



Palmo-Plantar Lichen Planus Keratoderma

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Abstract

Lichen planus is a widespread dermatosis of obscured pathogenesis and typical histopathological findings. Multiple polygonal papules on the flexural aspects of the upper extremities, shins and lumbal zone favor the classic diagnosis; hence, versatile clinical forms exist based on distribution and morphology of the clinical lesions. Palms and soles are very rarely affected. Herein, a challenging case of palmoplantar lichen planus keratoderma is presented to highlight its rarity and therapeutic resistance.

Keywords: Palmo-plantar; Lichen planus; Keratoderma

Introduction

Lichen Planus (LP) is a common inflammatory dermatosis with chronic-relapsing course, affecting the skin, mucous membranes, hair and nails. More prevalent among middle-aged women, the disease shows no racial predilections or certain geographical distribution. There are some familial cases, which tend to progress more aggressively [1].

LP occurs at highest frequency in association with some bacterial or viral infections, especially HCV, contact allergens, recently applied vaccinations, administration of certain drugs, thyroid dysfunction and underlying neoplasms [2].

The exact pathogenesis remains unclear. Latest theories describe LP as a chronic inflammatory disorder of altered basal cell layer keratinocytes, destroyed by auto-antibodies and effector T-cells in genetically predisposed individuals [3].

LP typically presents with pruritic red or violaceous flat papules with shiny surface, frequently covered with fine white scales, known as "Whickham's striae". A variety of other clinical forms are also described: Actinic, exanthematous, annular, atrophic, bullous, and hypertrophic (verrucosus), linear, pigmented and ulcerative [4]. Palmo-plantar forms are extremely rare and even more challenging to diagnose. To date, several clinical subtypes of LP of the palms and soles have been reported: Hypertrophic, diffuse scaly, psoriasiformed, plaque with collarette scale, keratotic plaque with pits, punctate keratosis, vesicular, macular and diffuse keratoderma [5]. Histologically identical, those variants share similar etiology and pathogenesis, however, differ in progression, prognosis and response to treatment.

We present one of the most uncommon, hard to diagnose and therapy-resistant subtype: palmoplantar keratoderma.

Case Presentation

A 58-year-old woman presented with pruritic thick eruption on her hands and feet of two-month duration. The lesions primarily appeared on the hands and rapidly coalesced to diffusely cover the entire palms. In two weeks, the eruption spread to her feet. No provocative triggers were identified with the exception of smoking. She had no personal or family history of concomitant disease, but revealed to be diagnosed with classic lichen planus 20 years ago.

The physical examination revealed diffuse yellowish thick hyperkeratotic plaques, symmetrically distributed over the entire palms and soles. The lesions were sharply demarked by infiltrated erythemo-livid borders (Figure1). Violaceous monomorphous papules in linear configuration were found on the medial aspect of her right lower leg on the site of a tiny excoriation (Koebner's phenomenon). The mucous membranes, nails and hair were spared.

The laboratory findings were in referential ranges. No metabolic or infectious underlying

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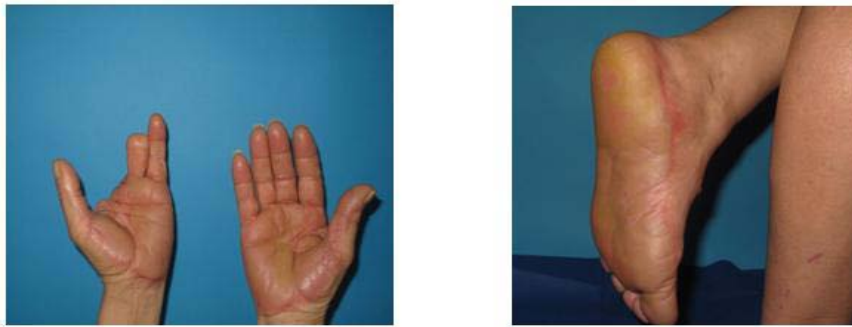


Figure 1: Diffuse keratoderma on the palms and soles, demarcated by well-defined elevated erythemo-livid border.

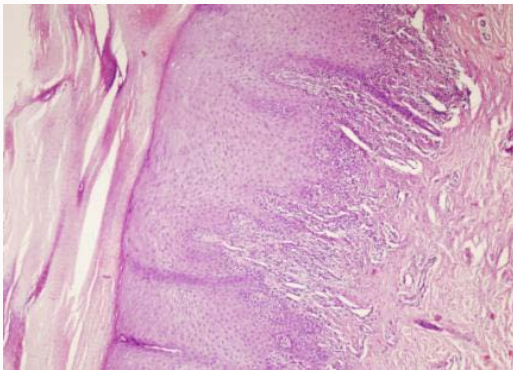


Figure 2: Compact hyperkeratosis, hypergranulosis, irregular acanthosis, vacuolar changes of the basal layer with necrotic keratinocytes and a dense band-like lymphocytic infiltration in the upper dermis (x200, HE).

conditions were found. The biopsy demonstrated compact hyperkeratosis, hypergranulosis, irregular acanthosis, vacuolar degeneration of the basal layer with necrotic keratinocytes in the lower portion of the epidermis, and a dense band-like lymphocytic infiltration in the upper dermis (Figure 2). Those findings were compatible with lichen planus.

Topical photochemotherapy in combination with acitretin (0.5 mg/kg/day) daily was introduced. On the first-month follow-up visit the patient was slightly improved.

Discussion

Palms and soles are unusual sites of LP involvement [6]. Only 12.9% of all LP patients have palmoplantar lesions and only a quarter of them present with exceptional lesions on palms and soles [5,7]. Palmo-plantar LP keratoderma is one of the most uncommon localized variants of LP, with only a few cases reported so far.

Palmo-plantar LP demonstrates a versatile clinical picture. To date, several subtypes have been described. The largest series with palmo-plantar LP included 18 patients. Of them, the majority (38%) had hypertrophic involvement, followed by diffuse scaly (11%), psoriasiform (11%), and plaque with collarette scale (11%) [5,6]. Anecdotal cases presented with keratotic plaque with pits, punctate keratosis, vesicular, and macular. Diffuse keratoderma was seen in only one patient (5.5%), which ranks it among the subtypes with lowest incidence. Our case showed isomorphic phenomenon of typical polygonal erythemo-livid papules on site of a linear excoriation that were clinically highly suggestive of LP. The diagnosis was confirmed afterwards by histological analysis.

Unlike the classic LP, where itching is not necessarily presented, palmo-plantar LP keratoderma is associated with severe pruritus, burning and pain [8,9]. Moreover, it usually demonstrates longer duration and an unpredictable chronic-relapsing course. Therefore, the patient's quality of life is extremely worsened. Our patient suffered severe itch and burning sensation, which intensified with walking. The subjective symptoms refracted despite proper therapy.

No standardized treatment protocol has been established. The best therapy outcomes are reported after a long-term administration of oral retinoids in combination with topical corticosteroids [10,11]. Several other modalities come as a second-line choice for severe and resistant cases: systemic corticosteroids, immune suppressors, dapsone, enoxaparine, etc. UVB-NB phototherapy and topical PUVA is still controversial [12]. Palmo-plantar LP keratoderma is considered more therapeutically resistant and hard to control.

Conclusion

Palmo-plantar keratoderma is a very rare chronic relapsing, recalcitrant subtype of lichen planus. The clinical diagnosis is extremely challenging, and requires proper histopathological verification. The case presented adds a humble contribution to the very few anecdotal cases worldwide.

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