CASE REPORT

Acroangiodermatitis of Mali: A Rare Case Report

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ABSTRACT

Acroangiodermatitis of Mali also known as pseudo-Kaposi sarcoma is an unusual, benign condition. It is clinically presents as purple-colored patches, which can be plaques or nodules. It is located mostly on the extensor surface of lower extremities and seen most commonly in patients with chronic venous insufficiency and arteriovenous malformation. It looks like aggressive conditions like Kaposi's sarcoma. Hence, histopathological examination is required for its diagnosis. Herein, we report a patient who presented with a chronic history of purple-colored papules and plaques over both legs which was later histopathologically confirmed to be case of acroangiodermatitis of Mali.

Key words: Acroangiodermatitis of mali, Arteriovenous malformation, Venous insufficiency

INTRODUCTION

Acroangiodermatitis of Mali is a reactive angiodysplasia of cutaneous blood vessels. It is generally associated with venous insufficiency or with vascular anomalies like klippel–trenaunay syndrome or in amputation stump dermatosis. Chronic history of stasis dermatitis started as violaceous macules and patches. These lesions progress into papules, nodules, and indurated plaques. The lesions are usually located on the lower extremities, mostly bilateral and presented with edema. It is a benign condition but it may resemble other malignant conditions like Kaposi's sarcoma [Table 1]. Hence, histopathological examination is essential for its diagnosis and differentiation.

CASE REPORT

A 36-year-old male presented with multiple dark-colored small lesions over both legs since 1 year associated with itching. There was no other significant history and general examination was unremarkable. Cutaneous examination revealed multiple, well defined, hyperpigmented to violaceous papules with few papules coalescence to form plaques over anterior aspect of bilateral legs [Figure 1]. Similar lesions are not seen elsewhere on the body. Dermoscopic examination showed radially arranged whitish lines on a brownish or reddish background similar to of prurigo nodularis "star burst" pattern [Figure 2]. Histopathological findings showed moderately dense superficial perivascular infiltrate with focal atrophy of overlying epidermis. Within the thickened papillary dermis, there is an increased number of thick-walled capillaries in clustered pattern. These thick-walled capillaries are surrounded by mucin and a moderate perivascular lymphocytic infiltrate with eosinophils occasionally. Minimal amount of hemosiderin deposits seen around vessels [Figure 3].

DISCUSSION

Acroangiodermatitis (other names: pseudo-Kaposi's sarcoma, acroangiodermatitis of Mali-Kuiper, gravitational purpura, stasis purpura) was first coined by Mali in 1965.[1] The condition occurs due to proliferation of pre-existing vasculature which is seen in venous insufficiency, arteriovenous malformation, or acquired iatrogenic arteriovenous fistula. In a study by Mehta *et al*., they reported 2 cases of acroangiodermatitis one of which had venous insufficiency and other had a normal Doppler study.[2]

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Table 1 : The histopathologic differentials for acroangiodermatitis are Kaposi's sarcoma

Differentiating features	Acroangiodermatitis of Mali	Kaposi's sarcoma
Histopathologically	Small dilated vessels lined by plump endothelial cells with hyperplasia of pre-existing vasculature	Slit-like spaces and spindle cell proliferation are independent of the existing vasculature
Periodic Acid Schiff positivity for the vessels	Present	Absent
Factor VIII-associated antigen in the endothelial cells	Present	Absent
Immunolabeling for the CD34 antigen	Positively seen on endothelial cells of hyperplastic vessels	Positively seen on both endothelial cells and the characteristic spindle-shaped perivascular cells
Dermal fibrosis, RBC extravasation, hemosiderin	Present	Present

RBC: Red blood cell

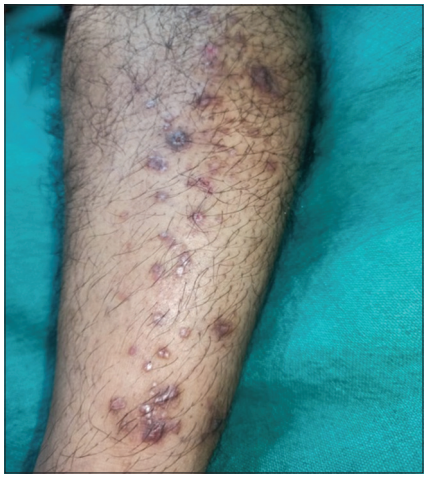

Figure 1: Clinical image: Multiple hyperpigmented to violaceous papules and plaques present over left leg

