

# Acroangiokeratitis of Mali: A rare vascular phenomenon

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## ABSTRACT

Acroangiokeratitis (synonym pseudo-Kaposi sarcoma) is an unusual, benign condition which clinically presents as purple-colored patches, plaques or nodules, mostly on the extensor surfaces of lower extremities in patients with chronic venous insufficiency and arteriovenous malformations. It resembles aggressive conditions like Kaposi's sarcoma and requires histopathological examination for its diagnosis. We report two such cases of acroangiokeratitis. Histopathology of both the cases showed dilated capillaries in the dermis with extravasated red blood corpuscles (RBCs), hemosiderin deposits, and hyperplastic granulation tissue. Both were treated with oral antibiotics and topical steroids. The ulcers showed a good response within 2 months of treatment.

**Key words:** Acroangiokeratitis of Mali, chronic venous insufficiency, pseudo-Kaposi's sarcoma

## INTRODUCTION

Acroangiokeratitis is a reactive angiodyplasia of cutaneous blood vessels associated with venous insufficiency or with vascular anomalies such as Klippel-Trenaunay syndrome or stump dermatosis in amputees. Exaggerated stasis dermatitis begins as violaceous macules and patches that gradually develop into papule, nodules, or indurated plaques often bilateral, and usually located on the lower extremities with edema. Although benign, it may mimic malignant conditions like Kaposi's sarcoma and therefore requires histopathological examination for its diagnosis and differentiation.

Here, we report two such cases presenting with nodulo-ulcerative lesions, with histological features of acroangiokeratitis, emphasizing the importance of histopathology in differentiating this from similarly presenting conditions.

## CASE REPORTS

### Case 1

A 41-year-old farmer in good health presented with a 4 year history of two raised lesions over

dorsum of the left foot. There was a 2 year history of recurrent bleeding and persistent swelling over the left ankle. He was a diagnosed case of varicose veins in both lower extremities for which he was treated with vein stripping and laser sclerotherapy, 2 years prior to presentation. On examination, there were two well-defined, soft, compressible hyperpigmented nodules of approximate dimensions 1 × 1 × 1 cm and 3 × 3 × 1.5 cm, respectively, over the dorsum of left foot. The larger nodule showed surface ulceration with a hemorrhagic crust [Figure 1]. There was no local tenderness or palpable thrill. The peripheral pulsations were felt. The saphenofemoral junction was competent on the Brodie-Trendelenburg test and perforators of the left leg were incompetent on the multiple tourniquet test. There were ill-defined hyperpigmented scaly, lichenified plaques on the volar aspect of the left foot, and left medial malleolus with minute hyperpigmented macules coalescing to form patches on inferomedial aspect of left leg. Examination of the right leg was within normal limits. A differential diagnosis of keratoacanthoma and Kaposi's sarcoma was considered in a patient of varicose veins.

### Case 2

A 26-year-old man presented with a single, pus

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discharging verrucous growth and swelling over the left ankle for 4 years. There was history of recurrent bleeding from the lesion. On examination, an ulcerated plaque with lobulated surface and serosanguinous discharge was found to be present on the lateral aspect of the left leg. The skin surrounding the ulcer showed dilated tortuous veins [Figure 2]. There was no local tenderness, palpable thrill, or bruit. The arterial pulsations were felt. The saphenofemoral junction was competent on the Brodie-Trendelenburg test but the lower perforator of the left leg was incompetent on the multiple tourniquet test. Examination of the right leg revealed no abnormality.

A differential diagnosis of Kaposi's sarcoma and chromoblastomycosis was considered.

In both the cases, hematological and biochemical investigations were normal. Serological testing for HIV was negative. A lesional skin biopsy from both cases showed hyperkeratotic stratum corneum, dilated capillaries with plump endothelial cells, extravasated RBCs, and hemosiderin, surrounded by hyperplastic granulation tissue with perivascular mononuclear infiltrate in the upper dermis [Figure 3a, b]. No vascular slits were seen. The hemosiderin deposits were confirmed on Perl's stain.

In the first case, venous Doppler showed multiple varicosities along course of the great saphenous vein with four incompetent perforators (mid thigh, upper leg, mid leg and lower leg) in the left lower extremity. The second case showed normal venous flow in left iliac vein with dilated left popliteal vein and patent venous valves. The saphenofemoral junction was competent in both the cases.

