# Acroangiodermatitis of Mali and Stewart-Bluefarb Syndrome

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PRACTICE **POINTS**

- Acroangiodermatitis (AAD) may mimic Kaposi sarcoma clinically and histopathologically. A human herpesvirus 8 stain is helpful to differentiate these two entities.
- Diagnosis of AAD should prompt investigation of an underlying arteriovenous malformation, as the disease may have systemic consequences such as congestive heart failure.

Acroangiodermatitis (AAD), also known as pseudo-Kaposi sarcoma, is a rare benign vascular proliferation mainly of the extremities. It is characterized by violaceous patches or plaques resembling Kaposi sarcoma. The term *pseudo-Kaposi sarcoma* encompasses 2 variants of acroangiodermatitis: Mali type and Stewart-Bluefarb syndrome (SBS). Mali-type AAD is more common and is associated with chronic venous hypertension, while SBS is more rare and is associated with arteriovenous malformations and iatrogenic arteriovenous fistulae. We report 2 patients, representing each type of AAD.

*Cutis.* 2019;103:336-339.

## Case Reports

*Patient 1*—A 56-year-old white man with a history of hypertension, hyperlipidemia, sleep apnea, bilateral knee replacement, and cataract removal presented to the emergency department with a worsening rash on the left posterior medial leg of 6 months' duration. He reported associated redness and tenderness with the plaques as well as increased swelling and firmness of the leg. He was admitted to the hospital where the infectious disease team treated him with cefazolin for presumed cellulitis. His condition did not improve, and another course of cefazolin was started in addition to oral fluconazole and clotrimazole–betamethasone dipropionate lotion for a possible fungal cause. Again, treatment provided no improvement.

He was then evaluated by dermatology. On physical examination, the patient had edema, warmth, and induration of the left lower leg. There also was an annular and serpiginous indurated plaque with minimal scale on the left lower leg (Figure 1). A firm, dark red to purple plaque on the left medial thigh with mild scale was present. There also was scaling of the right plantar foot.

Skin biopsy revealed a dermal capillary proliferation with a scattering of inflammatory cells including eosinophils as well as dermal fibrosis (Figure 2). Periodic acid–Schiff and human herpesvirus 8 (HHV-8) immunostains were negative. Considering the degree and depth of vascular proliferation, Mali-type acroangiodermatitis (AAD) was the favored diagnosis.

*Patient 2*—A 72-year-old white man presented with a firm asymptomatic growth on the left dorsal forearm of 3 months' duration. It was located near the site of a prior squamous cell carcinoma that was excised 1 year prior to presentation. The patient had no treatment or biopsy of the presenting lesion. His medical and surgical history included polycystic kidney disease and renal transplantation 4 years prior to presentation. He also had an arteriovenous fistula of the left arm. His other chronic diseases included chronic obstructive lung disease, congestive

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The authors report no conflict of interest.

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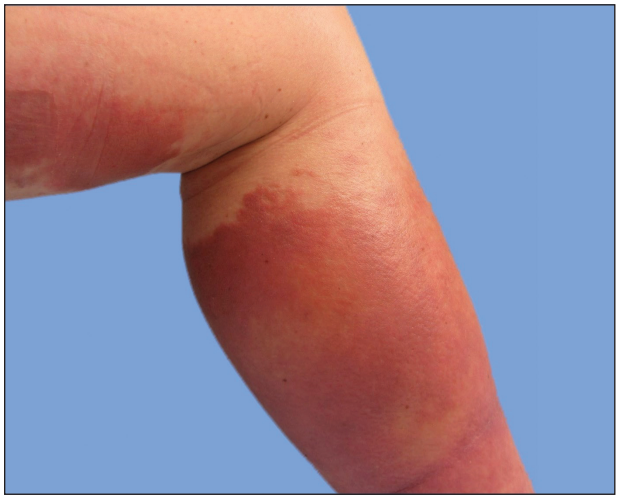
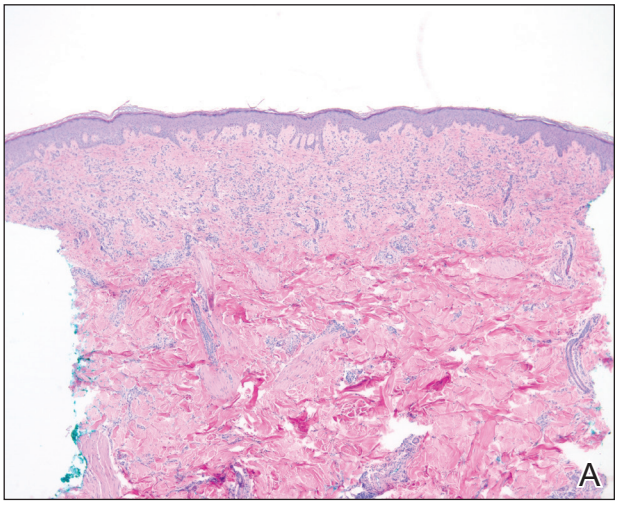


