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Bullous lichen planus in childhood – A case report

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VERSITA

Abstract

In Europe, only 1% of all patients with lichen planus are children. We report a case of lichen planus in a 5-year-old boy with blisters developing on papules. There was no history of Hepatitis B virus vaccination in the past 12 months. Routine laboratory analyses showed no abnormalities. Hepatitis B surface antigen and anti-Hepatitis C virus antibodies were negative. Our patient had disseminated and coalesced papules on the trunk and extremities with vesicles and bullae on the hands and feet. Histopathology confirmed the diagnosis of lichen planus and bullous lichen planus. Negative direct immunofluorscence test excluded lichen planus pemphigoides. The boy was treated with prednisone 1 mg/kg/day (the dose was gradually tapered and discontinued over the next 1.5 month), ultraviolet B phototherapy, fluocinolon acetonide, and topical pimecrolimus 1% cream till complete remission after 2.5 months. Bullous form of lichen planus is seen in 1-16% of all children with lichen planus. Two months upon the completion of therapy, there were no signs of relapse. In our case, short course of systemic corticosteroids and ultraviolet B phototherapy have been safe and effective. This was the only pediatric case of bullous lichen planus treated in our Clinic in the last 20 years. Long-term prognosis of childhood lichen planus is not predictable, and there is no consensus regarding the treatment of childhood lichen planus.

Key words

Child, Preschool; Skin Diseases, Vesicolobullous; Lichen Planus; Prednisone; Phototherapy

The term "lichen" describes discrete flat skin eruptions, or an aggregate of papules, giving a patterned configuration resembling lichens commonly found growing on rocks. Lichen planus (LP), first described in 1869 by Wilson, characteristically consists of very itchy eruptions containing flat-topped, polygonal and violaceous papules with fine linear white scales, referred to as Wickham's striae. The mucous membranes, especially the oral mucosa, may be affected. There are many clinical variants of LP: actinic, annular, atrophic, LP hypertrophic (LPH), guttate, linear, LP pigmentosus, erosive (ulcerative), follicular (lichen planopilaris), bullous LP and LP *pemphigoides* (LPP).

The etiology of LP remains unclear, although there are many theories: autoimmune mechanisms, association with certain HLA haplotypes and liver disease - Hepatitis C virus (HCV) infection (1) and Hepatitis B virus (HBV) vaccine (2, 3).

Case report

We report a case of a 5-year-old boy with a history of atopic dermatitis since the age of 3. A month and a half before admission, the first lesions appeared on the trunk and extremities. Otherwise, the boy was in good general health. Immunization was performed regularly, but not in the last 12 months. The family history was negative for LP. On admission, the lesions were disseminated all over the skin, except on the face and oral mucous membranes, with bilaterally symmetrical distribution. The eruptions consisted of flat-topped, polygonal, violaceous papules with Wickham's striae (Figure 1). They were a few millimeters in size, coalesced into plaques, predominantly on lower back,

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dorsal parts of hands and lower legs. Scalp skin was involved with whitish non-adherent scales. On several regions, linear papules (Koebner phenomenon) were noticed (Figure 2, 4). On hands and feet, bullae developed on papules and plaques (Figure 3). Bullae were hemorrhagic on the palmar surfaces (Figure 4).

Routine laboratory tests showed no abnormalities. HBsAg and anti-HCV antibodies were negative. Histopathologic examination revealed orthohyperkeratosis with hypergranulosis, irregular acanthosis with „sawtoothing" of the rete ridges, vacuolization of the basal cell layer, with Civatte bodies, and a dense, band-like infiltrate (Figure 5). Pigmentary incontinence was conspicuous under the epidermo-dermal junction (Figure 5, 6). In some segments, Max-Joseph spaces were progressing into frank subepidermal bullae (Figure 6). Direct immunofluorescence (DIF) test was negative.

The boy was treated with prednisone 1 mg/kg/day, broad band ultraviolet B (BB-UVB) radiation 4 times a week, (cumulative dose of 1145mJ/cm^2), topical fluocinolone acetonide ointment (0.025% and 0.2%) and pimecrolimus 1% cream. Prednisone dose was gradually tapered and discontinued over the next 1.5 month, but topical therapy was continued for one more month, until complete regression of lesions. The last check-up was performed 2 months after discontinuation of complete therapy, and until now, the boy presented no signs of relapse.