On the basis of the history, examination and biopsy findings, a final diagnosis of acroangiokeratitis of Mali was made in both the cases.

The first case was treated with oral erythromycin, topical clobetasol propionate (0.05%) cream, oral calcium dobesilate 500 mg twice daily, and emollients for 3 weeks. In addition, he was advised leg elevation and compression therapy with an elastocrepe bandage. After 3 weeks, calcium dobesilate was continued along with compression therapy. The ulcers responded well and healed with hyperpigmentation in 2 months but the nodular lesions persisted.

The second case was treated with oral doxycycline, oral metronidazole, topical clobetasol propionate (0.05%) cream, oral calcium dobesilate 500 mg twice daily, and emollients initially for 15 days. He was advised leg elevation and compression with elastocrepe bandage. The ulcers responded well and started to heal with scar formation. Antibiotics were discontinued and the calcium dobesilate and compression therapy were continued. The ulcers healed completely with scarring and dyspigmentation over the next 4 months [Figure 4].

## DISCUSSION

Acroangiokeratitis (Synonyms: pseudo-Kaposi's sarcoma, acroangiokeratitis of Mali-Kuiper, gravitational purpura, stasis purpura) was first coined by Mali in 1965.<sup>[1]</sup> It is a proliferation of pre-existing vasculature seen in venous hypertension, arteriovenous malformation, or acquired iatrogenic arteriovenous (AV) fistula. A variety of vascular conditions giving rise to this venous pathology include Klippel-Trenaunay syndrome, stump dermatosis in amputees, paralyzed limb, and intravenous drug abuse. It is rarely reported in hereditary coagulation defects (carrier of the thrombophilic 20210A mutation in the prothrombin gene and homozygous activated protein C resistance).<sup>[2,3]</sup> Most of the cases of acroangiokeratitis have been associated with some signs of venous insufficiency however Barbar *et al.* reported no venous insufficiency in 9 of their 10 cases of acroangiokeratitis, and similar spontaneous acroangiokeratitis in a young woman has been reported.<sup>[4,5]</sup> Our first case had venous insufficiency but in the second case, though there was normal Doppler study, the patient had localized dilated and tortuous veins around the lesion suggesting some degree of venous insufficiency as an etiologic factor in the development of the lesions.

There are various variants of acroangiokeratitis:

- Stewart-Bluefarb syndrome is a congenital arteriovenous malformation of the lower leg with multiple arteriovenous shunts. It begins early in life unilaterally over lower extremities with painful purple papules and macules, which may ulcerate. The affected limb may show increased warmth with varicose veins and a palpable thrill can be felt.<sup>[1]</sup>
- Mali type is an exaggerated stasis dermatitis seen in elderly patients, usually bilateral with chronic venous insufficiency on dorsum of feet, hallux,



Figure 1: Hyperpigmented nodule with surface ulceration and hemorrhagic crust (Case 1)



Figure 2: Ulcerated plaque with lobulated surface on lateral aspect of the left leg (Case 2)

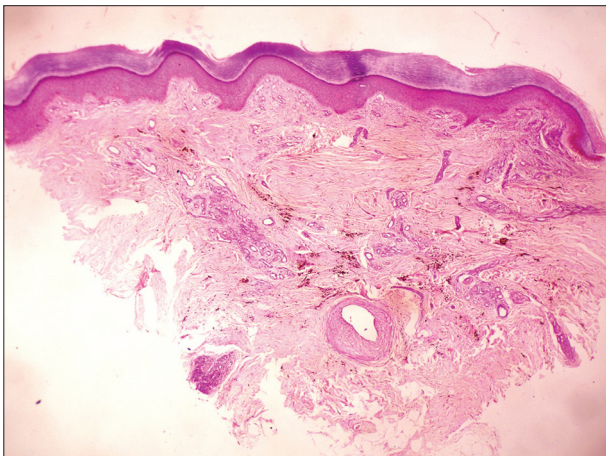


Figure 3a: Hyperkeratotic stratum corneum with proliferation of capillaries in upper and mid-dermis (H and E, x40)