**FIGURE 1.** Mali-type acroangiodermatitis. Annular and serpiginous indurated plaque with minimal scale on the left lower leg.



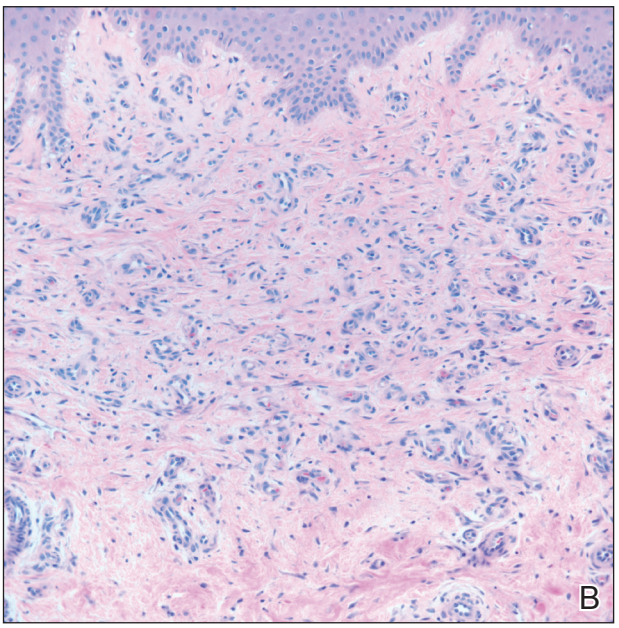




**FIGURE 2.** A and B, Histologic evaluation demonstrated dermal capillary proliferation with a scattering of inflammatory cells and dermal fibrosis (H&E, original magnifications ×4 and ×20).

heart failure, hypertension, type 2 diabetes mellitus, and obstructive sleep apnea.

On physical examination, the patient had a 1-cm violaceous nodule on the extensor surface of the left mid forearm. An arteriovenous fistula was present proximal to the lesion on the left arm (Figure 3).

Skin biopsy revealed a tightly packed proliferation of small vascular channels that tested negative for HHV-8, tumor protein p63, and cytokeratin 5/6. Erythrocytes were noted in the lumen of some of these vessels. Neutrophils were scattered and clustered throughout the specimen (Figure 4A). Blood vessels were highlighted with CD34 (Figure 4B). Grocott-Gomori methenamine-silver stain was negative for infectious agents. These findings favored AAD secondary to an arteriovenous malformation, consistent with Stewart-Bluefarb syndrome (SBS).

## Comment

*Presentation of AAD*—Acroangiodermatitis is a rare chronic inflammatory skin process involving a reactive proliferation of capillaries and fibrosis of the skin that resembles Kaposi sarcoma both clinically and histopathologically. The condition has been reported in patients with chronic venous insufficiency,[1] congenital arteriovenous malformation,[2] acquired iatrogenic arteriovenous fistula,[3] paralyzed extremity,[4] suction socket lower limb prosthesis (amputees),[5] and minor trauma.[6-8] The lesions of AAD tend to be circumscribed, slowly evolving, red-violaceous (or brown or dusky) macules, papules, or plaques that may become verrucous or develop into painful ulcerations. They generally occur on the distal dorsal aspects of the lower legs and feet.[110]

*Variants of AAD*—Mali et al[9] first reported cutaneous manifestations resembling Kaposi sarcoma in 18 patients with chronic venous insufficiency in 1965. Two years later, Bluefarb and Adams[10] described kaposiform skin lesions in one patient with a congenital arteriovenous malformation without chronic venous insufficiency. It was not until 1974, however, that Earhart et al[11] proposed the term *pseudo-Kaposi sarcoma*.[10,11] Based on these findings, AAD is described as 2 variants: Mali type and SBS.

