# Acroangiodermatitis presenting as unilateral hypertrophic verrucous plaques

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## Abstract

Acroangiodermatitis (AAD) is a rare vasoproliferative disorder often involving the extremities that has been classified into two variants. Mali-type AAD is more common and associated with chronic venous stasis. Stewart-Bluefarb syndrome, the other variant, is associated with underlying arteriovenous abnormalities. Mali-type AAD is a relatively benign diagnosis but it may mimic more harmful etiologies such as Kaposi sarcoma both clinically and histologically. A 67-year-old woman with a history of varicose veins, deep vein thrombosis, stroke, and obesity presented to our outpatient clinic with verrucous red-brown papules and plaques on her right lower extremity worsening for three years. Biopsy was consistent with a diagnosis of Mali-type AAD. Providers should be aware of AAD and its variants to accurately differentiate it from more harmful entities.

*Keywords: acroangiodermatitis, pseudo-Kaposi sarcoma, stasis dermatitis, venous insufficiency*

## Introduction

Acroangiodermatitis (AAD), also known as pseudo-Kaposi sarcoma, is a benign vascular proliferation often involving the lower extremities. Acroangiodermatitis typically presents clinically as purple papules and plaques on the lower extremities and dorsal feet/toes, although verrucous plaques may also be seen. Acroangiodermatitis is a reactive inflammatory disorder which has been classified into two variants: Mali-type AAD and Stewart-Bluefarb syndrome. Mali-type AAD is more common and associated with venous stasis of the lower extremities and severe chronic venous insufficiency. Stewart-Bluefarb syndrome is associated with arteriovenous fistulae and arteriovenous malformations. Because of the systemic nature of venous stasis, Mali-type AAD is classically bilateral [1]. Unilateral presentations can broaden the overall differential diagnosis and may also necessitate distinction from Stewart-Bluefarb syndrome, which is more often unilateral [1]. Herein, we present a patient with a unilateral presentation of Mali-type AAD secondary to chronic venous stasis and thromboembolic phenomena.

## Case Synopsis

A 67-year-old woman presented to the outpatient clinic at our academic institution with multiple hypertrophic verrucous purple-brown plaques with neighboring red-brown papules coalescing into plaques on her right lower leg (**Figure 1**). The lesions had onset three years previously and were initially small, progressively growing in number, and darkening since that time. The affected area was painful but the patient denied itching or bleeding. Her past medical history included varicose veins for 10+ years, a reported deep vein thrombosis (DVT) in the right lower extremity, a stroke in 2011, restless leg syndrome, hypertension, and hyperlipidemia. She had a BMI of 38kg/m$^2$. There were no similar lesions present on the left lower extremity. Bilateral significant pitting edema was present. All her toenails had yellowing and subungual dystrophy

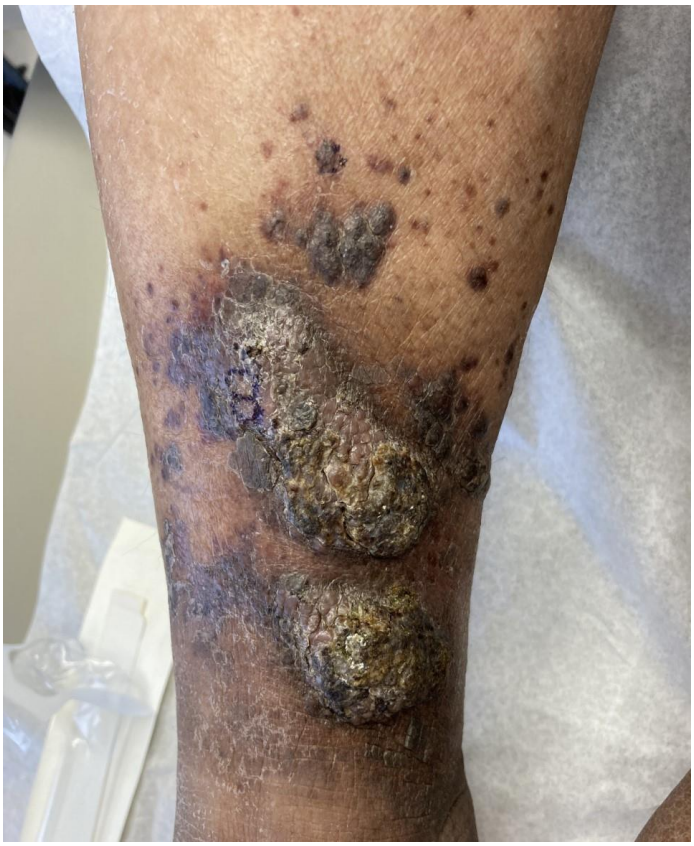

**Figure 1**. *Verrucous violaceous-brown plaques on the right lower leg with surrounding red-brown papules coalescing into plaques.*

