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Case report

Lichen planus pemphigoides in a child

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Abstract

Introduction: Lichen planus pemphigoides (LPP) is a rare autoimmune subepidermal blistering disease characterized by evolution of vesico-bullous skin lesions in patients with active lichen planus. We describe a case of LPP in a 12-year-old girl with clinical, histological and direct immunofluorescence findings.

Case report: A 12-year-old Moroccan girl presented, after sun burn, pruritic violaceous papules on hands and feet complicated by the apparition of bullous lesions on apparent normal skin and on lichenoid eruption. A white reticulated pattern was present on the oral mucosa. Histopathology of lichenoid papule and bulla was consistent with the diagnosis of LPP. Direct immunofluorescence of peribullous skin showed linear deposits of IgG and C3 at the basal membrane zone. Treatment with Dapsone was successful.

Discussion: LPP is exceptional in children; just fifteen cases were reported in the literature. This condition seems to be idiopathic. However, in rare cases it has been associated with some drugs or after PUVA therapy. In our patient, it was probably induced by prolonged sun exposure.

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Keywords: Lichen planus; Pemphigoides; Child

1. Introduction

Lichen planus pemphigoides (LPP) is a rare autoimmune subepidermal blistering disease that presents clinical and histological features of both lichen planus (LP) and bullous pemphigoid (BP) (Willsteed et al., 1991).

This disease is rare in adults and has been reported only occasionally in children (Cohen et al., 2009). We report a

case history of a 12-year-old girl with clinical, histological and direct immunofluorescence findings of LPP.

2. Case report

A 12-year-old Moroccan girl was admitted to our department with 6 weeks history of pruritic violaceous papules on hands and feet that gradually spread to limbs and trunk. Those cutaneous lesions were followed by appearance of blistering on her legs and forearm since a week. The patient had no past personal or familial medical history except erythematous sunburn in upper and lower extremities and a daily solar exposure in the beach during summer holidays, 2 months before her admission. The patient has denied cutaneous infection or drug intake recently.

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On clinical examination, she had widespread violaceous polygonal papules with numerous tense vesicles and erythematous bulla. The vesiculo-bullous lesions were seen both on apparent normal skin and on lichenoid eruption (Fig. 1). The Nikolsky's sign was negative. A white reticulated pattern was present on the oral mucosa. There was no nail involvement.

Laboratory investigations including full blood count, blood urea, electrolytes and liver function tests were normal. Hepatitis B and C serology were negative.

Histopathology of fresh bulla on lichenoid eruption was consistent with subepidermal blister containing lymphocytes, neutrophils and eosinophils, and lichen planus (irregular acanthosis, compact orthokeratosis, dense lichenoid lymphohistiocytic inflammatory infiltrate with vacuolar change, and pigmentary incontinence) (Fig. 2). Direct immunofluorescence of peribullous skin showed linear deposits of IgG and C3 at the basal membrane zone (BMZ). Indirect immunofluorescence, immunoblotting analyses and immunoelectron microscopy were not performed.

Based on all these findings, the diagnosis of lichen planus pemphigoides (LPP) was made. The patient received a daily bath with chlorhexidine solution, intact bulla was punctured and the resulting erosions were covered by silver sulfadiazine. Systemic treatment was started by Dapsone 50 mg daily after glucose-6-phosphate dehydrogenase

determination. Bullae formation and pruritus then ceased within 8 days. The blood workup for dapsone monitoring during treatment was normal and a decrease of level of hemoglobin by 1.5 g/dl was noted at the second week of drug intake. Dapsone was continued for 3 months until complete clearance of violaceous papules and absence of new lichen planus lesions. No relapse occurred after 9 months of follow up.

3. Discussion

LPP is a rare, acquired, autoimmune subepidermal bullous dermatosis characterized by tense bulla arising on lichen planus lesions and on uninvolved skin, histological demonstration of subepidermal bullae, and linear deposits of IgG and C3 along the BMZ on immunofluorescence of peribullous skin (Kuramoto et al., 2000).

LPP is rare in adults, and only 65 cases have been described in the literature (Zaraa et al., 2013). In children this condition is exceptional; indeed just fifteen cases were reported (Cohen et al., 2009; Zaraa et al., 2013; Ilknur et al., 2011). The male to female ratio in childhood is 3:1 (Cohen et al., 2009). The mean age is 11 years with the youngest case being that of a 2-year-old child (Duong et al., 2012).