Mali-type AAD is more common and typically occurs in elderly men. It classically presents bilaterally on the lower extremities in association with severe chronic venous insufficiency.[5] Skin lesions usually occur on the medial aspect of the lower legs (as in patient 1), dorsum of the heel, hallux, or second toe.[12]

The etiology of Mali-type AAD is poorly understood. The leading theory is that the condition involves reduced perfusion due to chronic edema, resulting in

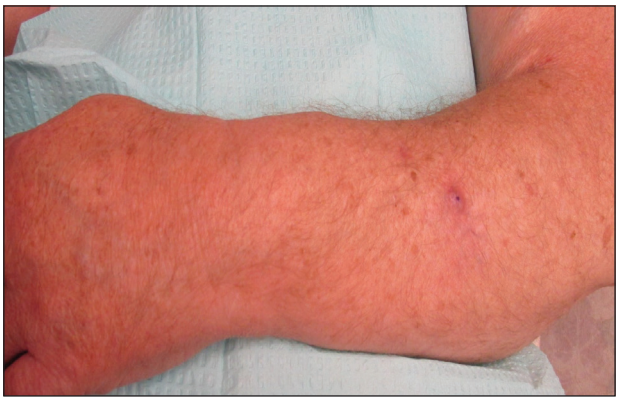


**FIGURE 3.** Acroangiodermatitis secondary to Stewart-Bluefarb syndrome. Violaceous nodule on the left mid forearm with an arteriovenous fistula proximal to the lesion.

neovascularization, fibroblast proliferation, hypertrophy, and inflammatory skin changes. When AAD occurs in the setting of a suction socket prosthesis, the negative pressure of the stump-socket environment is thought to alter local circulation, leading to proliferation of small blood vessels.[5,13]

Stewart-Bluefarb syndrome usually involves a single extremity in young adults with congenital arteriovenous malformations, amputees, and individuals with hemiplegia or iatrogenic arteriovenous fistulae (as in patient 2).[1] It was once thought to occur secondary to Klippel-Trenaunay-Weber syndrome; however, SBS rarely is accompanied by limb hypertrophy.[9] Pathogenesis is thought to involve an angiogenic response to a high perfusion rate and high oxygen saturation, which leads to fibroblast proliferation and reactive endothelial hyperplasia.[1,14]

*Diagnosis and Differential Diagnosis*—Prompt identification of an underlying arteriovenous anomaly is critical, given the sequelae of high-flow shunts, which may result in skin ulceration, limb length discrepancy, cortical thinning of bone with regional osteoporosis, and congestive heart failure.[1,5] Duplex ultrasonography is the first-line diagnostic modality because it is noninvasive and widely available. The key doppler feature of an arteriovenous malformation is low resistance and high diastolic pulsatile flow,[1] which should be confirmed with magnetic resonance angiography or computed tomography angiography if present on ultrasonography.

The differential diagnosis of AAD includes Kaposi sarcoma, reactive angioendotheliomatosis, diffuse dermal angiomatosis, intravascular histiocytosis, glomeruloid angioendotheliomatosis, and angiopericytomatosis.[15,16] These entities present as multiple erythematous, violaceous, purpuric patches and plaques generally on the extremities but can have a widely varied distribution. Some lesions evolve to necrosis or ulceration. Histopathologic analysis is useful to differentiate these entities.

