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Erosive Palmoplantar Lichen Planus[☆]

Liquen plano erosivo plantar

To the Editor:

Erosive palmoplantar lichen planus (LP), also known as ulcerative or bullous LP, was first described by Cram et al.¹ in 1966 as a rare variant of LP that can affect mucous membranes, palms, and soles. It presents clinically as erythematous, ulcerated plaques that run an extremely chronic course and are resistant to all types of treatment.

The patient was an 84-year-old woman with a personal history of hypertension, aortic valve fibrosclerosis, and polyarthritis who had been receiving treatment with paracetamol, acetylsalicylic acid, atorvastatin, and candesartan-hydrochlorothiazide for several years. She presented with a very painful ulcerated lesion on her right foot that had appeared 6 months earlier and made walking difficult. Physical examination revealed an ulcerated plaque, measuring 4 × 5 cm, on the medial aspect of the right heel. The base of the lesion was filled with granulation tissue, which in some areas was covered with whitish, macerated epithelium. Another ulcer with the same characteristics was present on the dorsal aspect of the first toe (Figs. 1 and 2). The dermatological examination also revealed the presence of whitish reticulated areas on the buccal mucosa and pterygium on several fingernails. In addition, the patient had total scalp alopecia, having lost her hair, as well as all 10 toenails, 6 years earlier. She did not have alopecia of the eyebrows, eyelashes, axilla, or pubic area, or any other cutaneous abnormalities. Blood tests, including liver function tests, hepatitis B serology, and hepatitis C serology, were normal or negative. Skin biopsy revealed marked orthokeratotic hyperkeratosis, acanthosis, and a band-like lymphocytic infiltrate in the upper dermis, without melanophages. Also present were spiculated dermal papillae and vacuolar degeneration at the dermal-epidermal junction. The findings were consistent with LP (Fig. 3).

On the basis of the clinical and histopathologic findings, the patient was diagnosed with erosive palmoplantar LP and was prescribed treatment with oral prednisone (1 mg/kg/d). Two months later, the ulcerated lesions had healed. Systemic treatment was continued with a tapering dose for a further 6 months and finally withdrawn. The patient then started



Figure 1 Absence of 5 toenails and ulcer on the dorsal aspect of the first toe similar to the ulcers on the heel.



Figure 2 Ulcerated plaque, located on the right heel, filled with granulation tissue and whitish, macerated epithelium in some areas.

a 1-month regimen of topical clobetasol propionate (500 mcg/g). She has remained asymptomatic since treatment was completed 24 months ago.

Erosive palmoplantar LP is a rare form of LP that mainly affects elderly people. The lesions initially present as erythema and blisters on the feet and evolve into intensely painful, disabling erosions and ulcers. They usually occur on

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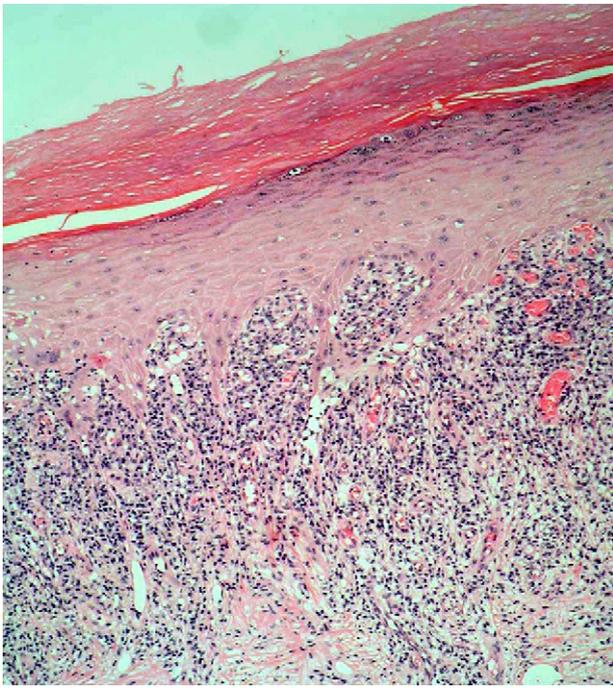


Figure 3 Histologic image of the edge of the lesion. Band-like lymphocytic infiltrate in the papillary dermis and prominent network of interpapillary ridges (hematoxylin-eosin, original magnification $\times 10$).

the heels but can affect the entire sole and even the dorsal surface of the toes.² The disease can lead to the total loss of all toenails.

Erosive palmoplantar LP can occur in isolation or in association with cicatricial alopecia of the scalp and diffuse alopecia of the axilla and pubic area. In many cases, it is accompanied by typical LP lesions.¹ It is important to monitor this condition, as squamous cell carcinoma³ and eccrine syringofibroadenoma⁴ have been known to arise in erosive palmoplantar LP lesions.

There have been numerous anecdotal reports of associations between erosive palmoplantar LP and autoimmune diseases such as primary biliary cirrhosis, autoimmune thyroiditis,⁵ diabetes mellitus, Sjögren syndrome, and rheumatoid arthritis, as well as other abnormalities, such as hypergammaglobulinemia, hypocomplementemia, and chronic liver diseases of infectious origin. Although it is accepted that the cellular immune response plays a key role in the pathogenesis of LP, the etiology and pathogenesis of erosive palmoplantar LP remain unknown.

