CASE REPORT

Cutaneous leiomyosarcoma on the trunk

Leiomiossarcoma cutâneo no tronco

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Abstract: O Leiomiossarcoma cutâneo é um tumor maligno que representa 7% dos sarcomas dos tecidos moles, afetando, mais frequentemente, homens entre os 50-70 anos. A localização, no tronco, é atípica, constituindo 10 a 15% dos casos. A radioterapia e o traumatismo prévio têm sido referidos como fatores de risco. Descrevemos um homem de 57 anos, com tumor eritemato-violáceo, indolor, de consistência dura, localizado na região pré-esternal, com cerca de oito meses de evolução. A histopatologia evidenciou tumor maligno de células fusiformes, com núcleos em forma de "charuto", pleomórficos, com alto índice mitótico, ocupando toda a espessura da derme. Estas células expressaram actina do músculo liso, desmina e vimentina e foram negativas para proteína S-100 e pancitoqueratina. Foi efetuada a exérese cirúrgica radical do tumor.
Palavras-chave: Imuno-histoquímica; Leiomiossarcoma; Sarcoma

Resumo: Cutaneous leiomyosarcoma is a relatively uncommon tumor that accounts for 7% of all soft tissue sarcomas. It occurs more frequently in males between 50-70 years and only 10-15% of cases are located on the trunk. Radiotherapy and previous trauma have been implied as risk factors. We report the case of a 57 year-old male with an eight-month history of a hard painless erythematous-violaceous tumor on the presternal region. Histopathology evidenced a malignant spindle cell tumor, "cigar" shaped, with pleomorphic nuclei and a high mitotic index that occupied the entire dermal thickness. Immunohistochemical staining of the tumor cells was positive for smooth muscle actin, desmin and vimentin and negative for S-100 protein and pan-cytokeratin, which supported the diagnosis of dermal leiomyosarcoma. Radical surgery was performed to remove the tumor.
Keywords: Immunohistochemistry; Leiomyosarcoma; Sarcoma

INTRODUCTION

The cutaneous leiomyosarcoma represents around 7% of all soft tissue sarcomas and its location on the trunk is rare (10-15%). The primary cutaneous leiomyosarcoma may be subdivided into two types: superficial (or dermal) and subcutaneous (or deep), according to the percentage of tumoral surface, which should be 90% in the former and less than 90% in the latter. The superficial leiomyosarcoma is a rare variant and does not exceed 3% of the total number of sarcomas. [1-4]

This division has an important prognostic value: the dermal variant is a locally aggressive tumor with frequent recurrence (30-50%), but it almost never metastasizes, while the subcutaneous variant is related to greater probability of metastasis and higher recurrence rates (50-70%). [5-8]

The dermal leiomyosarcoma may appear in any location: 50 to 75% of these tumors occur on the lower limbs, mainly on the thigh, 20 to 30% on upper limbs, 10-15% on the trunk and less than 5% on the face. [2,3,7,9]

The subcutaneous variant is clinically described

Received on 14.02.2010.
Approved by the Advisory Board and accepted for publication on 25.07.2010.
* Study carried out at the University Clinic of Dermatology, Santa Maria Hospital, Lisboa, Portugal.
Conflict of interest: None / *Conflito de interesse: Nenhum*
Financial funding: None / *Suporte financeiro: Nenhum*

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as a solitary, although well circumscribed, nodule of soft consistency; the longer axis usually measures more than 2 cm. It is rarely multinodular and the skin that covers the tumor may be pink, brownish or apparently normal. The dermal variant is usually adherent to the epidermis, its growth is slow and as a rule, the longer axis does not surpass 2 cm. Patients may refer pain when the tumors are pressed, pruritus, paresthesia and hemorrhage. [2,7,10]

Histologically, such tumors have a good or moderate degree of differentiation and are composed of bundles of fusiform smooth muscle cells, with vesicular nuclei with blunt extremities (the classical "cigar" shape) and eosinophilic cytoplasm. The leiomyosarcomas express desmin, vimentin and frequently actin.[8,9,11]

The treatment of choice for dermal or superficial leiomyosarcoma is radical surgical tumor excision, with free lateral margins measuring between 3 and 5 cm, including subcutaneous tissue reaching as deep as the fascia. Tumoral excision, without an adequate safety margin, may favor relapse with the involvement of deeper structures and a tendency to more aggressiveness than the primary tumor, due to the greater risk of metastasis. [4,7,12]

The leiomyosarcoma, when restricted to superficial tissues, is not an aggressive tumor and is associated with low mortality rate. [2,11] The local recurrence rate is around 30% for the dermal variant and 50% for the subcutaneous variant, and in around 30% of these patients metastases occur. [2,3]

CASE REPORT

A 57-year old male presented with an eight-month history of a hard, painless erythematous-violaceous tumor on the presternal region, with the longer axis measuring 3.1 x 2.8 cm (Figure 1).

