

CASE SERIES

Segmental lichen aureus: A report of two cases treated with methylprednisolone aceponate

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ABSTRACT

Two cases of segmental lichen aureus with a response to topical 0.1% methylprednisolone aceponate ointment are reported. A 9-year-old child and a 23-year-old man showed complete resolution of their lesions following treatment with the latter after 7 months and 4 months, respectively. Lichen aureus is a rare form of the pigmented purpuric dermatoses characterized by golden-brown and lichenoid macules and papules, most often on the lower extremities. Segmental presentations have seldom been described. Histology showed a lichenoid infiltrate with extravasation of red blood cells and haemosiderin deposition. The aetiology is unclear and treatment is disappointing. We report an uncommon segmental presentation of lichen aureus with resolution of the lesions after treatment with a topical corticosteroid.

Key words: Lichen aureus, Pigmented purpuric dermatoses, Methylprednisolone aceponate.

INTRODUCTION

Lichen aureus is a rare form of the pigmented purpuric dermatoses (PPD) characterized by golden-brown and rust coloured or lichenoid macules and papules, most often on the lower legs, but other sites can also be affected. The lesions usually arise suddenly, remain stable or enlarge slowly, and may resemble a bruise with no history of preceding trauma. The condition generally affects young adults and less frequently children. The overall poor response of

this condition to therapy remains unexplained. We report two cases of segmental lichen aureus that resolved after treatment with 0.1% methylprednisolone aceponate (MPA) ointment.

CASE REPORTS

Case 1

A 9-year-old girl presented with a 1-year history of itchy and painful lesions on her left leg. There was no history of prior trauma, or use of oral or topical medication. Physical examination revealed numerous golden-brown macules admixed with lichenoid papules in a segmental distribution involving mainly the anteromedial aspect of her left leg (Fig. 1a). There was no associated swelling of the leg or evidence of a vascular malformation. Routine blood chemistry and full blood count were normal. Duplex Doppler studies of the affected leg did not reveal any abnormalities. Histological examination of the skin punch biopsy specimen showed a normal epidermis with a narrow grenz zone in the upper dermis, extravasation of red blood cells, and a confluent perivascular and band-like lymphohistiocytic infiltrate with vascular endothelial swelling (Fig. 1b). Perls' stain revealed haemosiderin, both free-lying and within the siderophages (Fig. 1c). A diagnosis of segmental lichen aureus was made.

Acroangioidermatitis and PPD of Gougerout and Blum, which were considered as differential diagnoses, were excluded on the basis of clinical and histological differences with this case. The lesions of PPD of Gougerout and Blum are lichenoid, not golden-brown in colour, and are often symmetrical. Acroangioidermatitis presents with lichenoid papules on the lower legs, often on a background of chronic venous insufficiency. The histology shows extravasation of red blood cells, haemosiderin deposition and vascular proliferation with no lichenoid infiltrate. Our patient was

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Abbreviations:

