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Acro-angiodermatitis: Review of the Literature and Report of a Case

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Acro-angiodermatitis is a very common disorder, with a close clinical, anatomical and morphological resemblance to Kaposi's sarcoma. Several types of this disorder can be found in different settings. However, these conditions are often misdiagnosed and therefore mistreated. A review of the literature and a classification of all types of acro-angiodermatitis are presented. We also describe a case of a patient with acro-angiodermatitis which completely regressed following a course of dapsone combined with leg elevation and elastic support stockings. ***Key words: pseudo-Kaposi's sarcoma; dapsone; regression; classification.***

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The term "acro-angiodermatitis" was introduced in 1965 by Mali et al. (1), who described peculiar mauve-coloured macules and plaques developing on the extensor surfaces of the feet in 18 patients with chronic venous insufficiency. This disorder, which Mosto had termed "disseminated pruriginous angiodermatitis" (2), was later named "acro-angiodermatitis of Mali".

Randall et al. (3) and Steigleder (4) discussed the striking clinical and histopathological similarity between the members of a group of pigmented purpuric eruptions appearing mainly on the legs. These included Schamberg's progressive pigmentary dermatitis, angioma seurpiginosum of Hutchinson, Majocchi's purpura annularis teleangiectodes and Gougerot & Blum's pigmented purpuric lichenoid dermatitis. However, despite the close similarity in the end-stage histopathology of the latter to that of acro-angiodermatitis, these entities are clinically distinct.

In 1967 Stewart (5) and Bluefarb & Adams (6) independently described similar lesions on the legs of patients with arteriovenous malformations. This condition later became known as the Stewart-Bluefarb syndrome.

The term "pseudo-Kaposi's sarcoma" has often been used for both these conditions, as the clinical and histopathological appearance is similar to early Kaposi's sarcoma. Other names for this disorder are acro-angiodermatitis, angiodermatitis, congenital dysplastic angiopathy and Kaposi-like arteriovenous (A-V) malformation. The importance of differentiating it from Kaposi's sarcoma cannot be overemphasized.

Acro-angiodermatitis may be encountered in five different clinical conditions (Table I):

(1) Chronic venous insufficiency
(2) A-V malformation in the legs
(3) Iatrogenic A-V shunts in hemodialysis patients
(4) Paralyzed limbs
(5) Amputation stumps

ACRO-ANGIODERMATITIS ASSOCIATED WITH CHRONIC VENOUS INSUFFICIENCY (Refs. 1, 7–11, present case):

A total of 34 cases have been reported, comprising 22 males and 12 females. The disease usually appears in the fourth to sixth decades. The lesions are bilateral and often symmetrical, involving mostly the medial aspect of the lower legs, the dorsum of the foot, or the first and second toes. Other signs of chronic venous insufficiency, such as swelling, stasis dermatitis and hyperpigmentation, are often present. The lesions gradually progress and may ulcerate. Favre et al. did not find any relationship between the volume of varices and the extent of cutaneous changes (12). Therapy with compression bandages usually results in complete regression of the lesions.

ACRO-ANGIODERMATITIS ASSOCIATED WITH A-V MALFORMATION (Refs. 5–7, 13–38):

A total of 31 cases have been reported, 26 males and 5 females, of which 5 were associated with the Klippel-Trenaunay-Weber syndrome. The lesions, appearing mostly in the second and

Table I. *Summary of the clinical features of acro-angiodermatitis*

CVI = chronic venous insufficiency, AVM = arteriovenous malformation, AVS = arteriovenous shunt, PL = paralyzed limb, AS = amputation stump.

	CVI	AVM	AVS	PL	AS
No. of cases	34	31	7	6	3
Mean age of onset	53	30	51	46	54
Sex (M:F)	2:1	5:1	6:1	5:1	1:0
Sites	Bilateral, symmetrical, lower limbs	Unilateral, lower limbs, dorsal foot, 1st, 2nd and 3rd toes	Arm, hands, distal to shunt	Lower limbs	In stump
Prognosis	Good	Poor (especially when congenital)	Good	?	?

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third decades of life, are usually unilateral and occur on the dorsal aspect of the first to third toe. Other signs and symptoms related to the A-V shunt, such as pain, oedema, varices, limb hypertrophy, hyperthermia, hyperhydrosis and a vascular mass with a palpable thrill, may be noted. The course of the disease is variable. In most patients, the pain persists or increases. Ulcerations with arterial bleeding may develop and congestive heart failure can occur. Treatment includes embolization, surgical ligation of the shunts, or, in intractable cases, limb amputation.

