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Case Report

Acroangiodermatitis of Mali secondary to chronic venous insufficiency

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ABSTRACT

Acroangiodermatitis is an unusual, benign angioproliferative condition that affects most commonly the lower extremities in patients with chronic venous insufficiency or other vascular anomalies. It is Indistinguishable clinically from aggressive conditions like Kaposi's sarcoma, making histopathologic examination helpful in its diagnosis. We report a case of acroangiodermatitis of Mali in a patient with superficial venous insufficiency in a young patient.

Key words: Acroangiodermatitis of Mali; Pseudo-Kaposi's sarcoma; Chronic Venous Insufficiency; Dermoscopy

INTRODUCTION

Pseudo Kaposi's sarcoma or acroangiodermatitis is a rare vasoproliferative entity most commonly seen in the lower extremities, related to chronic venous insufficiency and arteriovenous malformations.

Clinically it is a real differential diagnosis with kaposi sarcoma and requires histopathological examination for its diagnosis.

We report a case of acroangiodermatitis of Mali in a patient with superficial venous insufficiency.

CASE REPORT

A 35-year-old man with 4 year history of chronic venous insufficiency of his left lower extremity.

He presented with itchy, thick and diffuse pigmented lesions involving the left lower extremity appearing 2 years ago with loss of substance.

Clinical examination revealed edema and 7 ulcers of variable size with infiltrated base and warty surface located on anterior left leg, left medial and lateral malleolus (Fig 1).

Surrounding skin was xerotic and ochre dermatitis with adherent hemorrhagic crust were present (Fig. 2).

Dermoscopy showed papillomatous appearance, hemorrhagic suffusion, White rail lines, and red lacunae (Figs. 3a-3c).

Multiple varicose veins and varicosities were noted on his lower extremities.

All peripheral pulses were palpable.

A venous Doppler was performed which revealed superficial venous insufficiency of the left lower extremity.

Initially, we evoked verrucous sqamous cell carcinoma, venous ulcer and Pseudo Kaposi sarcoma.

A skin biopsy was performed showing hyperplastic epidermis and presence of a proliferation in the dermis of small vessels with dilated light and lobulated disposition (Figs. 4a-4c). This proliferation evolves on a fibrous and oedematous background of the dermis, which is strewed with many positive Perls heme pigments scattered over its entire height and even at the superficial hypodermis.

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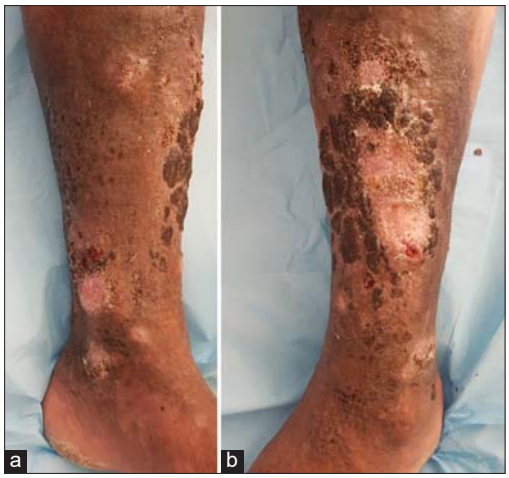

Figure 1: (a and b) Ulcers of variable size with infiltrated base and warty surface taking the left leg, left medial and lateral malleolus.

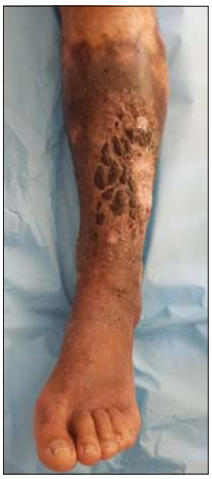

Figure 2: Ochre dermatitis with hemorrhagic crust.

Finally diagnosis of acroangiodermatitis of Mali was made.

The patient was prescribed compression stockings and then referred to the vascular surgery department for evaluation for surgical intervention.

DISCUSSION

Acroangiodermatitis (pseudo-Kaposi's sarcoma, AAD), was first described by Mali in 1965 [1]. It is a rare benign vascular proliferation most commonly seen in the lower extremities. Clinically, it is presents as violaceous patches, plaques or nodules that can progress to painful ulcerations [2].

It resembles to Kaposi sarcoma and requires histopathological examination for its diagnosis.