Clinically, lichen planus lesions precede, between 2 weeks and 16 weeks, the appearance of bulla in all reported pediatric cases had a mean time of 7.9 weeks (Cohen et al., 2009). As seen in our patient, LPP is characterized by developing blisters on lichenoid pruritic lesions and on uninvolved skin. The distribution of the blisters shows a marked predilection for the distal extremities, and half of the pediatric cases had involvement of the palms or soles (Cohen et al., 2009). White retiform network on the oral mucosa has also been described in three cases prior to our case (Borrego Hernando et al., 1992; Harjai et al., 2006; Bouloc et al., 1998).

Differential diagnoses include bullous LP or association of LP with erythema multiforme (Zaraa et al., 2013), but histological and immunological confrontation analysis rectifies diagnosis by showing coexistence of histological features of LP and BP, and linear IgG and/or C3 deposits along BMZ on direct immune-fluorescence. Indirect immune-fluorescence can be positive and the presence of IgG antibodies to either one or both, BP180 and BP230 antigens can help to assess diagnosis (Zaraa et al., 2013).

We were not able to conduct ELISA or immunoblotting studies on our patient; however, our patient's clinical, histological and direct immunofluorescence features were compatible with LPP.

LPP is usually idiopathic, but some cases, especially in adults, have been reported in association with some conditions like some drugs [Chinese herbs, angiotensin-converting enzyme inhibitors (captopril, ramipril), or simvastatin] (Xu et al., 2008; Ben Salem et al., 2008) or PUVA therapy (Kuramoto et al., 2000). In pediatric



Fig. 1. Violaceous polygonal papules with numerous tense bullae and erosion on lower extremities.

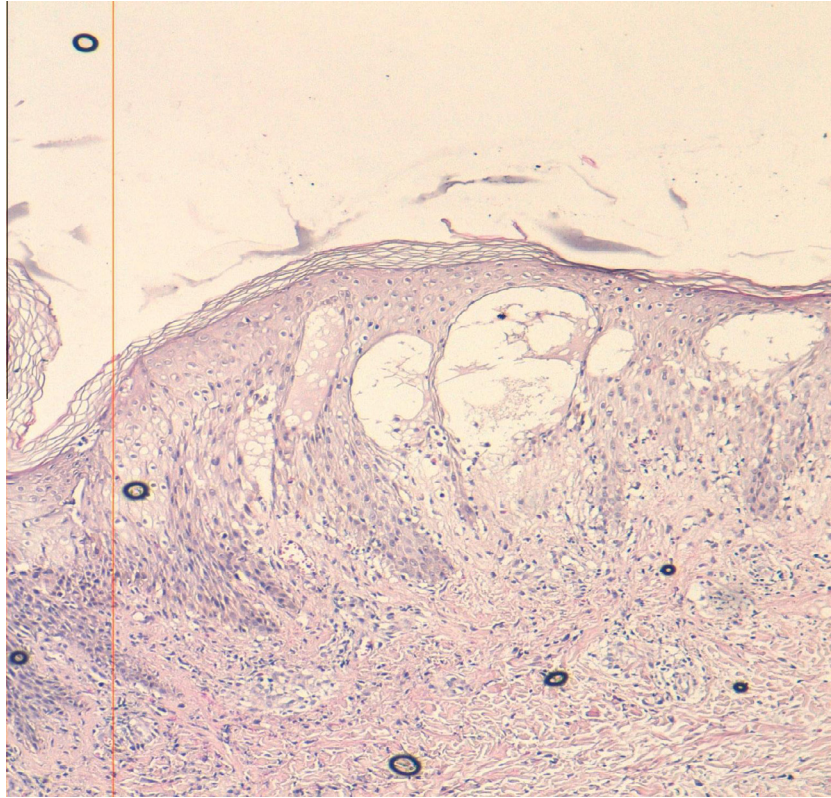


Fig. 2. Histopathology of bulla on lichenoid eruption showed subepidermal detachment, acanthosis, orthokeratosis, and dermal lymphocytic infiltrate with vacuolar change consistent with LPP. Hematoxylineosin stain; original magnifications $\times 20$.