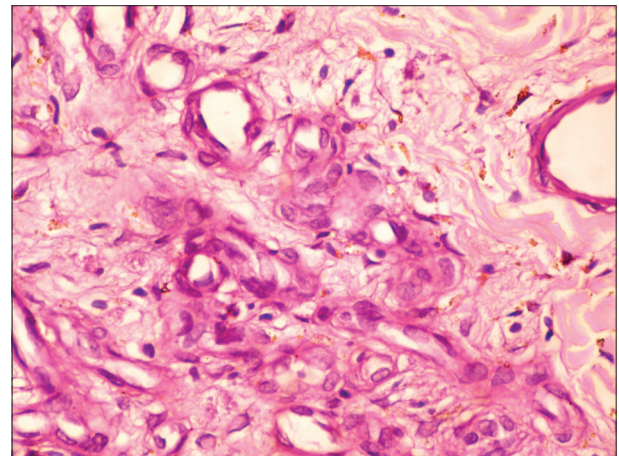


Figure 3b: Capillaries showing plump endothelial cells and perivascular mononuclear infiltrate (H and E, x400)



Figure 4: Healing of ulcer at 2 months of treatment (Case 2)

and second toe or on medial aspect of lower legs. The lesions begin as violaceous macules and

patches that develop slowly into soft, nontender, red to purple papules and nodules or indurated plaques.<sup>[1]</sup> Both our patients had this variant of acroangiokeratosis.

- Acroangiokeratosis in first pregnancy appears as gravity purpura (Dermite ocre of Favre) on lower legs over site of venous varicosities extending to the dorsa of feet and toes.<sup>[6]</sup>
- Angiodermatosis occurring after placement of the arteriovenous shunt for hemodialysis in chronic renal failure.<sup>[6]</sup>

Although precise etiology is unknown, it is postulated that severe chronic venous stasis with insufficiency of the calf muscle pump elevates the capillary pressure and leads to chronic tissue hypoxia from chronic edema which induces neovascularisation and fibroblast proliferation.<sup>[6,7]</sup>



**Table 1: Differentiating features between Kaposi's sarcoma and acroangiokeratitis on histopathology**

Differentiating features	Acroangiokeratitis of Mali	Kaposi's sarcoma
Histopathologically <sup>[3,4]</sup>	Small dilated vessels lined by plump endothelial cells with hyperplasia of pre-existing vasculature	Slit-like spaces and spindle cell proliferation. Vascular proliferation is independent of the existing vessels
Periodic acid Schiff (PAS) positivity for the vessels <sup>[3]</sup>	Present	Absent
Factor VIII-associated antigen in the endothelial cells <sup>[1,8]</sup>	Present	Absent
Immuno-labelling for the CD34 antigen <sup>[1,6]</sup>	Positivity seen on endothelial cells of hyperplastic vessels	Positivity seen on both endothelial cells and the characteristic spindle-shaped, perivascular cells
Dermal fibrosis, RBC extravasation, and hemosiderin deposition <sup>[4]</sup>	Present	Present

The histopathologic differentials for acroangiokeratitis are Kaposi's sarcoma [Table 1], pigmented purpuric dermatitis (PPD), vasculitis, and stasis dermatitis.<sup>[4]</sup>

PPD has dilated capillaries with the absence of plump endothelial cells, dermal fibrosis, or eosinophils in dermal infiltrates as seen in acroangiokeratitis. Fibrinoid necrosis of the vessels is seen in vasculitis as opposed to acroangiokeratitis. Stasis dermatitis differs from acroangiokeratitis by epidermal changes (parakeratosis and spongiosis), deeper dermal involvement, and greater hemosiderin deposition.<sup>[4]</sup>

Treatment of acroangiokeratitis involves correction of the underlying vascular pathology with mainstay of therapy being compression stockings or a compression pump for venous stasis and local wound care for ulcers. Various medical modalities of therapy have been tried with favorable results but options are limited. Oral erythromycin 500 mg four times a day or dapsone 50 mg twice a day for 3 months in combination with compression therapy has been tried with good results.<sup>[3]</sup> Topical therapy with local corticosteroid preparations is often useful.<sup>[6]</sup> Underlying vascular pathology like arteriovenous malformations can be surgically managed with elimination of the shunts and embolization for small fistulae.<sup>[1,2,9]</sup> Laser ablation, such as pulsed-dye laser, may clear some individual lesions.<sup>[2]</sup>

We would like to emphasize the importance of histopathology in distinguishing this benign condition from similar looking malignant conditions, even in the absence of predisposing factors such as varicose veins.

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