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An Atypical Primary Subcutaneous Leiomyosarcoma of the back

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Abstract

Subcutaneous leiomyosarcomas are rare tumors representing 1% to 2% of all superficial soft tissue malignancies. Although they can appear anywhere in the body, they most often occur in the lower limbs. The incidence of subcutaneous LMS located at the upper back is very rare.

We report an historic case of a 47-year-old man admitted to the dermatology department for the management of a large subcutaneous upper back leiomyosarcoma, measuring 08 cm of size, confirmed by histopathology.

Keywords Subcutaneous Leiomyosarcomas; Back; Malignant Tumor; Aggressive Prognosis

Abbreviations LMS: Leiomyosarcomas; SLMS: Superficial leiomyosarcomas

Introduction

Leiomyosarcoma is an uncommon malignant tumour that develops in the smooth muscle. Subcutaneous leiomyosarcomas represent 1% to 2% of all superficial soft tissue malignancies. It often affects patients between the ages of 50 and 70 years. It divided into two groups: cutaneous LMS of dermal origin arising from the arrector pili with possible extension into underlying tissues, while the subcutaneous LMS are arising from the smooth muscle wall of blood vessels and has a severe prognosis in terms of recurrence and metastasis [1]. The diagnosis of these tumors is often delayed due to their rarity and non-specificity; hence the interest of our case report.

Case Report

A 47-year-old man, without significant pathological history, admitted for a painless budding lesion of the upper back, evolving since 5 months with weight loss (10 kilogram in 5 months). On clinical examination, the tumor was measuring 8 cm long with a firm consistency, indurated base and fibrinous surface easily bleeding on contact (Figure 1). A computed tomography study showed a mass of bulky multi-lobed, tissue density, infiltrating the subcutaneous tissue by adhering to the deltoid muscle, there was 3 hypodense lesions in the segment VI of the liver and multiple dense nodules distributed diffusely in the two pulmonary fields without regional lymphadenopathy.

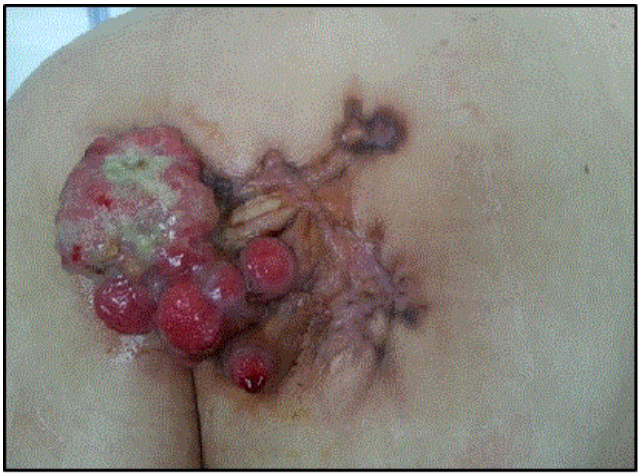

Figure 1: Tumor of the upper back, measuring 8 cm long axis, with indurated base and fibrinous surface easily bleeding on contact.

Under general anaesthesia an en block wide excision of the mass was performed with a partially removal of the surrounding muscles (latissimus dorsi and infraspinatus). Histology revealed the presence of mesenchymal tumor proliferation in the dermis made up of fusiform cells arranged in bundles in a fibrous stroma with nuclear atypia. The Ki67 proliferation index was high at 30%. The immunohistochemical staining was positive for smooth muscular actin (AML) and H-caldesmone and negative for CD34 and CD31. The conclusion of the histological analysis was: subcutaneous leiomyosarcoma grade 2 (Figure 2). With the support of these findings, the diagnosis of a subcutaneous leiomyosarcoma with hepatic and pulmonary metastases was made. Anthracycline chemotherapy was indicated for this patient, but he died after the first chemotherapy session.

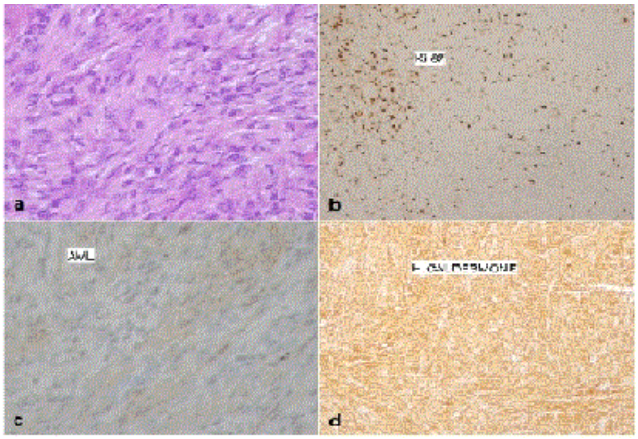

Figure 2: Histological aspect of the tumor. (a) Fascicles of spindle cells (Hematoxylin-eosin stain; x 40); (b, c and d) Immunohistochemical staining positive for smooth muscular actin (AML) and H-caldesmone, Ki67 proliferation index at 30% (x 40)

