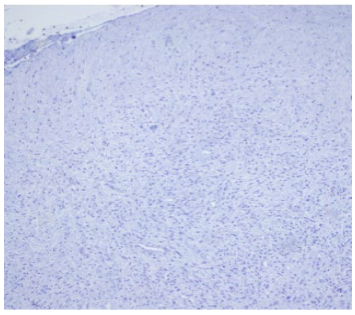

Figure 6: Immunohistochemistry was negative for S100, CD34, desmin, CD68, Melan A and ASM.

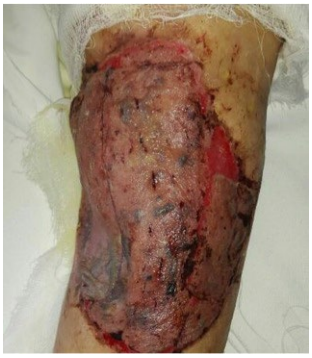

Figure 7: One week postoperative: The patient underwent surgery to remove the tumor, and the skin defect was corrected by means of a connective tissue graft.

Discussion

LGFMS is an infrequent tumor whose histopathological appearance can simulate a benign tumor, making its diagnosis a great challenge, and sometimes could be a confusing diagnosis like in our case [2,3]. LGFMS macroscopically as a well-defined mass ranging in size from 1 cm up to 20 cm, with a white fibrous cut surface, often with glistening myxoid areas [9]. On histological examination, these tumors can be circumscribed or infiltrative, and are predominantly composed of bland-appearing spindle cells with small, angulated nuclei with inconspicuous nucleoli and scant, wispy cytoplasm. Mitotic figures tend to be absent or sparse, although a mitotic index of > 5/50 high power fields and tumor cell necrosis can be seen in < 10% of cases [1,2,9,11]. The tumor cells are arranged in a patternless or whorled growth pattern usually showing an abrupt transition from myxoid to densely collagenized areas. A fascicular growth pattern is sometimes seen, which can be associated with cellular atypia and increased cellularity, and herringbone or storiform patterns can be focally present.

Perivascular aggregates of these cells may form incipient rosettes. Intranuclear inclusions may be seen in a few of the rounded cells at the edge of the rosettes [1,2,12]. The rosettes often coalesce into long cords or bands of dense hyalinization [1,2,4,5,12].

Immunohistochemistry can help distinguish these malignancies from LGFMS, characteristically shows strong and diffuse granular cytoplasmic immunoreactivity with MUC4 which is a highly sensitive marker in its diagnosis, labeling up to 100% of LGFMS, and has shown to be absent in most other soft tissue neoplasms [12,13].

The first immunohistochemical studies of LGFMS showed that most tumor cells were strongly positive for vimentin. Since then, epithelial membrane antigen (EMA) expression has shown a consistent finding. CD99 and Bcl-2 expression has also been demonstrated in the majority of LGFMS. However, these markers are commonly expressed in other soft tissue neoplasms such as synovial sarcoma and solitary fibrous tumor, therefore, it has a low diagnostic value. Focal positivity for smooth muscle actin (SMA), desmin, CD34, and cytokeratin is rarely seen, whilst S100 protein, GFAP, h-caldesmon, beta-catenin, MDM2 enolase, epithelial membrane antigen, CD68, ALK1, S-100 and CD99, and CD117 are typically negative [2,5,10]; as in our case those markers were negative.

The spindle cells are positive for vimentin and negative for enolase, epithelial membrane antigen, cytokeratin, desmin, CD34, CD68, ALK1, S-100, and CD99 [2,3,10,12]; as in our case where the markers were negative.

It is considered a pathology with high local recurrence and late metastases of up to 15 years with an average of 3 and a half years; the lung and pleura being the most frequently involved [1-3], as in our case. However, these patients have prolonged survival even after finding metastases. Metastatic lesions in existing reports do not have a follow-up of more than 5 years, neither the risk of limb loss [1-3], but we know that it is a latent reality of this disease. Unfortunately, the literature until today only indicates the rapid progression, so after the analysis carried out of this case, the initial margin should be taken as malignancy, especially in countries such as ours where the patient is unlikely to revisit for the same cause [12].

The reconstruction of this type of secondary deffects, in our opinion, considering the case-patient evolution, should be focused using a skin graft of partial or total thickness, because the size, not exposure of noble structures and not compromised extremities folds, it allows to reduce the time in surgery, pedicles are not sacrificed and the donor area recovers quickly, avoiding the risk of the local growth limits use of flaps [14,15].

Conclusions

The case of a patient with a tumor in the proximal third of the right leg is presented with clinical characteristics of malignancy, but in this particular case, the pathway until the final diagnosis wasn't easy. Initially, histopathology was mistaken for a dermatofibroma; fortunately, to comprehensive management linking the clinic, pathology, and immunohistochemistry, the diagnosis of low differentiation fibromyxoid sarcoma was reached. The knowledge of this disease and early detection using immunohistochemistry, will allow timely surgical treatment, prolong survival and prevent local relapses of these patients.

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