

Acroangiodermatitis (Pseudo-Kaposi sarcoma): a rarely-recognized condition. A case on the plantar aspect of the foot associated with chronic venous insufficiency *

Acroangiodermatite (Pseudossarcoma de Kaposi): uma condição raramente reconhecida. Um caso na planta do pé associado a insuficiência venosa crônica

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Abstract: Acroangiodermatitis, often known as pseudo-Kaposi sarcoma, is an uncommon angioproliferative entity related to chronic venous insufficiency, arteriovenous fistulae, paralysed limbs, amputation stumps, vascular syndromes and conditions associated with thrombosis. It presents most frequently as purple macules, papules or plaques in the dorsal aspects of the feet, especially the toes, and the malleoli. We report a case of acroangiodermatitis in the plantar aspect of the foot, misdiagnosed for two years, in which haematoxylin-eosin histopathological stain and immunolabeling with CD34 histochemistry examination were decisive for diagnosis. Patient had chronic venous insufficiency. The lesion responded well to the treatment with a combination of leg elevation and compression.

Keywords: Peripheral Vascular Diseases; Skin Diseases, Vascular; Vascular Malformations

Resumo: A acroangiodermatite ou pseudossarcoma de Kaposi é entidade angioproliferativa incomum relacionada a insuficiência venosa crônica, fístulas arteriovenosas, membros paralisados, cotos de amputação, síndromes vasculares e condições trombóticas. Apresenta-se, em geral, como máculas, pápulas ou placas purpúricas no dorso dos pés (especialmente hálux) e maléolos. Relatamos um caso de acroangiodermatite afetando a região plantar, por dois anos sem diagnóstico, para o qual a coloração histológica por hematoxilina-eosina e a marcação imuno-histoquímica com CD34 foram decisivas. A paciente tinha insuficiência venosa crônica e a lesão respondeu bem ao uso de bandagens elásticas e repouso com a perna elevada.

Palavras-chave: Dermatopatias Vasculares; Doenças Vasculares Periféricas; Malformações Vasculares

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INTRODUCTION

Acroangiodermatite é uma entidade incomum caracterizada por uma proliferação reativa de pequenos vasos sanguíneos em resposta a distúrbio circulatório crônico. Acroangiodermatitis is an uncommon condition characterized by a reactive proliferation of small blood vessels in response to chronic circulatory disturbance. It can occur due to various vascular conditions such as chronic venous insufficiency,¹ congenital or acquired (traumatic or iatrogenic) arteriovenous fistulas, arteriovenous fistulas and syndromes such as the Prader-Labhart-Willi syndrome and Klippel-Trenaunay syndrome.²⁻⁵ It is also described as being linked with amputation stumps, the use of poorly-fitting suction-type devices, or affecting patients with paralyzed extremities or congenital myopathy.⁶⁻⁹ Other conditions occasionally associated with the appearance of acroangiodermatitis lesions are thrombotic events that may be related to genetic changes.¹⁰

Most cases concern the lower extremities, except for those associated with hemodialysis arteriovenous fistulas, which can occur in the upper limbs.³ In the lower limbs, the lesions mainly appear on the dorsal aspects of the foot, particularly on the toe and ankle, including the malleolus, but occasionally also affects the anterior and posterior faces of a leg. It can however sometimes affect both lower extremities, depending on the nature of the circulatory disturbance.¹¹

The lesions initially appear as brown, red or violet spots which can evolve into papules or plaques and sometimes become verrucous or ulcerated.¹¹ They can be painful or not.¹

We describe a case of acroangiodermatitis on the sole of the foot, associated with chronic venous insufficiency of the corresponding leg, as evidenced by Doppler ultrasound examination, and with good response to treatment with the use of elastic compression bandages and leg elevation.

CASE REPORT

A 67-year-old white woman complained of a painful lesion on the left heel from which she had suffered for two years, with the lesion progressively increasing in size. A previous biopsy led to a diagnosis of chronic pigmented purpura with indication of ferric pigment in the tissues. The patient reported having been treated with oral cephalexin, azithromycin, amoxicillin + clavulanate, and topically with neomycin + bacitracin and potassium permanganate baths, with little improvement and subsequent worsening of the lesion. She suspended the use of oral medications due to "stomach ache".

The patient's past medical history revealed

nothing of importance, except for a snake bite on her left foot suffered in adolescence. Nothing of use for diagnosis emerged from her physiological, family and social history.

Physical examination was normal except for the presence of varicose veins in the left leg and a plaque lesion on the sole of the left foot (calcaneal region), measuring 8.0 x 7.5 cm in diameter, with a violet tone and a consistency similar to that of the surrounding skin, reaching the medial region of the calcaneus and the medial and posterior left ankle (Figure 1).

Blood count and evaluation of serum glucose, renal and liver function were all normal.

Histopathological examination of the lesion showed lobular proliferation of small capillaries in a loose stroma with extravasated erythrocytes and a sparse mononuclear cell infiltrate throughout the reticular dermis. Macrophages with hemosiderin pigment inside were detected. Interstitial fibroblast cells were slightly increased in number, mainly along thickened and sclerotic collagen bundles that were seen adjacent to an area of more pronounced vascular proliferation. The overlying epidermis showed hyperkeratosis and erasure of the epidermal ridges (Fig. 2). Immunohistochemical assay with CD34 antiserum showed strong positive reaction in the endothelial cells of the vascular channels, with no marking on the perivascular cells (Figure 3).

A Doppler ultrasound test detected superficial venous insufficiency of the left lower extremity. There was no failure of the deep venous system in the lower extremities or sonographic signs of thrombotic events, either in the superficial or deep venous systems. Segmental insufficiency was observed in the left saphenous vein.