Discussion

Leiomyosarcoma is a very rare superficial soft tissue sarcoma. It represents 2.3 to 5.3% of all these sarcomas. It's appeared most often between 40 and 60 years, with male predominance. The etiology is unknown. Predisposing factors include leiomyomas, trauma and radiation exposure [2]. Our observation is characterized by an atypical clinical manifestation and the location on the upper back is unusual, without any notion of trauma or precancerous lesions.

This tumor can be anywhere in the body, but there is a predilection for the extremities, especially the thighs. Clinically, SLMS can be presented as a solitary red nodule, from 5 to 60 mm. It can have a smooth, rough, warty, ulcerated or indurated and hemorrhagic surface. Clinicals features of leiomyosarcoma may occasionally lead to confusion with basal cell carcinomas, squamous cell carcinomas, dermatofibromas, lipomas, neurofibromas, and benign papillomas [3].

Histologically, the tumour is made up of a cellular proliferation organized into fusiform cell bundles with irregular disposition. The immunohistochemical study is a complementary technique to the eosine hematoxylin study which is performed on fixed and paraffin-embedded tissues. It is useful in the diagnosis of badly differentiated forms. Classical immune-phenotyping are based of positive staining for vimentin, desmin and smooth muscle actin [4]. In our case, it was positive for actin but negative for desmine. Actin is more sensitive than desmin for smooth muscle tumours and has been signaled to be positive in 100% of the cases in many series of leiomyosarcomas. A recent study on cutaneous sarcomas showed that transgeline, which is a new myogenic marker, is the best marker in LMS [5].

Treatment of subcutaneous LMS is based on surgical excision of the tumor with a lateral margin of 3-5 cm and a depth that includes subcutaneous tissue and fascia to avoid recurrence. The interest of adjuvant therapy in patients with superficial leiomyosarcoma is controversial. SLMS are often resistant to radiation and chemotherapy. Chemotherapy has been recommended in the treatment of metastatic cases but the response rate was 15-30% and the median survival was one year. Radiotherapy is indicated for high-grade tumours to reduce the risk of recurrence, in case of tumors larger than 5 cm and in combination with surgery for recurrent tumours.

Factors of unfavorable prognostic involve the size of the tumor (5 cm), deep extension with fascia involvement, high degree of malignancy and presence of necrosis and intratumoral vascular invasion [6]. After surgical excision, the tumour may recur locally in 40% to 60% of patients or give distant metastases in 20% to 40% of cases, hence the importance of good follow-up by clinical examination and CT scan [7].

Conclusion

We report an historic case of large subcutaneous upper back leiomyosarcoma with defavorable prognosis. Dermatologist must recognize this clinical entity, and quickly conduct a histological and immunohistochemical study to be able to eliminate others differential diagnosis and ensure proper management.

The authors certify that they have obtained all appropriate patient consent forms.

Conflicts of Interest

The authors declare no conflict of interest.

References:

1. Weiss SW, Goldblum JR (2001) Leiomyosarcoma: Enzinger and Weiss's soft tissue tumors (4th Edn) Saunders Company, USA.
2. Auroy S, Contesso G, Spatz A, Genin J, Marqulis A, et al. (1999) Primary cutaneous leiomyosarcoma: 32 cases. Ann Dermatol Venereol 126: 235-242.
3. Deneve JL, Messina JL, Bui, MM, et al. (2013) Cutaneous leiomyosarcoma: treatment and outcomes with a standardized margin of resection. Cancer Control 20: 307-312.
4. Guillen DR, Cockerell CJ (2001) Cutaneous and subcutaneous sarcomas. Clinics in Dermatology 19: 262-268.
5. Hassouna JB, Saadallah F, Mezghani B, Ali Ayedi M, Charfi L, et al. (2015) Primary cutaneous leiomyosarcoma of the flank: Case report and review. J Dermatol Dermatol Surg 19: 51-54.
6. Svarvar C, Böhling T, Berlin O, Gustafson P, Follerås G et al. (2007) Clinical course of non-visceral soft tissue leiomyosarcoma in 225 patients from the Scandinavian sarcoma group. Cancer 109: 282-291.
7. Palla L, Gentile P, Cannatà C, Ascenzi P, Buonomo O et al. (2009) A neglected giant subcutaneous leiomyosarcoma of the shoulder: a case report. Eur Rev Med Pharmacol Sci 13: 389-392.