CASE REPORT

Acroangiodermatitis: Review of the Literature and Report of a Case Associated with Symmetrical Foot Ulcers

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Introduction

Acroangiodermatitis (AAD) is a rare vasoproliferative disorder, usually of the lower limb, associated with a number of congenital and acquired vascular conditions. Only a few cases have been reported over the last 30 years, mainly in dermatological and plastic surgery journals. We have reviewed the previous literature and present the first case of AAD in a patient who presented to the vascular clinic with chronic venous insufficiency and foot ulcers that were resistant to conventional treatment.

Literature Review

In 1964 Kopf and Gonzalez1 first described a condition that they termed “congenital dysplastic angiopathy”. In 1965 Mali et al.2 reported 18 cases of a form of angiodermatitis mainly affecting the lower limbs, due to chronic venous insufficiency and histologically resembling Kaposi’s sarcoma, and adopted the term “acroangiodermatitis” (AAD). In 1967 Stewart3 and Bluefarb and Adams4 reported the same condition associated with a congenital arterio-venous malformation (AVM) and in 1974, Earhart et al. also reported a case associated with a congenital AVM and suggested the name pseudo-Kaposi’s sarcoma.5 Since then sporadic cases of this rare condition have been reported in dermatology and plastic surgery journals. Mali described the characteristic lesions as peculiar purple macular plaques on the extensor aspects of the digits or foot. There were occasionally ulcers associated with these plaques that occurred in sites where no direct pressure was being exerted on the skin. All patients described had elevated venous pressure in the toes that did not fall following exercise.

AAD has been reported in association with Klippel-Trenaunay syndrome,6 after surgical arteriovenous fistula for haemodialysis,7 in a patient with paralysed legs8 and Kolde et al.9 reported AAD in an amputee with poorly fitting suction prosthesis in the absence of arterio-venous malformation.

Aetiology

The aetiology of AAD is not known and although chronic venous insufficiency is a common condition only a small percentage of patients develop AAD and various workers have suggested different theories. Proliferation of fibroblasts and small vessels secondary to high perfusion rate in tissues has been suggested.10 Another theory is that a PGE1 or heparin like factor, which has angiotensin promoting activity, is responsible for the development of this lesion.11–13 A possible role of micro trauma has been suggested by Pleger.14

Histology

Histologically AAD resembles Kaposi’s sarcoma (Fig. 1) as both conditions show a proliferation of
small blood vessels and fibroblasts, extravasated erythrocytes and haemosiderin deposits within the dermis. However the regularity of the vessels, the lack of vascular slits, the relative paucity of inflammatory cell infiltrates and the presence of more oedema, extravasated erythrocytes and haemosiderin** differentiate it from Kaposi’s sarcoma. Electron microscopy should be applied when conventional microscopy is inconclusive as in AAD, there are no spindle cells that is the characteristic neoplastic cell in Kaposi’s sarcoma.

**Treatment**

Surgical and conservative methods have been tried for the treatment of AAD. Surgical ligation of a feeding vessel, laser therapy and elastic support stockings has been tried by different workers. Utermann *et al.* successfully treated AAD by embolisation of the associated arteriovenous fistula. Complete regression following a course of Dapsone and support stockings has been reported by Rashkovsky *et al.* and erythromycin have also been successfully used in treating this rare condition. The exact mechanism by which dapsone and erythromycin act is not known. Erythromycin appears to have anti-inflammatory effect and has been shown to inhibit the chemotaxis of leukocytes, monocytes and eosinophils.

**Case Report**

A 60-year-old man presented to the vascular clinic with 20-year history of venous disease and recurrent bilateral leg ulcers. There was no history of DVT and he had undergone surgery for bilateral varicose veins and had worn graduated support stockings for many years. On examination he had gross bilateral lipodermatosclerosis with evidence of healed ulceration in the gaiter areas. There were some small ulcers on the left leg laterally and chronic inflammation in the left first interdigital cleft but no evidence of critical ischaemia or gangrene.

Further examination revealed normal arterial pulses and on Doppler assessment he had recurrent left long saphenous reflux and both long and short saphenous reflux on the right. A venous duplex scan was requested and Class II below knee stockings and topical antifungal agent for the interdigital area prescribed.

On review a month later he had symptomatic relief with Class II stockings and skin scrapings from the foot were negative for fungi. A duplex scan confirmed bilateral superficial venous reflux and he underwent left saphenofemoral ligation, following which the left leg ulcer healed with Class II socks. On review 3 months later he had healed but he developed an ankle ulcer on the right side, so underwent further venous surgery on the right side and the Class II compression was continued. Three months later the right leg ulcer had healed but he had developed a new ulcer on his left index toe. A plain X-ray ruled out osteomyelitis and as the ulcer in his toe remained resistant to simple treatment and an excision biopsy was done which confirmed acroangiodermatitis. Postoperatively the wound did not heal and within 3 months and he had developed an identical ulcer on the index toe of the right foot. These symmetrical ulcers were resistant to treatment with topical antibiotics and regular dressings so he was prescribed erythromycin...
500 mg four times a day, following which there has been a significant improvement (Figs 2 and 3).

**Conclusion**

Acroangiodermatitis is a rare, benign condition associated with chronic venous insufficiency and may progress to foot ulcers. It resembles Kaposi’s sarcoma histologically but appears to respond to a combination of surgical treatment of the venous hypertension and medical treatment with erythromycin.

**References**


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