cases, all seem to be idiopathic, and just one case has been described after varicella infection (Ilknur et al., 2011). The mechanism of the bullous eruption is still not clear; it has been explained by the phenomenon of epitope spreading. This concept was first proposed by Vanderlugt and Miller and suggested that a primary inflammatory process as in LP causes the release and exposure of a previously “sequestered” antigen, leading to a secondary autoimmune response against the newly released antigen (Vanderlugt and Miller, 1996). PUVA therapy was reported to expose BMZ components to autoreactive lymphocytes, thereby inducing autoimmunity against BMZ components (Kuramoto et al., 2000). Our patient may be an example of epitope spreading resulting from injuries by an inflammatory dermatitis and by prolonged exposure to ultraviolet: She does not have history of drug intake, but a daily sun exposure and the sun burn could be responsible for an alteration of dermal–epidermal junction and the appearance of LPP.

Unlike the adult in whom oral corticosteroid is the treatment of choice, topical corticosteroids or dapsone seem to be first-line therapies in children (Cohen et al., 2009). In case of failure of these drugs, oral corticosteroids or methotrexate can be used (Duong et al., 2012).

The prognosis of LPP seemed good. In a recent review of all LPP cases, the rate of recurrence was about 20% in adults and 23% in children (Zaraa et al., 2013).

In conclusion, we report the first Moroccan case of LPP. This condition is exceptional in children and was probably induced by sun exposure during summer in our patient.

Conflict of interest

None declared.

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References

- Ben Salem, C., Chenguel, L., Ghariani, N., Denguezli, M., Hmouda, H., Bouraoui, K., 2008. Captopril-induced lichen planus pemphigoides. *Pharmacoepidemiol. Drug Saf.* 17, 722–724.
- Borrego Hernando, L., Vanaclocha Sebastian, F., Hergueta Sanchez, J., Ortiz Romero, P., Iglesias Diez, L., 1992. Lichen planus pemphigoides in a 10-year-old girl. *J. Am. Acad. Dermatol.* 26 (1), 124–125.
- Bouloc, A., Vignon-Pennamen, M.D., Caux, F., Teillac, D., Wechsler, J., Heller, M., Lebbé, C., Flageul, B., Morel, P., Dubertret, L., Prost, C., 1998. Lichen planus pemphigoides is a heterogeneous disease: a report of five cases studied by immunoelectron microscopy. *Br. J. Dermatol.* 138, 972–980.
- Cohen, D.M., Ben-Amitai, D., Feinmesser, M., Zvulunov, A., 2009. Childhood lichen planus pemphigoides: a case report and review of the literature. *Pediatr. Dermatol.* 26 (5), 569–574.
- Duong, B., Marks, S., Sami, N., Theos, A., 2012. Lichen planus pemphigoides in a 2-year-old girl: response to treatment with methotrexate. *J. Am. Acad. Dermatol.* 67 (4), e154–156.

- Harjai, B., Mendiratta, V., Kakkar, S., Koranne, R.V., 2006. Childhood lichen planus pemphigoides – a rare entity. *J. Eur. Acad. Dermatol.* 20, 117–118.
- Ilknur, T., Akarsu, S., Uzun, S., Ozer, E., Fetil, E., 2011. Heterogeneous disease: a child case of lichen planus pemphigoides triggered by varicella. *J. Dermatol.* 38 (7), 707–710.
- Kuramoto, N., Kishimoto, S., Shibagaki, R., Yasuno, H., 2000. PUVA-induced lichen planus pemphigoides. *Br. J. Dermatol.* 142 (3), 509–512.
- Vanderlugt, C.J., Miller, S.D., 1996. Epitope spreading. *Curr. Opin. Immunol.* 8, 831–836.
- Willsteed, E., Bhogal, B.S., Das, A.K., Wojnarowska, F., Black, M.M., McKee, P.H., 1991. Lichen planus pemphigoides: a clinicopathological study of nine cases. *Histopathology* 19 (2), 147–154.
- Xu, H.H., Xiao, T., He, C.D., Jin, G.Y., Wang, Y.K., Gao, X.H., Chen, H.D., 2008. Lichen planus pemphigoides associated with Chinese herbs. *Clin. Exp. Dermatol.* 34, 329–332.
- Zaraa, I., Mahfoud, A., Kallel Sellami, M., Chelly, I., El Euch, D., Zitouna, M., Mokni, M., Makni, S., Ben Osman, A., 2013. Lichen planus pemphigoides: four new cases and a review of the literature. *Int. J. Dermatol.* 52 (4), 406–412.