Acroangiodermatitis Secondary to Chronic Venous Insufficiency

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Acroangiodermatitis (AAD) is a benign uncommon vasoproliferative disorder that affects the lower extremities. It appears to be a reactive phenomenon related to severe chronic venous insufficiency and stasis of the lower extremities. The clinical presentation of this condition often is similar to Kaposi sarcoma. We report a case of AAD in a patient with severe hypertension and chronic venous insufficiency.

**Case Report**

An 89-year-old woman with a long-standing history of hypertension, chronic venous insufficiency, aortic stenosis, and bilateral total knee replacement presented with multiple asymptomatic, firm, violaceous nodules and plaques distributed extensively over her left lower extremity. The lesions developed approximately 4 years prior and gradually increased in number with worsening of her chronic venous insufficiency. Review of her clinical history did not reveal evidence of human immunodeficiency virus infection or Kaposi sarcoma.

Physical examination revealed multiple 1- to 2-cm well-circumscribed, violaceous, scaly nodules and plaques extending from the left lower knee to the medial ankle (Figure 1). Similar lesions were seen on the left calf. The lesions were not tender to palpation. The remainder of the physical examination was unremarkable.

A punch biopsy specimen of the left medial shin was obtained. Histologic examination demonstrated a marked proliferation of numerous dilated, thick- and thin-walled vessels throughout the dermis (Figure 2). Many of the vessels were oval, lined by plump endothelial cells, and had luminal thrombi. Extravasated red blood cells and increased numbers of spindle-shaped cells were seen perivascularly. The vessel walls were CD34+ and negative for human herpesvirus 8. Additionally, scattered lymphocytes were seen throughout the dermis. The epidermis was unremarkable and a narrow band of uninvolved dermis was present between the epidermis and dermal infiltrate.

Acroangiodermatitis (AAD) was diagnosed based on the clinical and histologic findings. The patient was prescribed compression stockings and referred back to her primary care physician with a recommendation to better improve her hypertension and chronic venous insufficiency. The patient was subsequently lost to follow-up.
Acroangiodermatitis

Comment
Acroangiodermatitis, first described in 1965 by Mali et al., is a benign uncommon vasoproliferative disorder that affects the lower extremities. It appears to be a reactive phenomenon related to severe chronic venous insufficiency and stasis of the lower extremities. Paralysis of the affected limb, damage to vessels caused by an ill-fitting prosthesis, arteriovenous malformation, and the presence of Klippel-Trenaunay-Weber syndrome are predisposing factors. Additionally, AAD may be associated with other hypercoagulable genetic conditions, such as carriage of the thrombophilic 20210A variant (mutation) in the prothrombin gene, and homozygous protein C resistance. The formation and deposition of thrombi in the lower extremities appear to further compound venous stasis, leading to AAD.

Clinically, the patient may present with well-circumscribed; slowly evolving; red, violaceous, brown, or dusky macules, papules, or plaques. The lesions may be pruritic or painful. Histopathology of a biopsy specimen may reveal epidermal hyperplasia with slight acanthosis, proliferation of small blood vessels and fibroblasts, and extravasated erythroblasts in the dermis with abundant hemosiderin deposition. Also, a perivascular superficial infiltrate consisting of lymphocytes, histiocytes, and eosinophils may be present as well as small luminal thrombi.

Although the pathogenesis of AAD and Kaposi sarcoma is quite different, AAD is sometimes referred to as pseudo-Kaposi sarcoma because it can clinically and histologically resemble Kaposi sarcoma. However, unlike Kaposi sarcoma, the blood vessels in AAD have a fairly regular profile. The vascular spaces appear more oval shaped compared to the jagged ones found in Kaposi sarcoma. These vascular spaces are lined by plump rather than thin endothelial cells. Additionally, positive staining of affected spindle cells for human herpesvirus 8 is characteristically found in Kaposi sarcoma but is not found in AAD.

Many modes of therapy have been employed for AAD. Correction of the underlying venous stasis using compression garments or surgery is of paramount importance. Laser ablation has been demonstrated to be effective in the treatment of solitary lesions. Dapsone, stanozolol, and erythromycin also have improved the condition, but the exact mechanisms are poorly understood. Responses to these treatments have been underwhelming.

REFERENCES