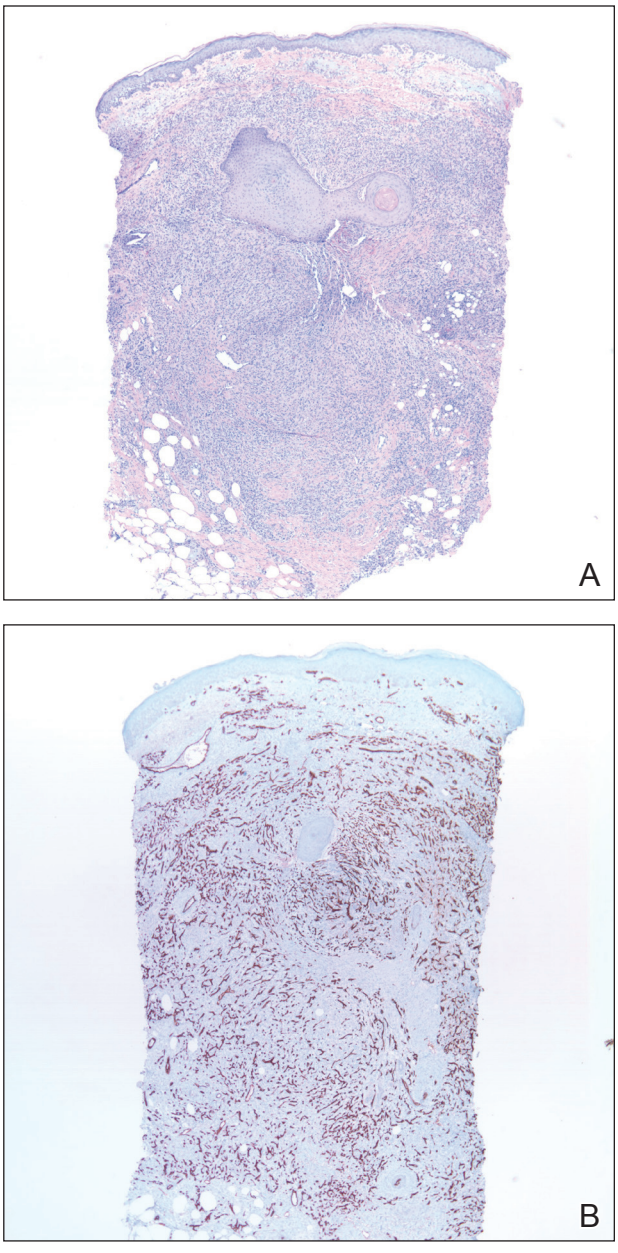



**FIGURE 4.** A, Histologic evaluation demonstrated a proliferation of small vascular channels. Intraluminal erythrocytes with neutrophils scattered throughout (H&E, original magnification ×4). B, CD34 stain highlighted blood vessel proliferation (original magnification, ×2).

*Histopathology*—The histopathologic features of AAD can be nonspecific; clinicopathologic correlation often is necessary to establish the diagnosis. Features include a proliferation of small thick-walled vessels, often in a lobular arrangement, in an edematous papillary dermis. Small thrombi may be observed. There may be increased fibroblasts; plump endothelial cells; a superficial mixed infiltrate comprised of lymphocytes, histiocytes, and eosinophils; and deposition of hemosiderin.[2,5] These characteristics overlap with features of Kaposi sarcoma;

AAD, however, lacks slitlike vascular spaces, perivascular CD34+ expression, and nuclear atypia. A negative HHV-8 stain will assist in ruling out Kaposi sarcoma.[1,17]

*Management*—Treatment reports are anecdotal. The goal is to correct underlying venous hypertension. Conservative measures with compression garments, intermittent pneumatic compression, and limb elevation are first line.[18] Oral antibiotics and local wound care with topical emollients and corticosteroids have been shown to be effective treatments.[19-21]

Oral erythromycin 500 mg 4 times daily for 3 weeks and clobetasol propionate cream 0.05% healed a lower extremity ulcer in a patient with Mali-type AAD.[21] In another patient, conservative treatment of Mali-type AAD failed, but rapid improvement of 2 lower extremity ulcers resulted after 3 weeks of oral dapsone 50 mg twice daily.[22]

A tissue matrix–protective agent (a heparan sulfate mimetic) was reported to completely resolve a patient's lower extremity ulcer secondary to SBS after other treatment modalities failed.[19] In the SBS variant of AAD, treatment should be directed toward obliterating the underlying arteriovenous malformation, which can be achieved by selective embolization, endovenous ablation, sclerotherapy, or surgical intervention.[1,2]

## Conclusion

Acroangiodermatitis is a rare entity that is characterized by erythematous violaceous papules and plaques of the extremities, commonly in the setting of chronic venous insufficiency or an arteriovenous shunt. Histopathologic analysis shows proliferation of capillaries with fibrosis, extravasation of erythrocytes, and deposition of hemosiderin without the spindle cells and slitlike vascular spaces characteristic of Kaposi sarcoma. Detection of an underlying arteriovenous malformation is essential, as the disease can have local and systemic consequences, such as skin ulceration and congestive heart failure.[1] Treatment options are conservative, directed toward local wound care, compression, and management of complications, such as ulceration and infection, as well as obliterating any underlying arteriovenous malformation.

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