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.
(Accepted June 27, 1995.)

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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 purpuric 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 pigmented purpuric dermatitis, angiosarcoma sequestrum of Hutchinson, Majochi’s purpura annularis telangiectodes 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 arterio-venous (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

<table>
<thead>
<tr>
<th>Feature</th>
<th>CVI</th>
<th>AVM</th>
<th>AVS</th>
<th>PL</th>
<th>AS</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of cases</td>
<td>34</td>
<td>31</td>
<td>7</td>
<td>6</td>
<td>3</td>
</tr>
<tr>
<td>Mean age of onset</td>
<td>53</td>
<td>50</td>
<td>51</td>
<td>46</td>
<td>54</td>
</tr>
<tr>
<td>Sex (M:F)</td>
<td>2:1</td>
<td>5:1</td>
<td>6:1</td>
<td>5:1</td>
<td>1:0</td>
</tr>
<tr>
<td>Sites</td>
<td>Bilateral, symmetrical, lower limbs</td>
<td>Unilateral, lower limbs, dorsal foot, 1st, 2nd and 3rd toes</td>
<td>Arm, hands, distal to shunt</td>
<td>Lower limbs</td>
<td>In stump</td>
</tr>
<tr>
<td>Prognosis</td>
<td>Good</td>
<td>Poor (especially when congenital)</td>
<td>Good</td>
<td>?</td>
<td>?</td>
</tr>
</tbody>
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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, hyperhidrosis 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 leg 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 kine-
otherapy (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 etiol-
ogy 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 papil-
lar dermis, is oedematous with marked small vessel prolifera-
tion in the upper and mid-dermis. In addition, marked red
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 hyper-
chromatic spindle cells, and the paucity of inflammatory cell
infiltrate also help distinguish acro-angiodermatitis from
Kaposi’s sarcoma (15).

Healy & 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 ultra-
structural 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, or heparin, which has angiogenis-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 pannicul-
tis involving her breasts, axillae, abdomen and back. The panniculitis
was still partially active despite aggressive long-term therapy with oral
steroids, colchicine, hydroxychloroquine and salicylates.

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 aspect 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 sapheno-
fermal 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

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patients’ concurrent immunosuppressive therapy, an initial clinical diagnosis of Kaposis’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.

REFERENCES