present bilaterally consistent with onychomycosis and onychogryphosis. At this visit, two punch biopsies were performed including hematoxylin and eosin staining and tissue culture for aerobic bacteria, acid-fast bacillus, and fungal organisms. Pan-sensitive *Staphylococcus aureus* only was grown. On pathology, nodular angioplasia was seen with tufts of small-caliber vascular channels in the papillary and upper reticular dermis (**Figure 2**). The endothelial cells were benign-appearing, without cytologic atypia. Extravasated erythrocytes were observed along with hemosiderin and a perivascular lymphocytic infiltrate. No slit-like vascular channels were present. Periodic acid-Schiff stains were negative. A multi-institution dermatopathology consensus conference reviewed the case; no dermatopathologist present favored Kaposi sarcoma and a diagnosis of AAD was made. The patient was referred to vascular surgery. A venous duplex ultrasound performed revealed significant reflux in bilateral great saphenous veins and multiple varicose veins. The patient was advised to wear compression stockings. She was also advised that non-thermal ablation of her right great saphenous vein might offer further alleviation of her symptoms. At the last visit the patient reported she would consider the procedure.

## Case Discussion

In 1965 Mali et al. documented 18 cases of purple plaques on extensor aspects of the lower extremities attributed to chronic venous insufficiency and histologically resembling Kaposi sarcoma [2]. In 1967 Bluefarb and Adams and Stewart reported a similar condition associated with congenital arteriovenous malformations [3,4]. In 1974 Earhart et al. proposed the name pseudo-Kaposi sarcoma [5]. Acroangiodermatitis has been reported in patients

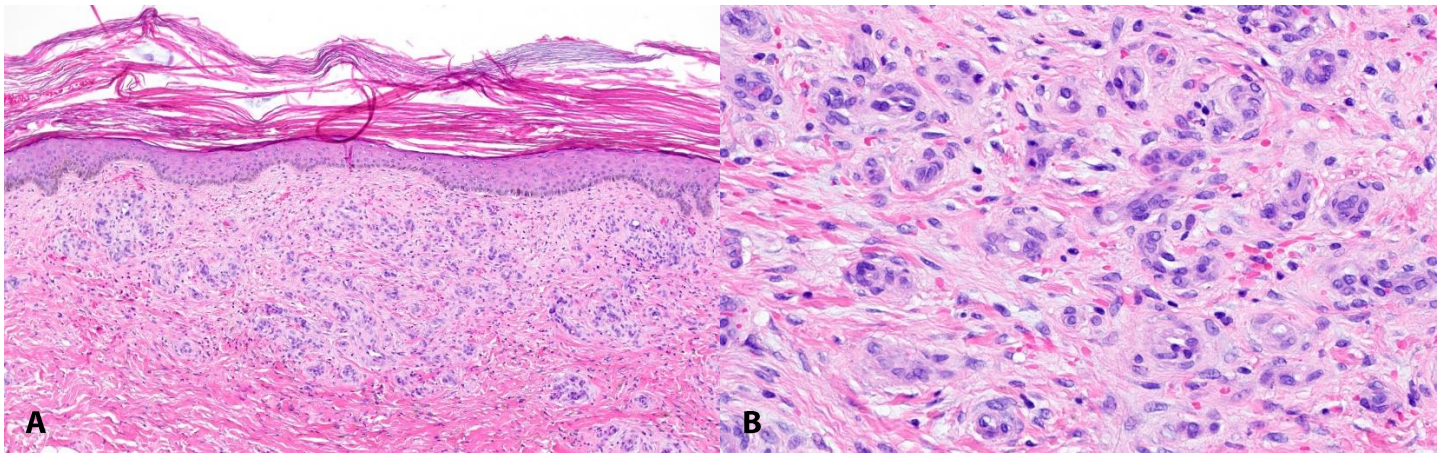


**Figure 2**. ***A)*** *H&E histopathology. Lobular angioplasia present in the dermis with perivascular lymphocytic infiltrate, 100×.* ***B)*** *Plump endothelial cells lining dermal vessels without the presence of slit-like vascular channels or cytologic atypia. Interspersed extravasated erythrocytes and hemosiderin deposits present, 400×.*

with many conditions including chronic venous insufficiency, paralyzed extremities, suction socket lower limb prosthesis, minor trauma, congenital arteriovenous malformation, and acquired arteriovenous fistula [1].

Mali-type AAD is more common in elderly patients. It is associated with chronic venous stasis and venous insufficiency. The characteristic lesions are slowly evolving, red-violaceous macules, papules, or plaques which may ulcerate or develop a verrucous quality. Lesions typically occur on the medial aspect of the lower leg, as occurred in our patient, the dorsal foot, hallux, heel, and/or toes [1].

In contrast to Mali-type AAD which usually involves bilateral extremities, Stewart-Bluefarb syndrome often involves a single extremity, as occurred in our patient. However, Stewart-Bluefarb syndrome often occurs in young adults with arteriovenous fistulae or arteriovenous malformations or in amputees [1]. Our patient had none of these predisposing factors.