MPA	methylprednisolone aceponate
PPD	pigmented purpuric dermatosis

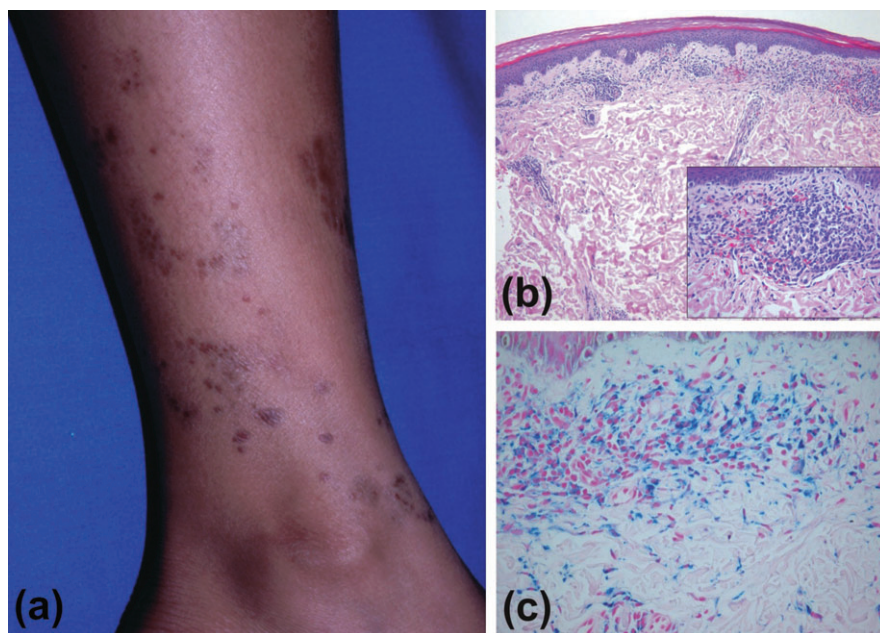


Figure 1 (a) Golden-brown macules and lichenoid papules on the anteromedial aspect of the left leg. (b) Biopsy showing lichenoid and perivascular infiltrate with a grenz zone in the upper dermis and extravasation of the red blood cells (H&E; ×200). Insert showing lichenoid infiltrate (H&E; ×400). (c) Perls' stain showing haemosiderin deposition.

treated with 0.1% MPA ointment daily and oral paracetamol for pain. The lesions improved significantly within 3 months and cleared after 7 months of therapy. No new lesions were seen up to 11 months later.

Case 2

A 25-year-old man presented with a 6-month history of itchy lesions on his abdomen. There was no antecedent rash or trauma and no use of medication. On examination golden-brown macules and patches with petechiae were noted on the left side of his lower abdomen in a segmental distribution (Fig. 2a). The differential diagnoses included eczema, lichen aureus and purpuric mycosis fungoides. Full blood count, urea, electrolytes and liver function tests were normal. Skin punch biopsy revealed an attenuated epidermis with a prominent perivascular lymphohistiocytic infiltrate in the upper dermis, associated with endothelial swelling and extravasation of erythrocytes (Fig. 2b). There were no atypical lymphocytes and the presence of haemosiderin was confirmed using Perls' stain (Fig. 2c). T-cell receptor gene rearrangement studies were done and did not reveal any monoclonal T-cell population. On the basis of the clinical and histopathological findings, a diagnosis of segmental lichen aureus was made. Treatment with daily application of 0.1% MPA ointment was commenced. Lesions resolved after 4 months and therapy was stopped 1 month thereafter. No recurrence of the lesions was noted 1 year later.

DISCUSSION

Lichen aureus is a rare dermatosis first described by Martin in 1958.¹ Calnan in 1960 coined the term lichen aureus to describe the golden-brown colour of the lesions.² It is one of the rarest forms of the PPD, which include: Schamberg's

disease (progressive pigmentary dermatosis), Majocchi's disease (purpura annularis telangiectodes), pigmented purpuric lichenoid dermatosis of Gougerot and Blum, eczematoid-like purpura of Doucas and Kapetanakis, itching purpura of Lowenthal, granulomatous PPD, as well as linear and quadrangular PPD.³ A familial variant of Schamberg's disease has also been reported. There is significant clinical and histological overlap between these entities, but lichen aureus is usually distinct. The lesions of Schamberg's disease generally affect the lower limbs and have a typical 'cayenne pepper' appearance. Majocchi's disease presents with bluish-red annular macules with red telangiectatic puncta and affects the upper and lower extremities. Pigmented purpuric lichenoid dermatosis of Gougerot and Blum may resemble lichen aureus, but the eruption consists of lichenoid papules that fuse into plaques in a symmetrical distribution affecting the legs and rarely the trunk.

Lichen aureus affects young male adults and less frequently children.⁴ It may exhibit a segmental pattern, follow the lines of Blaschko or the course of an underlying vein. The eruption is typically asymptomatic, but may be intensely pruritic and associated with severe pain. Its aetiology is unknown and has been reported in association with trauma and perforator vein incompetence.^{5,6} No firm association of drugs and lichen aureus has been observed however. Lichen aureus has recently been reported in a child following regular consumption of energy drink, with resolution of the lesions after the drink was stopped.⁷

Pathogenetic mechanisms that have been proposed to play a role in lichen aureus include venous hypertension, capillary fragility with leakage of red blood cells, disturbed humoral and/or cell-mediated immunity. The histology of lichen aureus shows a band-like lymphohistiocytic infiltrate with extravasation of red blood cells and haemosiderin