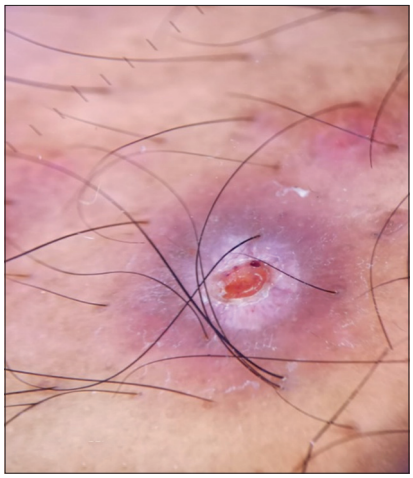

Figure 2: Dermoscopic images: ×10: "Star Burst" pattern

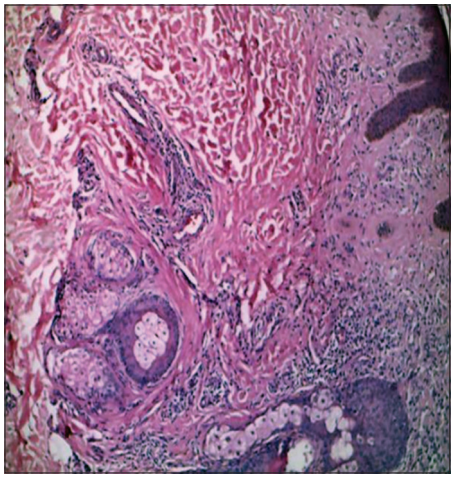

Figure 3: Histopathological image: ×40

Various variants of acroangiodermatitis are:

- Stewart-Bluefarb syndrome: It is a congenital arteriovenous malformation of the lower limb with multiple arteriovenous shunts. It begins in early life and presents with painful purple papules and macules, which can ulcerate. The condition is unilateral and presents over lower extremities.[1]
- Mali type: It is severe form of stasis dermatitis seen in elderly patients. The condition is generally bilateral. It is associated with chronic venous insufficiency. Located on dorsum of feet, hallux, and second toe or on medial aspect of lower limbs. It starts as violaceous macules and patches which gradually progress into soft, non-tender, red-to-purple papules and nodules or indurated plaques.[1]
- Acroangiodermatitis in first pregnancy: It is a gravity purpura also known as Dermite ocre of Favre. Located on lower legs over the site of varicosities of veins which may extend to the dorsa of feet and toes.[3]

- Angiodermatitis occurring after placement of the arteriovenous shunt: It is seen in patients of chronic renal failure and on hemodialysis. Angiodermatitis develops in these patients after placement of the arteriovenous shunt for hemodialysis.[3]

The exact etiology is not known, but it is suggested that severe chronic venous stasis with insufficiency of the calf muscle pump increases the capillary pressure and it leads chronic edema due to which there is chronic tissue hypoxia which causes neovascularization and fibroblast proliferation.[3]

Treatment of acroangiodermatitis contains first correction of the underlying vascular pathology with the help of compression stockings or a compression pump for venous stasis and it is the mainstay of therapy with local wound care for ulcers with daily dressing. Oral erythromycin 500 mg 4 times a day or dapsone 50 mg twice a day for 3 months in combination with compression stocking and pump therapy has been tried with good results.[4] Topical therapy with corticosteroid is also often helpful.[3] Laser ablation, such as pulsed-dye laser, may be useful to clear some localized lesions.[2]

CONCLUSION

In clinical practice, acroangiodermatitis of Mali can be presented in such features which may looks like aggressive malignant conditions like Kaposi's sarcoma. Hence, histopathology should be considered as a gold standard in the differentiation of such similar presenting conditions.

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