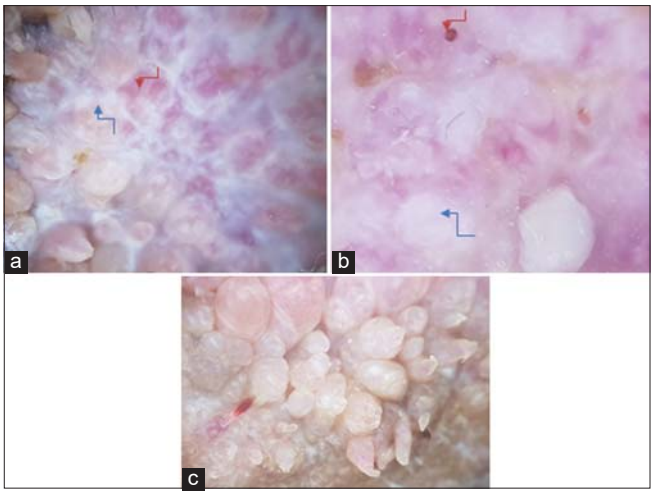

Figure 3: Dermoscopy showing: a: White rail lines (blue arrow) and red lacunae (red arrow). b: White structureless areas (blue arrow), and hemorrhagic suffusion (red arrow). c: Papillomatous appearance.

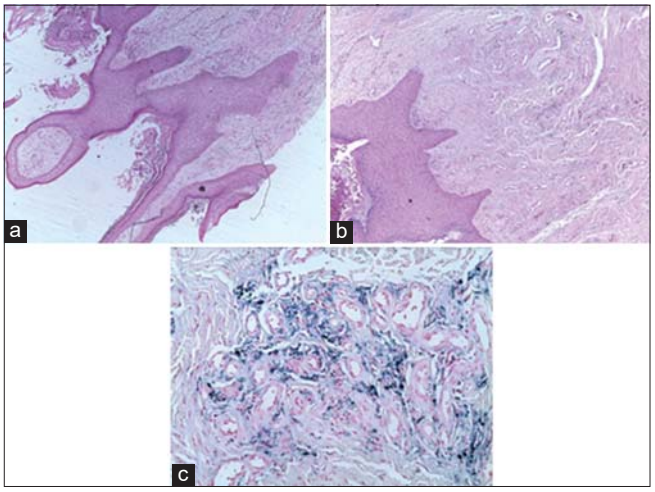

Figure 4: Histological findings (a): Hyperplastic epidermis (HES G × 50). (b): Proliferation in the dermis of small vessels with dilated light and lobulated disposition (HES G × 50). (c): Positive Perls heme pigments (Perls coloration G × 200).

There are four types of acroangiodermatitis: the Mali type, which is associated with chronic venous insufficiency, the Stewart-Bluefarb type accompanying congenital arteriovenous malformations, a type accompanying the first pregnancy, and a type accompanying arteriovenous shunts in patients with chronic kidney failure [3].

Several underlying conditions may be associated with AAD: venous insufficiency, hemodialysis, post-traumatic, congenital or acquired arterio-venous shunt, paralysis of the affected limb, residual limbs, intravenous drug abuse,

hereditary thrombophilia and vascular malformations (Klippel-Trenaunay syndrome) [4-6].

The Mali type, mainly affects elderly patients, usually bilateral with history of chronic venous insufficiency. It presents clinically as violaceous patches, macules that develop into papules and nodules [7].

Given its relative rarity, acroangiodermatitis is often misdiagnosed clinically. It can mimic several pathologies such as Kaposi's sarcoma, hemangioma, angiokeratomas, lymphangioma, lymphangiosarcoma, pigmented purpura, lichen aureus, vasculitis, lichen simplex chronicus and stasis dermatitis.

Even if it is not specific,dermoscopy seems useful for the diagnosis of acroangiodermatitis. It shows irregularly distributed polymorphic vessels, white structureless areas, red and blue lacunae [8].

Histopathological 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.

Immunohistochemical staining with CD34 anti-serum helps to distinguish between acroangiodermatitis and Kaposi's sarcoma, since in the first there is an absence of perivascular CD34, unlike the latter (CD34 staining on the endothelial cells as well as the perivascular spindle cells) [9].

Treatment is essentially based on the correction of the underlying pathology.

Various medical modalities of therapy have been tried with favorable results such as Oral erythromycin and dapsone [10].

Topical therapy with local corticosteroid preparations is often useful [11].

CONCLUSION

Acroangiodermatitis of Mali remains a rare pathology, the interest of our case lies in the occurrence in a young patient, the unilateral location and the interest of the histology in order to confirm the diagnosis.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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