The personal history points out to chronic alcoholism, tobacco use and toxicophilic habits, bipolar disorder and chronic hepatitis C. The patient also referred traumatism of the sternum region caused by a stab wound, approximately 18 years before.

The histological examination revealed a malignant dermal tumor with fusiform, pleomorphic cells, "cigar shaped" nuclei and innumerable atypical mitoses.

Immunomarking was positive for smooth muscle actin, desmin and vimentin and negative for S-100 protein and pan-cytokeratin (Figure 2).

Computerized tomography showed two small, solid, hypoechogenic nodular images close to the hepatic hilum, suggestive of adenopathy.

A radical surgical excision of the tumor was performed, with partial skin graft and good graft viability.

A Nuclear Magnetic Resonance of the liver was proposed to study the hepatic hilum nodules, which was not done because the patient abandoned our services.

DISCUSSION

Cutaneous soft tissue sarcomas are clinically not very expressive and encompass a great histological variety, which makes its diagnosis difficult. [10]

When investigating a malignant spindle cell cutaneous tumor the importance of utilization of an ample antibody panel (smooth muscle actin, desmin, vimentin, cytokeratins and S-100 protein) should be emphasized. [3,4,8,9,10] In this case, immunomarking allowed diagnosis of a leiomyosarcoma based on the expression of tumoral cells for smooth muscle actin, desmin and vimentin and negativity for the remaining markers, permitting to establish differential diagnosis with other neoplasias such as melanoma (whose cells positively mark S-100 protein), atypical fibroxanthoma (whose cells are habitually negative for smooth muscle actin), spindle cell carcinoma (positive cells for cytokeratin) or dermatofibrosarcoma protuberans (positive cells for CD34) (Chart 1). [13]

The dermal variant of leiomyosarcoma is rare and the therapeutic approach is not consensual. [7,8,11,14] As the main prognostic factors are tumor size, its distal location, the depth of tumoral invasion and the

Chart 1: Immunohistochemistry of malignant spindle cell cutaneous tumors

	Leiomyo-sarcoma	Melanoma	Atypical Fibroxanthoma	Spindle cell carcinoma	Dermato-fibrosarcoma protuberans
Smooth muscle actin	Positive	-/+	N	N	N
Desmin	+/-	N	N	N	N
Pan-cytokeratin	-/+	N	N	Positive	N
Vimentin	Positive	Positive	Positive	N	Positive
S-100	-/+	Positive	N	N	N
CD 34	-/+	N	-/+	N	Positive

Positive, expressed by the majority of tumoral cells; +/-, sometimes positive; -/+, rarely positive; N, negative.

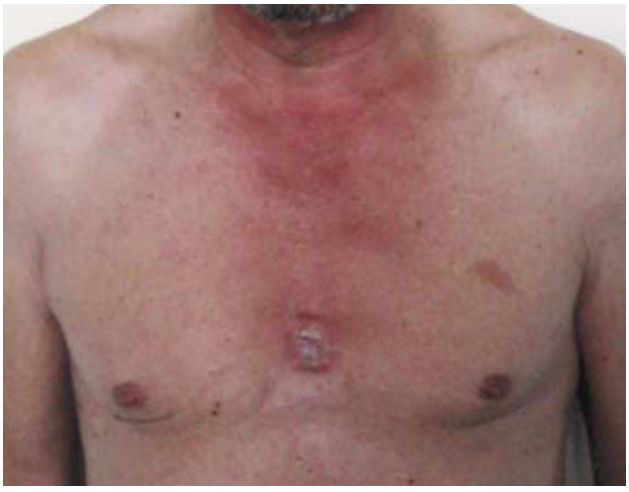

Figure 1: Erythematous-violaceous tumor, with central depression and parchment-like surface similar to scar tissue

degree of histopathological displasia. [7,8,14] In our case, it was a dermal tumor of small dimensions and little depth, which constitutes a good prognostic index. Similarly to what has been described in literature, our patient had a history of previous traumatism at the site of the tumor. Although local recurrence has been described in some cases, its potential for distant metastasis is much smaller. [1,2,3]

