CASE REPORT

Cutaneous Leiomyomatosis – Case Report and Literature Review

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Abstract

Cutaneous leiomyomas are uncommon benign smooth muscle tumors that comprise of three distinct types such as piloleiomyoma, angioliomyoma, and genital leiomyoma. The discovery in 2001 of the association between cutaneous leiomyomas, uterine leiomyomas in women, and an aggressive form of renal cell cancer (RCC) underscores the importance of accurate dermatologic diagnosis of CL so that appropriate cancer screening and counseling of patients and at-risk relatives can be instituted. The present case report will discuss the pathogenesis, clinical manifestations, diagnosis, and management of a piloleiomyoma.

Keywords: Cutaneous leiomyomas, piloleiomyoma.

INTRODUCTION

Cutaneous leiomyomas, also called piloleiomyomas, are uncommon smooth muscle tumors that are benign and derive from the muscles responsible for piloerection of hair follicles, the arrector pili muscles1. When Cutaneous leiomyomas occur in multiple numbers, they are most likely part of a cancer syndrome abbreviated...
CASE REPORT

Our patient was a 36-year-old male presenting with piloleiomyoma for approximately 15 years. He had undergone a surgical treatment in 2010, where the affected area of skin, which was at the time limited to the left pectoral and deltoid region, was excised and primarily skin grafted. The patient was now readmitted due to the progression of the tumors, with some adjacent to the skin graft, distally but with most of them encompassing the whole left upper limb (except for the left hand).

On clinical examination were identified multiple pink nodules ranging in size between 1 cm and 3 cm in diameter, on the dorsal side of the left upper limb from the radiocarpal joint to the level of the scapulohumeral joint, which infiltrate the dermis and are extremely painful to the touch (Figure 1). The patient also has painful episodes accompanied by sweating and anxiety that are slowly ameliorated by calcium-blocking medication.

Paraclinical evaluation showed normal blood tests, EKG showed sinus rhythm without signs of heart blocks or any other abnormalities and the heart rate was about 70 beats per minute. Thoracic radiographs showed free costophrenic sinuses, and no pleuropulmonary evolutionary lesions; heart and aorta were within normal limits; nodular opacities projected at soft parts in the left axillary region with maximum dimensions of 32/24 mm.

Surgical intervention: the nodular formations on the dorsal side of the left forearm were resected “en bloc”,...
Cutaneous leiomyomatosis – Case Report and Literature Review

ign tumors of smooth muscle cells that can be classified into 3 types depending on the place of origin. Piloleiomyomas are benign formations that arise from the arrector pili muscles, angioleiomyomas are formations that derive from smooth muscles within the vascular walls and dartoic leiomyomas are formations that originate in the smooth muscles of the genital skin. Of these, the most common form is Piloleiomyoma.

Approximately 75% of all extra-uterine leiomyomas are represented by Cutaneous leiomyomas. Their incidence does not appear to be related to race. Solitary cutaneous leiomyomas typically occur in the lower extremities, and mostly in adulthood, whereas multiple piloleiomyomas tend to occur anywhere on the body, with various distribution patterns and most commonly between the ages of 10 and 30 years.

When autosomal dominant syndromes are excluded, the incidence of piloleiomyoma appears to be equal in women and men, compared to angioleiomyomas, which are statistically more common in women than men (2:1). The exception to this is represented by the cavernous and venous subtypes; they are more common in men.

Clinically patients present with skin formations having the appearance of red or brown nodules or papules located mainly on the limbs and trunk. These benign tumors respond by pain to pressure and low temperatures, but multiple painful episodes are described without an illusory cause that requires medication to release it. The incriminated physiological mechanism is not completely elucidated, but there are multiple theories that justify the appearance of pain. Theories that have been proposed: the contraction of smooth muscle fibers, the invasion of cutaneous nerve threads or numerous nerve elements located inside the formations.

The evolution and extent of the lesions are variable even within the same family. Their size and number are gradually increasing, but without a constant rate. In some cases, patients suffer from an extensive disease with lesions covering large areas of the body and others have only a few imperceptible papules.

The histopathological examination revealed multiple firm, pinkish tumors, with diameters ranging from 0.2 cm to 5 cm, all with regular edges (Figure 4).

The microscopic examination revealed the proliferation of elongated cells, presenting a low degree of histological aggressiveness, which suggested a leiomyoma. As an immuno histochemical evaluation, the streptavidin-biotin/horseradish peroxidase complex method was used to detect antigens such as desmin, S100, SMA (smooth muscle actin) and CD34. The tumor cells were slightly positive for desmin and SMA stains, but negative for S100 and CD34. The histological pattern corresponded to a cutaneous leiomyoma with a very high probability of hereditary leiomyomatosis and renal cell cancer syndrome (HLRCC).

The patient’s evolution is favorable, at 7 days postoperatively the graft is integrated and the patient is discharged.

DISCUSSIONS

Cutaneous leiomyomas was first described in the literature by Rudolf Virchow in 1854. These are rare benign tumors of smooth muscle cells that can be classified into 3 types depending on the place of origin. Piloleiomyomas are benign formations that arise from the arrector pili muscles, angioleiomyomas are formations that derive from smooth muscles within the vascular walls and dartoic leiomyomas are formations that originate in the smooth muscles of the genital skin. Of these, the most common form is Piloleiomyoma.

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and imaging examinations throughout life. Also, these examinations have the role of early detection of a renal tumor formation being known the association of these two diseases (risk of 10-16%)².

CONCLUSIONS

Cutaneous leiomyomas are a rare condition that can be associated with other systemic manifestations and has clinical diagnostic difficulties due to the unusual form of presentation. The patient’s health is severely affected due to sporadic pain.

The treatment of choice that improves the patient’s quality of life is the surgical operation associated with calcium-blocking and alpha-blocking drug therapy. Recurrences are common, these patients require monitoring for the rest of their lives.

Compliance with ethics requirements: The authors declare no conflict of interest regarding this article. The authors declare that all the procedures and experiments of this study respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2008(5), as well as the national law. Informed consent was obtained from all the patients included in the study.

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