

Angiodermatitis Associated with Congenital Arteriovenous Malformation on the Elbow

Sir,

Angiodermatitis is a common disorder with a clinical and morphological resemblance to Kaposi's sarcoma. In 1965, Mali et al. described the condition "acro-angiodermatitis" in 18 patients with chronic venous insufficiency (1). Other names for this disorder are Stewart-Bluefard syndrome, angiodermatitis, congenital dysplastic angiography, Kaposi-like arteriovenous malformation (AVM) and pseudo-Kaposi's sarcoma. Most of cases associated with congenital AVM have developed in the lower leg. We report here a case of angiodermatitis associated with congenital AVM on the elbow.

CASE REPORT

A 37-year-old Korean man presented with an asymptomatic purple 4×5 cm plaque on the left elbow (Fig. 1). Small papules had developed 3 years before and were slowly becoming confluent. On palpation of the lesion, pulsation was observed. He had stated that the pulsation had been present ever since his childhood. Routine haematological examination and urinalysis was normal. An arteriogram revealed AVM. Histopathological findings showed marked proliferation of small vessels and perivascular spindle cells in the upper dermis (Fig. 2).

DISCUSSION

At first, Mali et al. termed the disease acro-angiodermatitis since in most of the 18 patients the condition had developed on the surfaces of the feet (1). Nowadays the term "pseudo-Kaposi's

sarcoma" is more often used. In 1995, Rashkovsky et al. reviewed the cases of acro-angiodermatitis and categorized it as 5 different clinical conditions (2); (a) chronic venous insufficiency; (b) AVM in the leg; (c) iatrogenic A-V shunts in haemodialysis patients; (d) paralysed limbs; and (e) amputation stumps.

In angiodermatitis-associated AVM, the lesions appear mostly in the second and third decades of life, are usually unilateral and occur on the dorsal aspect from the first to the third toe (2). To our knowledge, this is the first case of angiodermatitis on the elbow associated with congenital AVM.

The aetiology of angiodermatitis is unknown. A high perfusion rate of tissue (3, 4) or hypoxia of the tissue distally to AVM could lead to progressive vascular and endothelial proliferation (5). We also think that the possible mechanism in our case may be circulatory compensation for hypoxia, since the hydrostatic pressure of the arm is lower than that of the lower leg. Other factors such as PGE1, heparin and repeated micro-trauma are thought to be involved in this condition (6–8).

Angiodermatitis associated with congenital AVM can develop not only in the leg but also in any other site, such as the elbow. Therefore, we suggest that second category of Rashkovsky's criteria (2) may be corrected to "angiodermatitis-associated congenital AVM" instead of "acro-angiodermatitis associated with AVM in the leg".

REFERENCES

1. Mali JWH, Kuiper JP, Hamers AA. Acroangiodermatitis of the foot. *Arch Dermatol* 1965; 92: 515–518.
2. Rashkovsky I, Gilead L, Schamroth J, Leibovici V. Acro-angiodermatitis: review of the literature and report of a case. *Acta Derm Venereol (Stockh)* 1995; 75: 475–478.
3. Steigleder VJK. Die hamorrhagisch-pigmentaren Dermatosen-ein syndrome oder eine selbständige Erkrankung? *Hautarzt* 1953; 4: 515–520.



Fig. 1. Asymptomatic purple 4×5 cm plaque and papules on the elbow.

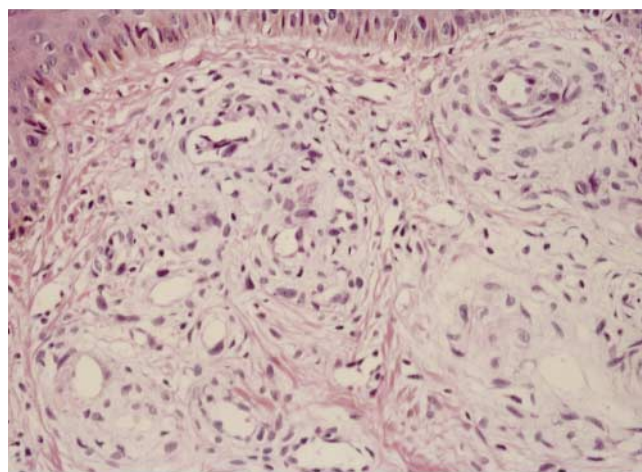


Fig. 2. Proliferation of small vessels and perivascular spindle cells in the upper dermis (Hematoxylin-eosin stain; original magnification $\times 200$).

