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ORIGINAL ARTICLE

Clinical and Histopathologic Findings of Cutaneous Leiomyosarcoma: Correlation With Prognosis in 12 Patients☆

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KEYWORDS
Leiomyosarcoma;
Soft tissue;
Sarcoma;
Cutaneous metastasis;
Atypical intradermal smooth muscle neoplasm

Abstract

Introduction: Cutaneous leiomyosarcoma is a malignant neoplasm derived from smooth muscle cells. Its low incidence hampers the development of specific protocols for diagnosis and treatment.

Objectives: To describe the clinical and histopathologic characteristics of a series of primary and secondary cutaneous leiomyosarcomas and to determine how these characteristics correlate with prognosis.

Material and methods: We performed an observational, descriptive, retrospective study based on 17 cutaneous leiomyosarcomas in 12 patients diagnosed between January 1, 2000 and December 31, 2015. We recorded demographic data, clinical and histopathologic characteristics, outcome, and response to treatment.

Results: We included 5 men and 7 women, all aged more than 50 years at diagnosis. There were 4 cutaneous leiomyosarcomas (23%) in 4 patients, 2 subcutaneous leiomyosarcomas (11.5%) in 2 patients, and 11 skin metastases of leiomyosarcoma (65%) in 6 patients. The most frequently affected sites were the scalp (41%), lower limbs (17%), and trunk (17%). During follow-up, 50% of the cutaneous leiomyosarcomas recurred, 50% of the subcutaneous leiomyosarcomas presented distant metastases, and 83% of the patients with skin metastases of leiomyosarcoma died of their disease.

Limitations: Ours was a retrospective review of a small case series at a single center.

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Conclusions: Cutaneous leiomyosarcoma is an uncommon malignant neoplasm. Our approach to diagnosis and therapy must take into account the marked heterogeneity in the prognosis of the various subtypes.

PALABRAS CLAVE
Leiomiosarcoma;
Partes blandas;
Sarcoma;
Metástasis cutánea;
Neoplasia atípica intradérmica de músculo liso

Leiomiosarcoma cutáneo: características clínicas, histopatológicas y correlación pronóstica en 12 pacientes

Resumen
Introducción: El leiomiosarcoma de piel es una neoplasia maligna de estirpe muscular cuya baja incidencia dificulta el desarrollo de protocolos específicos de diagnóstico y manejo terapéutico.
Objetivos: Describir las características clínicas e histopatológicas de una serie de leiomiosarcomas cutáneos primarios y secundarios, junto con su correlación pronóstica.
Material y métodos: Se realizó un estudio retrospectivo, descriptivo y observacional. Se seleccionaron 17 casos de leiomiosarcoma cutáneo en 12 pacientes, diagnosticados entre el 1 de enero de 2000 y el 31 de diciembre de 2015. Se recogieron sus datos demográficos, características clínicas e histopatológicas, evolución y respuesta al tratamiento.
Resultados: Se reclutaron 5 varones y 7 mujeres, todos ellos mayores de 50 años al diagnóstico. Se recogieron 4 leiomiosarcomas dérmicos (4/17, 23%) en 4 pacientes, 2 leiomiosarcomas hipodérmicos (2/17, 11,5%) en 2 pacientes, y 11 metástasis cutáneas de leiomiosarcoma (11/17, 65%) en 6 pacientes. Las localizaciones más frecuentes fueron cuero cabelludo (7/17, 41%), miembros inferiores (3/17, 17%) y tronco (3/17, 17%). Durante el seguimiento, un 50% de leiomiosarcomas dérmicos recidivaron, un 50% de leiomiosarcomas hipodérmicos presentaron metástasis a distancia y 5/6 pacientes con metástasis cutáneas de leiomiosarcoma (83%) fallecieron a causa de su enfermedad.
Limitaciones: Este estudio es una revisión retrospectiva de una serie de casos de tamaño limitado en un centro único.
Conclusiones: El leiomiosarcoma cutáneo es una neoplasia maligna poco frecuente. A la hora de adoptar una actitud diagnóstico-terapéutica en estos pacientes debemos tener en cuenta la marcada heterogeneidad pronóstica entre sus diferentes subtipos.

Introduction

Skin leiomyosarcomas are rare malignant neoplasms that derive from smooth muscle tissue. They account for around 2% to 3% of all soft-tissue sarcomas in skin and 0.04% of all neoplasms. They are currently thought to arise de novo from smooth muscle, even though there have been reports of tumors that derived from leiomyomas.[1]

Leiomyosarcomas have traditionally been grouped in 3 large subtypes according to clinical and pathologic features that have prognostic implications: 1) cutaneous, or dermal forms; 2) subcutaneous, or hypodermal forms; and 3) secondary metastatic forms. The most important clinical and prognostic differences between the classes are summarized in Table 1.[2,3]

The origin of cutaneous leiomyosarcomas seems to be the pilorector muscle in the dermis. However, there have been reports of cases in which the tumor developed in the dartos, or its equivalent muscle in the vulva, or in the areola. This form is not usually aggressive but tends to recur locally after surgical excision with narrow margins. The risk of distant metastasis is considered low (5%–10%), and these tumors have therefore been called ''atypical intradermal smooth muscle neoplasms.''[4] Subcutaneous leiomyosarcomas arise in the smooth muscle fibers of the tunica media of arteries and veins. Unlike more superficial, or dermal forms, these leiomyosarcomas have high rates of local–regional recurrence and metastasis (30%–40%), presenting a poor prognosis. Finally, metastatic leiomyosarcoma of the skin is associated with a particularly poor prognosis. Metastasis usually derives from a primary leiomyosarcoma in the retroperitoneum, the uterus, or the subfascial soft tissue of the extremities.[5–7]Even though the World Health Organization's 2005 taxonomy distinguishes true cutaneous from subcutaneous forms[8] (the latter of which are included among soft-tissue tumors[9]), this distinction can become complicated in clinical practice because of overlap of histopathologic features with prognostic implications.[10] In addition, metastases of noncutaneous leiomyosarcomas to the skin simulate the features of primary subcutaneous leiomyosarcomas, a situation that makes metastasis a diagnosis of exclusion.[6]

Because prognosis is difficult in some cases of leiomyosarcoma in the skin, we analyzed features of cases diagnosed in our hospital in the past 15 years. We included all leiomyosarcomas regardless of origin of the neoplasm, that is, both primary tumors and metastatic disease.