Our patient did have an obese body habitus and a reported history of DVT. Acroangiodermatitis Mali-type has been documented in multiple patients with a variety of hypercoagulable states including heterozygous *prothrombin* G20210A mutation, homozygous activated protein C resistance, and protein C deficiency [6]. Given our patient's history of stroke and DVT in her affected extremity, this may have predisposed her right lower extremity to AAD, yielding her unilateral presentation.

The etiology of AAD has yet to be completely elucidated. In Mali-type AAD it is hypothesized that the chronic edema patients experience reduces perfusion. This lack of perfusion leads to fibroblast proliferation, hypertrophy, inflammatory skin change, and neovascularization [1]. On the other hand, in Stewart-Bluefarb syndrome, instead of reduced perfusion, a high perfusion, high oxygen saturation state occurs. This leads to reactive endothelial hyperplasia and fibroblast proliferation.

Discerning Mali-type AAD from Stewart-Bluefarb syndrome can be accomplished via duplex ultrasonography [1]. Ultrasound may be indicated in patients with clinical suspicion for Stewart-Bluefarb syndrome, such as young patients without signs of venous insufficiency or a warm lesion with a pulse or thrill associated with it.

Outside of discerning between Mali-type AAD and Stewart-Bluefarb syndrome, the differential diagnosis is broad. From a clinical perspective the differential includes stasis dermatitis, lichen planus, pigmented purpura, and Kaposi Sarcoma. Potential entities which may require more histologic examination include glomeruloid angioendotheliomatosis, angiopericytomatosis, diffuse dermal angiomatosis, intravascular histiocytosis, reactive angioendotheliomatosis, vasculitis, as well as Kaposi sarcoma. In cases with unilateral and verrucous presentations, as in our patient, the differential diagnosis broadens to infectious etiologies such as deep fungal infection and malignancy including squamous cell carcinoma or angiosarcoma.

Histopathologically, AAD will often show a proliferation of papillary dermal thick-walled capillaries and venules lined by plump endothelial cells. Edema within the papillary dermis may be present and the vessels are often organized in a lobular pattern. There may be an increased amount of fibroblasts present with fibrosis or sclerosing panniculitis present in older lesions [7]. Venules can become hypertrophic and tortuous. A superficial or perivascular infiltrate composed of lymphocytes, histiocytes, and eosinophils may be present [1]. Deposits of hemosiderin and extravasated erythrocytes may also be seen [7]. Many of these features are also common to Kaposi sarcoma. However, when compared to Kaposi sarcoma, the proliferation of jagged vascular channels with slit-like lumina encompassing pre-existing capillaries is not seen in AAD [8]. Acroangiodermatitis lacks cytologic atypia seen in Kaposi sarcoma. Kaposi sarcoma often also spares the papillary dermis early in its evolution [8]. With regard to immunohistochemistry, CD34 positivity is detected in both endothelial cells and perivascular cells of Kaposi sarcoma, compared to AAD where there is an absence of CD34 expression in the perivascular cells [8]. However, the gold standard for differentiation from Kaposi sarcoma is testing for human herpesvirus 8 [8]. Human herpesvirus 8 may be

immunohistochemically seen in the nuclei of proliferating cells in true Kaposi sarcoma but will be absent in AAD.

Management of AAD is directed at normalizing the underlying elevated venous pressure and managing complications. Conservative therapy begins with compression stockings and garments. Local wound care is also important especially in cases of ulceration. Topical corticosteroids such as clobetasol propionate 0.05% have demonstrated some success when combined with oral antibiotics [9]. Oral antibiotic regimens have included erythromycin and dapsone when secondary infection is present [9,10]. Other treatments which have been explored include a heparan sulfate mimetic which functions as a tissue matrix-protective [11]. However, definitively halting the progression of AAD is difficult without addressing the underlying cause. It is important to emphasize the role of compression, elevation, physical activity, and other lifestyle alterations to patients seeking relief. Treating underlying venous insufficiency and arteriovenous malformations may also be accomplished with surgical interventions, selective embolization, sclerotherapy, or endovenous ablation [1].

## Conclusion

Acroangiodermatitis is a rare vasoproliferative disorder occurring in the setting of chronic venous insufficiency or arteriovenous abnormalities. It is important to differentiate this relatively benign disorder from more harmful entities such as Kaposi sarcoma. This can be accomplished with histopathology and immunohistochemical testing correlated with clinical presentation. In patients with possible underlying arteriovenous abnormalities or no signs of venous insufficiency, duplex ultrasonography may provide diagnostic utility. Although medical therapies have been used successfully, evidence is limited. The mainstay of improving symptoms for patients will stem from compression, elevation, proper wound care, and lifestyle modifications in inactive or obese patients. Surgical or more invasive intervention may provide relief if more conservative measures are not sufficiently effective.

## Potential conflicts of interest

The authors declare no conflicts of interest.

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