ACRO-ANGIODERMATITIS ASSOCIATED WITH IATROGENIC A-V SHUNTS (Refs. 39–44):

A total of 7 cases have been reported, 6 males and 1 female. The ages ranged from 43 to 67 years. In 6 cases, acro-angiodermatitis developed distal to a radial artery/cephalic vein shunt, and in one case distal to a femoral artery/saphenous vein shunt. The lesions developed gradually with a latent period of 2 months to 7 years after placement of the shunt. Persistent ulcerations following skin biopsy were noted. In one case (39), the skin lesions cleared following a spontaneous thrombosis of the A-V shunt. In the remaining 6 cases, the lesions resolved subsequent to surgical ligation of the A-V shunts.

ACRO-ANGIODERMATITIS ASSOCIATED WITH A PARALYZED LIMB (Refs. 14, 45):

Six cases have been reported, 5 males and 1 female. The age of onset varied from 27 to 69 years. Typical Kaposi-like lesions were present on the lower legs and dorsa of the feet. It is doubtful whether this is a separate variety of acro-angiodermatitis, as the paralyzed limb often has features of chronic venous insufficiency due to a disturbance of the muscle pump mechanism. The support prosthesis often used by such patients has been shown to increase the venous pressure in the affected limb (46). The choice of treatment as well as prognosis are unclear. Improvement was noted in one case after kinesiotherapy (14). A cessation of lesion progression was reported in one case treated with elastic support stockings (46).

ACRO-ANGIODERMATITIS ASSOCIATED WITH AMPUTATION STUMPS (Refs. 15, 47, 48):

Three cases have been reported, all males, ranging in age from 31 to 65 years. All had above-knee amputations. Typical kaposiform lesions developed in the amputation stump.

Additional cases have been reported under the heading of "acro-angiodermatitis" without reference to the possible etiology and classification (49, 50).

HISTOLOGY OF ACRO-ANGIODERMATITIS

In all varieties of acro-angiodermatitis, the histological picture resembles an advanced stage of the changes seen in stasis dermatitis. The epidermis is characterized by mild acanthosis with some hyperkeratosis. The dermis, particularly the papillary dermis, is oedematous with marked small vessel proliferation in the upper and mid-dermis. In addition, marked red blood cell extravasation, abundant hemosiderin deposition and fibrosis may be present. In contrast to Kaposi's sarcoma, the vessels are regular, lack vascular slits, and are devoid of the promontory sign. The lack of collagen bundle dissection by new vascular channels, the absence of pleomorphic hyperchromatic spindle cells, and the paucity of inflammatory cell infiltrate also help distinguish acro-angiodermatitis from Kaposi's sarcoma (15).

Headly & Cole (41), in an ultrastructural study, were the first to report vascular slits, spindle cells and occasional mitotic figures in acro-angiodermatitis. They described features that were identical to those of true Kaposi's sarcoma (51). These findings, however, have not been confirmed by other ultrastructural studies.

ETIOLOGY

The etiology of acro-angiodermatitis is unknown. Some authors believe that a high perfusion rate of susceptible tissue may cause small vessels and fibroblasts to proliferate (4, 5). It should, however, be noted that although venous congestion is a common disorder, only a small percentage develop acro-angiodermatitis. It is thus suggested that a distinct factor, such as PGE_1 or heparin, which has angiotensin-promoting activity, is responsible for the development of this condition (52–54). Pfleger thought that an exogenous microtrauma might play a role in the pathogenesis of this disease (55).

CASE REPORT

A 65-year-old female was admitted to the Department of Dermatology, Hadassah University Hospital, with bilateral purple-brown macules and ulcerations on the medial aspect of her ankles and lower calves. Three years prior to admission, she had developed severe episodes of pain and swelling of her feet. The skin lesions had appeared 1 year prior to admission and had slowly enlarged and coalesced. Ulcerations ultimately developed in some of the lesions. There was no history of thrombophlebitis, significant leg trauma, parasthesia, intermittent claudication or impaired ambulation.

The patient also had a history of chronic Weber-Christian panniculitis involving her breasts, axillae, abdomen and back. The panniculitis was still partially active despite aggressive long-term therapy with oral steroids, colchicine, hydroxychloroquine and salazopyrin.

Examination of the skin revealed numerous, discrete to confluent, sharply bordered, purple-brown, tender, 5–10 cm-sized macules on the ankles and lower calves (Fig. 1). Two ulcers, with sharp borders and necrotic centres, were present on the medial aspects of both ankles. The complete blood count, serum electrolytes, liver function tests, immunoglobulins, ANA and anti-cardiolipin were all within normal limits. Doppler examination revealed markedly incompetent valves of the superficial calf veins, the perforators and the saphenofemoral junctions on both sides. Arterial blood flow was normal.