According to the best treatment referred in the literature, the patient was subjected to radical surgical exeresis of the lesion, without local recurrence up to the date of the last note taken during appointment. [5,7,14,15,16] As the patient did not choose to cooperate, we considered his stage classification incomplete, since it was impossible to carry out an imaging exam to clarify the nature of the liver "nodules" (even though they might be related to comorbidity and not to neoplasia), which may raise doubts regarding their prognostic value. ❑

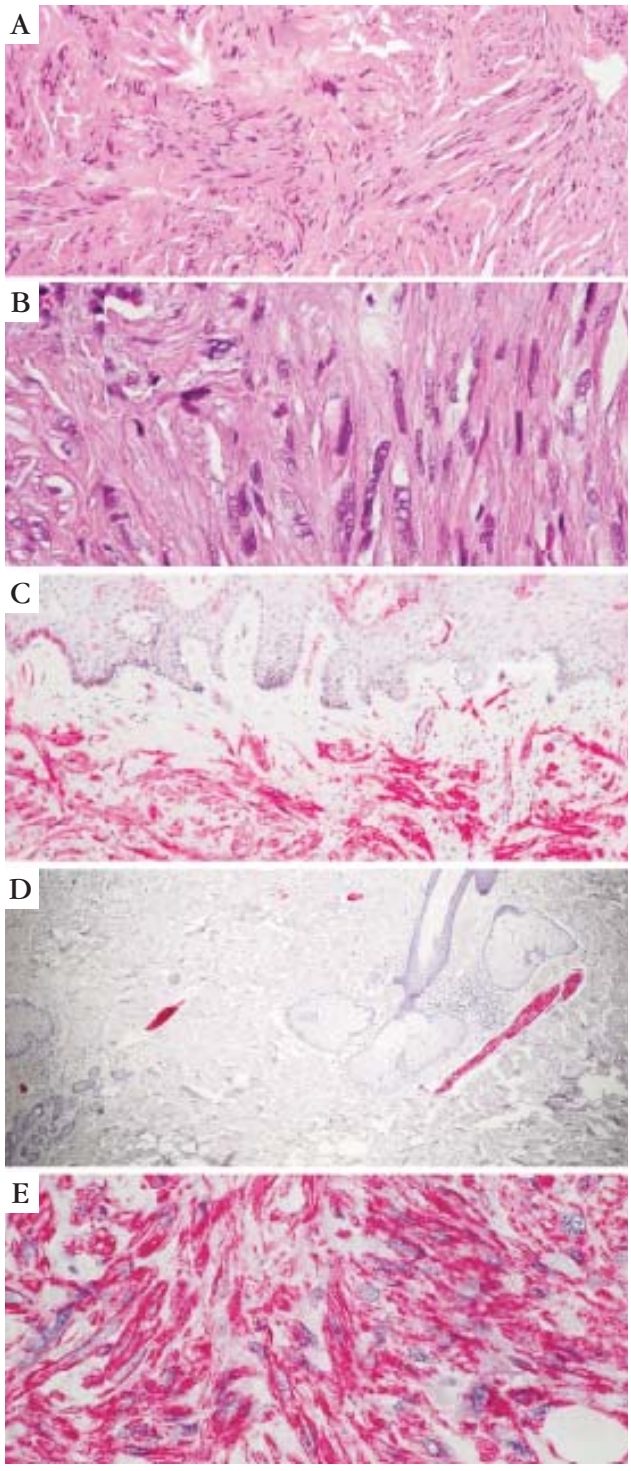

Figure 2: A. Histological aspects of Leiomyosarcoma - HE: Nuclear pleomorphism and eosinophilic cytoplasm of tumor cells; **B.** Histological aspects of Leiomyosarcoma – fusiform smooth muscle cells, with vesicular nuclei and blunt edges; "cigar"- contrasting with sharp, pointy extremities of fibroblast nuclei; **C.** Histological aspects of Leiomyosarcoma. Immunomarkers; smooth muscle actin; **D.** Histological aspects of Leiomyosarcoma. Immunomarker; desmin; **E.** Histological aspects of Leiomyosarcoma. Immunomarker; vimentin

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How to cite this article/*Como citar este artigo*: André MC, Antunes JV, Reis MD, Filipe PL, Almeida LMS. Cutaneous leiomyosarcoma on the trunk. An Bras Dermatol. 2011;86(5):999-1002.