

A Pediatric Case of Erythrodermic Lichen Planus Pemphigoides

Safae Maouni^{*}, El Anzi Ouiam, Sqalli Asmae, Zenati Kaoutar, Meziane Mariam, Senouci Karima and Hassam Badredine

Department of Dermatology-Venereology, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco

^{*}Corresponding author: Safae Maouni, Department of Dermatology-Venereology, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco, Tel: +212635342053; E-mail: maouni.safae88@gmail.com

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Abstract

Lichen planus pemphigoides (LPP) is an uncommon skin condition that is characterized by signs of lichen planus associated to subepidermal detachment with deposits of autoantibodies along dermal-epidermal junction basement membrane. LPP is infrequent in adults, but extremely rare in pediatric population; indeed just 17 cases were reported. We report a rare case history of a 6-year-old boy with clinical and histological findings of an erythrodermic LPP.

Keywords: Lichen planus pemphigoides; Child; Erythrodermia; Subepidermal bullous dermatosis

Introduction

Lichen planus pemphigoides (LPP) is an uncommon skin condition that is characterized by signs of lichen planus associated to subepidermal detachment with deposits of autoantibodies along dermal-epidermal junction basement membrane. LPP is infrequent in adults, but extremely rare in pediatric population; indeed just 17 cases were reported. We report a rare case history of a 6-year-old boy with clinical and histological findings of LPP.

Clinical Case

This is a 6 years old boy, who consulted in our dermatology department for pruritic erythrodermia. There was no particular medical history apart a pharyngitis that appeared 3 weeks before an eruption made of papules on the hands and feet, evolving in erythrodermia in 3 months, with blisters appearing. The dermatological examination found a patient with dry erythrodermia, with the presence of purplish papular lesions on the hands and wrists, as well as tense blisters and post-bullous erosions in the thighs and legs (Figure 1). Nikolsky's sign was negative. The examination of the oral mucosa found a reticular pattern on the cheeks. The examination of the skin appendage found a scaly condition of the scalp without nail disorder. Histological examination of a skin biopsy performed on a recent bullous lesion revealed a subepidermal blister containing neutrophils associated to dense lichenoid lymphohistiocytic inflammatory infiltrate and pigment incontinence (Figure 2). Direct immunofluorescence showed linear IgG deposits along the basement membrane. Indirect immunofluorescence was positive (1/160).

According to these clinical and histological features, the diagnosis of lichen planus pemphigoides in its erythrodermic form was made. The serologies of hepatitis B and C were negative. The patient received systemic corticosteroid (prednisolone) treatment at 0.5 mg/kg/day, with good clinical progress and dryness of the bullous lesions. No recurrence was noted after 4 months of follow-up.



Figure 1: Erythrodermia in a 6-year-old boy (a) with tense bullae and erosion on the thigh (b).

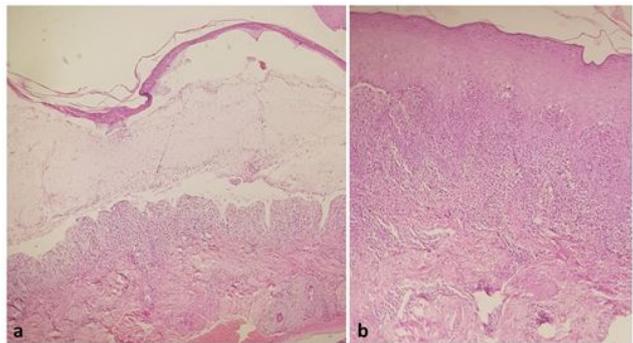


Figure 2: Cutaneous histology (HE, G x 4), subepidermal blister containing neutrophils (a), dense lichenoid lymphohistiocytic inflammatory infiltrate (b).

Discussion

LPP is a rare, acquired, autoimmune subepidermal bullous disorder, characterized by the co-existence of both signs of lichen planus and bullous pemphigoid in the same patient.

LPP affects mostly adults, while it is unusual in children; only 17 cases have been reported in the literature. Male to female sex ratio is 1 [1]. The lowest age reported in child is 2-year-old [2].

Clinically, LPP is characterized first by lichenoid pruritic lesions and after 2 weeks to 16 weeks, bullous lesions appear on lichen planus lesions or on uninvolved skin [1]. The erythrodermic form is rare, it has been described in just 11 cases in adult, but it has never been reported in the literature in children. Our patient was admitted with erythrodermia occurring 3 months after the first eruption. The presence of oral lichen was noted in 4 cases before ours.

Histologically, there is a co-existence of the signs of lichen planus (lympho-histiocytic infiltrate with pigment incontinence) and bullous pemphigoid (detachment subepidermal), with the presence of linear deposits of IgG and/or C3 along the junction of the basement membrane [3]. Serum IgG antibodies directed against the PB180 antigen or PB230 can be demonstrated.

The exact etiology of LPP is still unknown, it is usually idiopathic. In children, 2 cases were triggered by chickenpox [4,5] another case after exposure to the sun [6]. In our patient, LPP seems to be triggered by the episode of pharyngitis that preceded the rash by 3 weeks.

The pathophysiology of this dermatosis is still unclear, it can be explained by the phenomenon of "epitopic spreading", that was proposed by Vanderlugt and Miller in 1996 [7]. Indeed there is a sensitization of autoreactive B and T cells during a chronic autoimmune or inflammatory response, as in the lichen planus, which leads to a production of auto-antibodies directed against the components of the basement membrane such as Ag BP 180, ultimately responsible for the onset of bullous lesions [8].

In children, the treatment of LPP is mainly based on dapsone and dermocorticoids in the first place and oral corticosteroid therapy or methotrexate in second intention [1]. Generally, the LPP has a good prognosis, with a recidivism rate estimated at 23% in children.

Conclusion

We report the first case of erythrodermic LPP in children. It is a serious form that poses a problem of differential diagnosis with the other causes of acquired erythrodermia in child and which requires an early and adequate care to avoid complications.

References

1. Cohen DM, Ben-Amitai D, Feinmesser M, Zvulunov A (2009) Childhood lichen planus pemphigoides: a case report and review of the literature. *Pediatr Dermatol* 26: 569-574.
2. 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: 154-156.
3. Zaraa I, Mahfoud A, Sellami MK, Chelly I, El Euch D, et al. (2013) Lichen planus pemphigoides: four new cases and a review of the literature. *Int J Dermatol* 52: 406-412.
4. 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: 707-710.
5. TS Mohanarao, Kumar GA, Chennamsetty K, Priyadarshini T (2014) Childhood lichen planus pemphigoides triggered by chickenpox. *Indian Dermatol Online J* 5: 98-100.
6. Meziane M, Lakjiri S, Harmouch T, Mikou O, Mernissi FZ (2014) Lichen planus pemphigoides in a child. *Journal of Dermatology & Dermatologic Surgery* 18: 31-34.
7. Vanderlugt CJ, Miller SD (1996) Epitope spreading. *Curr Opin Immunol* 8: 831-836.
8. Mignogna MD, Fortuna G, Leuci S, Stasio L, Mezza E, et al. (2010) Lichen planus pemphigoides, a possible example of epitope spreading. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 109: 837-843.