Histological examination of a skin biopsy (Fig. 2) revealed capillary proliferation, red blood cell extravasation, hemosiderin deposits and multiple siderophages throughout the dermis. Fibrosis, oedema and a sparse mixed infiltrate was evident in the upper dermis. No vascular slits, promontory sign, or other sarcomatous changes were seen.

Initial treatment consisted of leg elevation, debridement of the ulcers, and open-wet dressings. Due to a poor clinical response, however, dapsone at a dosage of 50 mg twice daily was administered. A rapid response was seen, and within 3 weeks the ulcers were considerably smaller and shallower. The patient also experienced a significant decrease in pain and was discharged with elastic support stockings. Within 3 months, the ulcers had closed and the skin lesions had completely resolved (Fig. 3).

This case presentation is a typical example of acro-angiodermatitis associated with chronic venous insufficiency, although, in view of the

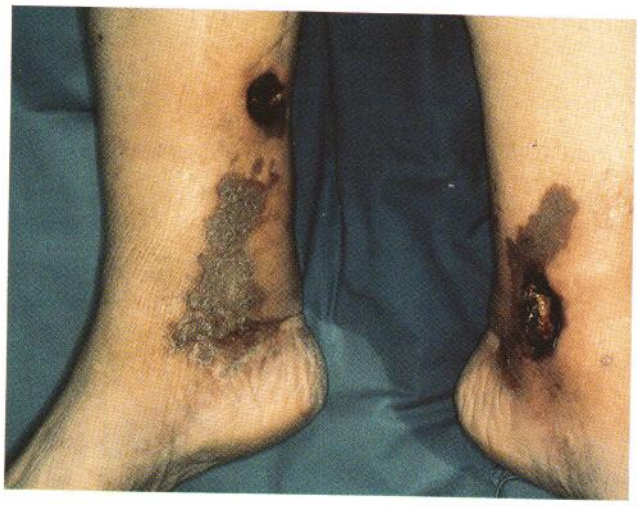

Fig. 1. Photograph of lower legs showing typical mauve-coloured lesions on ankles, with associated ulcerations.

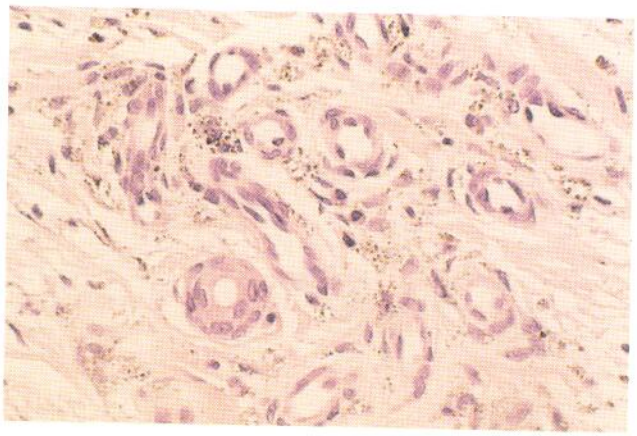

Fig. 2. H & E staining of a biopsy from one of the lesions, showing hemosiderin deposits, capillary proliferation and fibrosis.

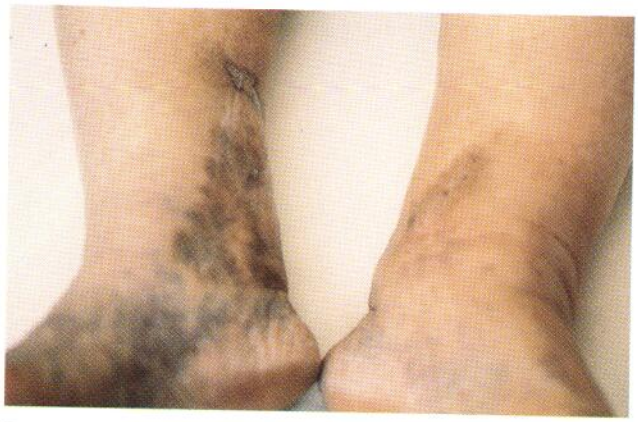

Fig. 3. Photograph of lower legs showing healed lesions.

patients' concurrent immunosuppressive therapy, an initial clinical diagnosis of Kaposi's sarcoma was suspected. Treatment by leg elevation and debridement with open-wet dressings did not result in any improvement, but dapsone therapy induced rapid improvement and eventually complete regression of the lesions.

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