Because erosive palmoplantar LP is chronic and resistant to both topical and systemic treatments, numerous treatment options have been proposed. Local treatments used to date, with variable results, include high-potency corticosteroids, retinoids, intralesional triamcinolone acetonide, topical ciclosporin,⁶ and psoralen-UV-A bath therapy.⁷ Topical tacrolimus 0.1% cream applied twice daily was recently shown to be effective in the treatment of ulcerative LP affecting mucous membranes.⁸

Oral treatments used to date include systemic corticosteroids, dapsone, etretinate, ciclosporin, mycophenolate mofetil, platelet-derived growth factor, griseofulvin, and thalidomide. Good response has been seen after 4 to 13 weeks of subcutaneous low-molecular-weight heparin (LMWH) administered at low doses (3 mg/wk); LMWH inhibits the type IV hypersensitivity response that appears to be involved in the pathogenesis of LP.^{9,10}

UV-A1 radiation (380-400 nm), which induces apoptosis of CD4⁺ T cells, has proved to be an effective treatment in some cases. Extracorporeal phototherapy has also been effective in isolated cases.

Surgery, with or without immunosuppressive therapy with ciclosporin, has occasionally been effective against chronic, stable, localized cases of erosive palmoplantar LP.¹¹

This case is remarkable for the patient's excellent response to conventional treatment with systemic corticosteroids and the absence of recurrence after 2 years, despite the fact that erosive palmoplantar LP is refractory to most available treatments. Nevertheless, because of the high recurrence rate associated with this disease, we consider that the patient should be monitored closely.

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Generalized Eruptive Multinucleate Cell Angiohistiocytoma in a Pregnant Woman: A Case Report and Review of the Literature[☆]

Angiohistiocitomas de células multinucleadas eruptivos y generalizados en una gestante. Presentación de un caso y revisión de la literatura

To the Editor:

Multinucleate cell angiohistiocytoma (MCAH) is a rare entity that takes the form of a benign dermatologic proliferation formed by small blood vessels and multinucleate mesenchymal cells. These lesions are probably underdiagnosed because of their similarity to other disorders, such as dermatofibroma, and because of their benign, asymptomatic nature.

We report the case of a 35-year-old woman in the 36th week of her 4th pregnancy. The child born as a result of her second pregnancy had died 24 hours post partum. The woman had been visiting our department for many years due to multiple asymptomatic lesions on her trunk and extremities; the lesions first appeared during the first pregnancy and had increased in number over the course of the subsequent pregnancies. The physical examination revealed more than 100 round maculopapular lesions with diameters ranging between 2 and 8 mm; the lesions were erythematous-violaceous with a smooth surface and were distributed unevenly across the trunk and extremities (Fig. 1). Dermoscopy showed a fine pigmented reticular pattern around the borders and a whitish central patch (Fig. 2); these findings were compatible with clinical suspicion of multiple eruptive dermatofibroma. Because this presentation of dermatofibroma has been associated with autoimmune disease, an immunological study was performed. The salient findings were the presence of anti-Ro 60 antibodies and antinuclear antibodies distributed in a speckled pattern.

Biopsy specimens of 2 papules revealed poorly defined lesions involving the full thickness of the dermis. Histology showed that the papules consisted of numerous small blood vessels with prominent endothelia distributed with no clear pattern throughout the dermal collagen. Other findings included scant perivascular histiocytes and

lymphocytes and multinucleate mesenchymal cells characterized by fewer than 10 nuclei, no abnormalities, and a large cytoplasm with angular contours (Fig. 3a). Immunohistochemical analysis showed estrogen receptor α expression in some perivascular spindle-shaped cells (Fig. 3b) and progesterone receptor expression in very few cells at the same location. Immune staining was negative for hormone receptors in the multinucleate cells. A diagnosis of generalized eruptive MCAH was reached.

The skin lesions were not treated because of the widespread nature of the outbreak. The patient was referred to obstetrics because of the positive anti-Ro 60 result and the history of the death of the newborn child after her earlier pregnancy. The neonate was monitored by the pediatrics department.

MCAH is considered to be a dermal dendrocyte proliferation. Some authors consider it to be a variant of dermatofibroma¹; as with dermatofibroma, it is difficult to determine whether MCAH is a true neoplasm or a reactive process.² MCAH presents in healthy persons and reported associations with other diseases appear to be coincidental. Several factors support the idea that this is a reactive entity: the fact that some patients present with multiple lesions, the location of lesions in sites exposed to trauma and insect bites, the absence of a familial association, and the spontaneous regression reported in some cases. A link to hormone factors has been suggested, as these lesions present more frequently in women (79% of cases) and have been associated with overexpression of estrogen receptor α .³



Figure 1 Multiple round erythematous-violaceous maculopapular lesions measuring up to 6 or 7 mm, on the abdomen.

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