FIGURE 1: Violet-colored plaque on the left leg, measuring 8.0 x 7.5 cm in diameter, reaching the medial calcaneal and medial and posterior regions of the left ankle

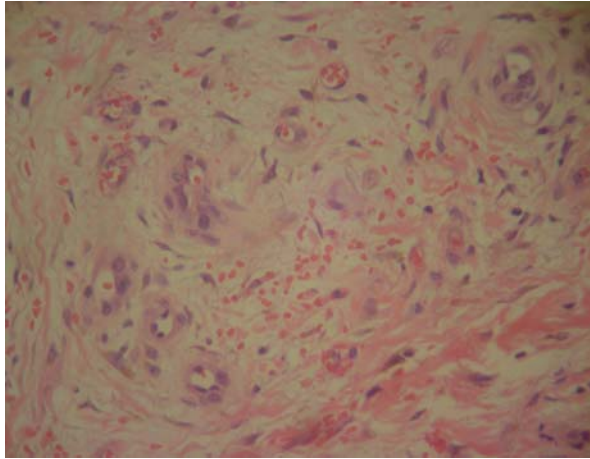


FIGURE 2: Proliferation of vessels, extravasated erythrocytes and hemosiderin pigment within macrophages (hematoxylin-eosin, 40X magnification)



FIGURE 4: Resolution of the lesion, with residual punctate hyperchromic macules on the medial left ankle, after continuous use of compression bandages and leg elevation

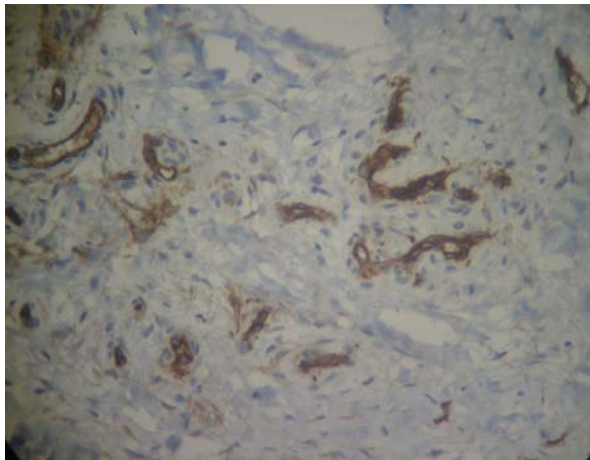


FIGURE 3: Immunohistochemistry: CD34 stained, with a strong positive reaction in the endothelial cells of proliferating vascular channels and no staining of perivascular cells (40X magnification)

nous vein (saphena magna) up to 16 cm from the sole of the foot, where it communicated with a competent perforating vein. The saphenofemoral junction was incompetent in the bottom left extremity.

The patient responded well to treatment with elastic compression bandages and leg elevation. 20 months after diagnosis the lesion was apparently resolved, leaving brownish pigmentation of the previously affected skin and some slight scaling. The painful symptoms had also disappeared (Figure 4).

DISCUSSION

Acroangiokeratitis can simulate a number of different clinical conditions such as Kaposi's sarcoma, lichen purple, pigmented purpura, lichen aureus, vas-

culitis, lichen simplex chronicus, actinic keratosis, basal cell carcinoma, stasis dermatitis, hemangioma, lymphangioma and lymphangiosarcoma.¹² Histopathologic examination shows proliferation of endothelial cells, newly-formed vessels with thick walls, often in a lobular pattern and surrounded by pericytes in the dermis. Extravasation of red blood cells, hemosiderin pigment deposition, dermal fibrosis, small thrombi in the lumen and superficial perivascular infiltrate of lymphocytes, histiocytes and occasional plasma cells are also found, and may resemble Kaposi's sarcoma.^{12,13} This, however, has vascular slits, proliferation of fusiform cells and atypical cells, and the vascular hyperplasia is independent of pre-existing vasculature.^{12,13} Other vascular tumors may be histologically confused with acroangiokeratitis such as hemangioendothelioma.¹¹

Immunohistochemical staining with CD34 antiserum helps to distinguish between acroangiokeratitis and Kaposi's sarcoma, because in the former an absence of perivascular CD34 is noted, unlike in the latter (CD34 staining on the endothelial cells as well as the perivascular spindle cells).^{14,15}

Given its relative rarity, acroangiokeratitis is often misdiagnosed clinically.^{11,12} The histopathological characteristics similar to Kaposi's sarcoma and other vascular tumors may also confuse professionals with little experience.¹¹⁻¹⁵ A faulty diagnosis can lead to inadequate treatment such as surgery, which often leads to complications.¹¹

Our patient remained undiagnosed for at least two years despite having been examined by several doctors. A previous biopsy for histopathology was wrongly interpreted.

The acroangiokeratitis lesions usually appear

on the dorsal aspects of the foot, particularly on the big toe, as well as on the ankles and legs.^{11,12} In the above case, the location of the lesions was different: on the sole of the foot in the region of the calcaneus. The histopathologic and immunohistochemical tests carried out by an experienced pathologist were essential for achieving a correct diagnosis. Doppler ultrasound was also useful for determining the cause of the injury as chronic venous insufficiency (the so-called 'Mali syndrome').¹

The treatment of this condition is barely discussed in the literature and depends on the nature of the underlying circulatory disturbance. In cases related to chronic venous insufficiency, the use of compression bandages can sometimes relieve symptoms.¹¹ The patient whose case we report here experienced long-term symptomatic relief after using a combination of compression and leg elevation, with apparent resolution of the lesion and only residual pigmentation of the area previously affected (Figure 4). □

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