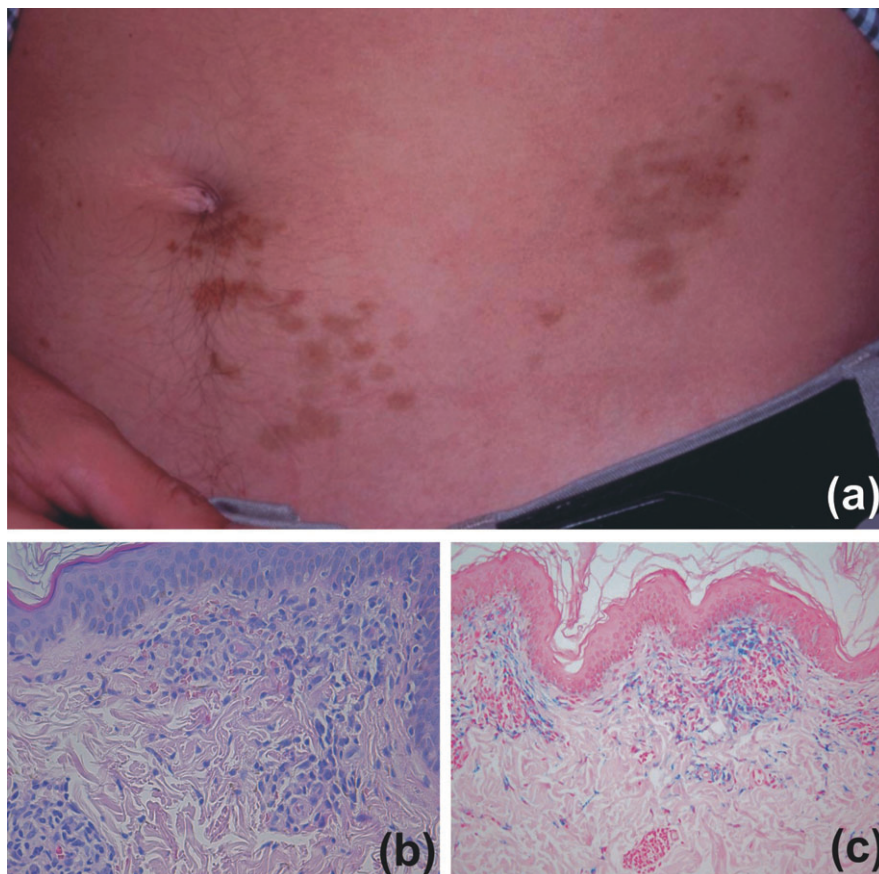


Figure 2 (a) Golden-brown macules and patches on the left lower abdomen. (b) Biopsy showing lichenoid infiltrate and extravasation of red blood cells in the upper dermis (H&E; $\times 200$). (c) Perls' stain demonstrating haemosiderin deposition.

deposition. Perivascular accentuation of the infiltrate in the upper and deep dermis has been described, and the grenz zone, which was seen in our first case, is an inconsistent finding.⁸ The clinical and histological overlap between lichen aureus and purpuric mycosis fungoides has been described.⁹ A more recent large clinicopathological study, with a mean follow-up period of 102.1 months, showed no progression of lichen aureus to mycosis fungoides in their series of patients.¹⁰ The molecular and phenotypical features of 43 cases of PPD were reported and suggested that they are a form of cutaneous T-cell lymphoid dyscrasia based on the frequency of monoclonality, T-cell clonotypes and extent of pan-T cell loss.¹¹

In this series, none of the patients with PPD and polyclonal population on polymerase chain reaction progressed to mycosis fungoides. T-cell gene receptor rearrangement studies in our second patient showed a polyclonal pattern and the lesions have since resolved with no recurrence. Lichen aureus is resistant to treatment and runs a chronic course. It is more liable to spontaneous resolution in children than in adults, with a mean of 5.4 years.⁴ Topical corticosteroids are usually ineffective. There are previous reports of successful treatment of two cases of lichen aureus with potent fluorinated topical corticosteroids.¹² Successful use of 0.1% MPA in the treatment of lichen aureus has recently been reported in a single patient.¹⁵

MPA is a potent, non-halogenated corticoid diester with anti-inflammatory, antipruritic and vasoconstrictive effects indicated mainly for the treatment of eczema.¹⁴ It is highly lipophilic and penetrates readily into the skin. It has a comparatively low atrophogenic potential and is well-tolerated. Recent reports have documented successful treatment of this condition with topical pimecrolimus, PUVA, as well as combination therapy of pentoxifylline and prostacyclin.¹⁵⁻¹⁷ In conclusion, we have described two rare clinical presentations of segmental lichen aureus with a response to 0.1% MPA.

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