4. Stewart WM. Fausse angiosarcomatose de Kaposi par fistules arterioveuses multiples. *Bull Soc Fr Derm Syph* 1967; 74: 664–665.
5. König A, Brüngger A, Schnyder UW. Kaposiform acro-angiodermatitis with arteriovenous malformation (Stewart-Blufarb syndrome). *Dermatologica* 1990; 181: 254–257.
6. Ashida ER, Johnson AR, Lipsky PE. Human endothelial cell lymphocyte interaction. *J Clin Invest* 1981; 67: 1490–1499.
7. Auerbach R, Sidky YA. Nature of the stimulus leading to a lymphocyte induced angiogenesis. *J Immunol* 1979; 123: 751–754.
8. Ziche M, Jones J, Gullino PM. Role of prostaglandin E1 and copper in angiogenesis. *J Natl Canc Inst* 1982; 69: 475–482.

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Pityriasis Lichenoides Chronica with Acral Distribution Mimicking Palmoplantar Syphilid

Sir,

Pityriasis lichenoides chronica (PLC) is an idiopathic dermatosis consisting of recurrent crops of erythematous, scaly papules, which exhibit histopathological features of an inter-face dermatitis, often with many necrotic keratinocytes. The trunk and proximal parts of the limbs are preferentially affected. We describe here a case of PLC involving only acral areas, mimicking palmoplantar syphilid.

CASE REPORT

A previously healthy 31-year-old Korean man developed asymptomatic reddish-brown scaly papules on his palms in December 1997. The lesion subsequently spread to the dorsa of the hands, the soles of the feet and distal parts of the limbs. He visited our department 1 month after the onset of skin eruptions. There was no relevant family history and the patient was not on any regular medication. On physical examination, there were crops of round, reddish-brown papules covered by dry mica-like scales. Some of them were crusted in the centre and older ones were flatter in appearance (Fig. 1). Laboratory investigations including VDRL and TPHA were within normal limits. The biopsy specimen of a papule revealed hyperkeratosis, focal parakeratosis, moderate acanthosis with elongation of rete ridges and superficial perivascular infiltrates. Higher magnification showed a few necrotic keratinocytes, moderate lymphocytic exocytosis, spongiosis and vacuolization of the basal layer of the epidermis. Extravasated red blood cells were also noted in both the epidermis and the papillary

dermis. These clinical and pathological features led us to make a diagnosis of PLC. The patient was treated with psoralen photochemotherapy (PUVA). After 21 treatments over 7 weeks, the lesions cleared leaving some hyperpigmentation. His palmar lesions recurred, however, 2 months after discontinuation of the PUVA therapy.

DISCUSSION

Although a few cases of PLC with palmoplantar involvement have been described in the literature, PLC affecting the palmoplantar area and distal parts of the limbs without involvement of the trunk, as seen in this patient, is rare (1, 2). Moreover, our patient is unique since eruptions developed on the palmoplantar areas and then spread to the distal parts of his extremities.

Clinical differential diagnosis of PLC includes secondary syphilis, guttate psoriasis, lichen planus, drug eruption, insect bite, lymphomatoid papulosis, pityriasis rosea and papular eczematous dermatitis. Because our patient showed papulosquamous eruptions on the palmoplantar areas, which did not affect the trunk, a diagnosis of papulosquamous palmoplantar syphilid was considered on the first visit. However, negative results of serological tests for syphilis and characteristic pathological features supported the diagnosis of PLC.

Phototherapy has been considered as the first line of therapy in PLC (3). Our patient initially responded well to PUVA therapy.

REFERENCES

1. Gelmetti C, Rigoni C, Alessi E, Ermacora E, Berti E, Caputo R. Pityriasis lichenoides in children: a long-term follow-up of eighty-nine cases. *J Am Acad Dermatol* 1990; 23: 473–478.
2. Marks R, Black M, Wilson Jones E. Pityriasis lichenoides: a reappraisal. *Br J Dermatol* 1972; 86: 215–225.
3. Levine MJ. Phototherapy of pityriasis lichenoides. *Arch Dermatol* 1983; 119: 378–380.

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Fig. 1. Crops of round, reddish brown papules covered by dry mica-like scales on the palm and distal parts of the arm.