

Dyshidrosiform Palmoplantar Pemphigoid in a Young Man: Response to Dapsone

Francesca Lupi¹, Cinzia Masini¹, Marina Ruffelli², Pietro Puddu¹ and Giuseppe Cianchini^{1*}

Departments of ¹Immunodermatology, ²Allergology and Immunology Service, Istituto Dermatopatico dell'Immacolata, IRCCS, Via dei Monti di Creta 104, IT-00167 Rome, Italy. E-mail: g.cianchini@idi.it

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Sir,

Dyshidrosiform pemphigoid (DP) is an unusual variant of localized pemphigoid presenting with a persistent vesicobullous eruption localized to the soles or palms or both, similar to dyshidrosiform dermatitis. This condition is rarely reported in the literature, it generally occurs in the elderly and there are no reported cases in young people. We describe here a rare case of DP that developed in a young man and was limited only to the palms and soles.

CASE REPORT

A 20-year-old man presented with recurrent and itchy vesicles localized on both palms which has developed over the previous year. The first clinical diagnosis was dyshidrosis and he was treated with topical steroids combined with oral antihistamine for 8 weeks, with a transitory improvement. Treatment with oral prednisone, 25 mg daily for 16 weeks, did not improve his skin condition, and while he was treated vesicles occurred also on his soles. On examination the patient showed multiple tense vesiculobullae of various sizes, with a base of normal or erythematous skin on the palms and soles (Fig. 1). The remaining skin was spared and the mucous membranes were normal. Nikolsky's sign was negative. Fungal culture of the lesions was negative and the blood chemistry tests detected marked eosinophilia 16.6% (1,250/ μ l). Biopsy specimens from the palms and the soles showed subepidermal vesicle containing erythrocytes and rare eosinophils with a dermal inflammatory infiltrate made of lymphocytes, histiocytes and eosinophils. Direct

immunofluorescence was positive for continuous linear deposition of immunoglobulin IgG and C3 at the dermo-epidermal junction (Fig. 2). Indirect immunofluorescence disclosed anti-basement membrane antibodies IgG at a titre of 1:80. An enzyme-linked immunosorbent assay (ELISA) for circulating antibodies against bullous pemphigoid antigen BP180 was positive with a titre of 50.7 U/ml (normal value: <9 U/ml). On the basis of these patterns a diagnosis of dyshidrosiform palmo-plantar pemphigoid was made. Treatment with topical steroids and dapsone 50 mg daily produced a rapid improvement. After 4 months the dapsone was tapered (25 mg daily) with a complete clinical remission, and now 12 months later, the patient is still in good condition.

DISCUSSION

DP is an unusual form of localized pemphigoid that was first described by Levine et al. in 1979 (1). The pathogenesis of localized pemphigoid is unknown. Some authors have suggested that physical agents, such as thermal burns or radiotherapy, might initiate the development of localized bullous pemphigoid (2). The diagnosis of DP may be misleading for clinicians because clinical lesions are often indistinguishable from dyshidrosiform dermatitis and the correct diagnosis has to be confirmed by immunopathological findings that reveal the typical hallmarks of BP (3). To the best of our knowledge only 21 cases of DP have been described in the literature, mainly in elderly subjects (3–8), and this is the first reported case in a young man with DP localized only to the palms and soles.



Fig. 1. Multiple vesiculobullae on palms.

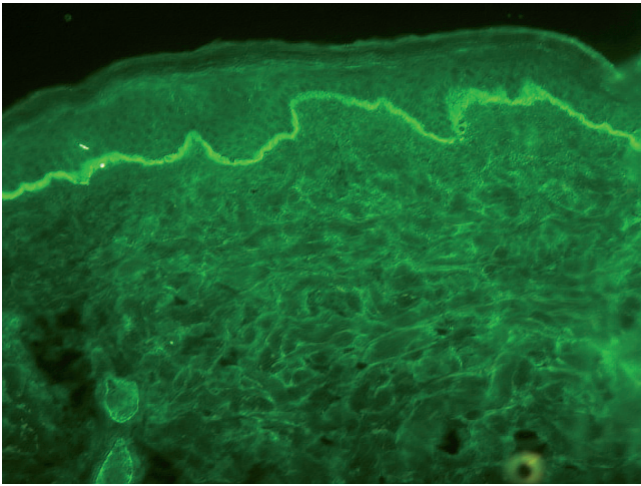


Fig. 2. Direct immunofluorescence test shows deposition of IgG at the dermo-epidermal junction (200 X). The biopsy was taken from the right palm.

In the majority of reported cases, DP is initially located on the palms and plantar skin and later involves other parts of the body. In the case described here the condition improved quickly and after one year of follow-up there was no sign of diffuse cutaneous involvement.

Topical steroids are reported to be effective as monotherapy in this condition, but usually a combination

with systemic corticosteroid or dapsone is necessary to keep the disease under control. In the case described here a short course of dapsone produced a rapid remission, while systemic corticosteroids were previously ineffective.

REFERENCES

1. Levine N, Abraham F, Barland P. Localized pemphigoid simulating dyshidrosiform dermatitis. *Arch Dermatol* 1979; 115: 320–321.
2. Forschner A, Fierlbeck G. Localized pemphigoid on the soles of both feet. *Int J Dermatol* 2005; 44: 312–314.
3. Patrizi A, Rizzoli L, Benassi L, Neri I. Another case of dyshidrosiform pemphigoid. *J Eur Acad Dermatol Venereol* 2003; 17: 370.
4. Sugimura C, Katsuura J, Moriue T, Matsuoka Y, Kubota Y. Dyshidrosiform pemphigoid: report of a case. *J Dermatol* 2003; 30: 525–529.
5. Kim YJ, Kim MY, Kim HO, Park YM. Dyshidrosiform bullous pemphigoid. *Acta Derm Venereol* 2004; 84: 253–254.
6. Forschner A, Fierlbeck G. Localized pemphigoid on the soles of both feet. *Int J Dermatol* 2005; 44: 312–314.
7. Seike M, Nakajima K, Ikeda M, Kodama H. Coexistence of nodular and dyshidrosiform pemphigoid. *J Dermatol* 2006; 33: 375–376.
8. Yasuda M, Miyachi Y, Utani A. Two cases of dyshidrosiform pemphigoid with different presentations. *Clin Exp Dermatol* 2009; 23: 1–3.