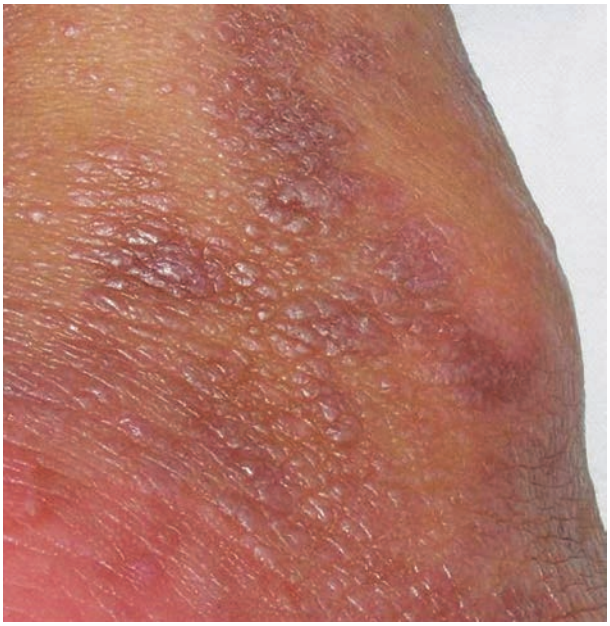

Figure 1. Flat-topped, polygonal, violaceous papules with Wickham's striae.

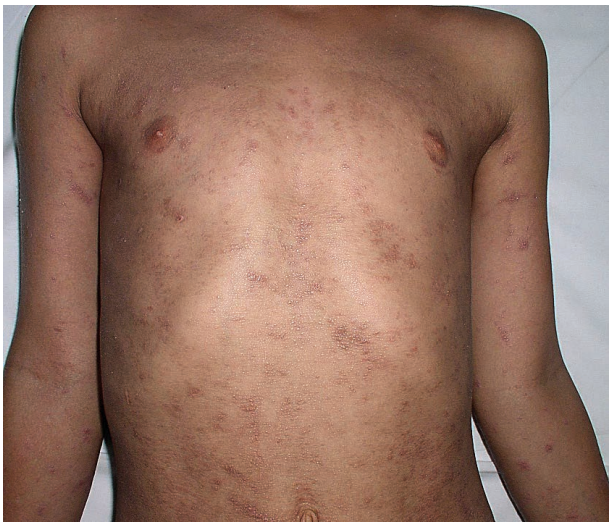

Figure 2. Disseminated violaceous papules and Koebner phenomenon on the trunk and arms.

Discussion

LP is an uncommon skin disease in childhood. It is most frequently found in patients aged 30 - 60 years (4). There are only a few studies about childhood LP. In Kanwar's study (5), the earliest age of onset was 2 weeks of age. According to major published studies, the age of onset varied from 5 months to 13 years (6-8), with slight male predominance 1.1 - 3 : 1 (5, 6, 9). LP starting in childhood accounts for 1% in London

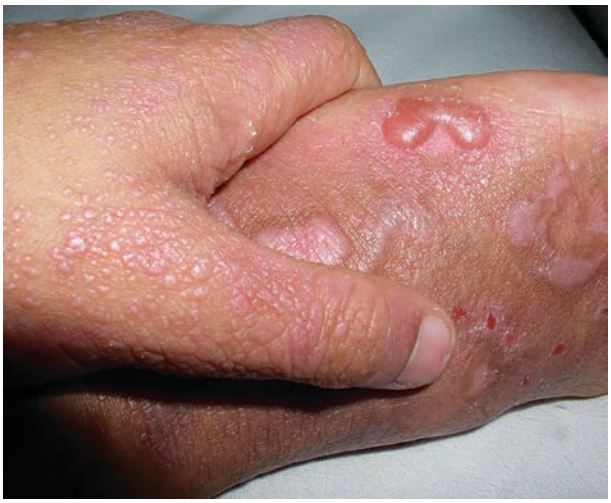

Figure 3. Papules on the hand, bullae surmounting papules on the foot.

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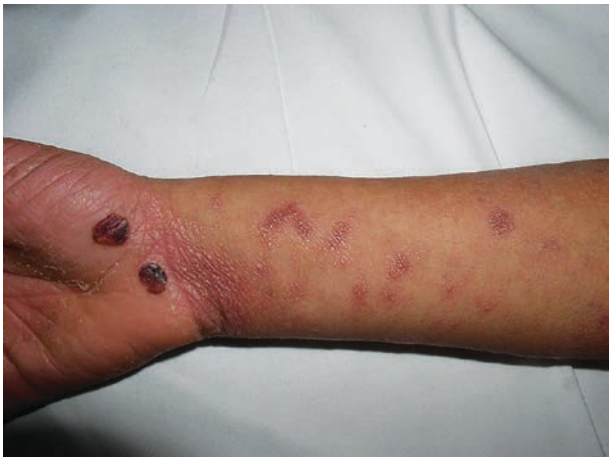

Figure 4. Hemorrhagic bullae on the palmar surface.

(10), 7.5% among Libyans (11), and 17.2% in India (12). Most studies of LP in children have been from India. There is an increased incidence of childhood LP in the Tropics and Subtropics (7, 12-14). Bullous LP occurs in 1 - 16% of all children with LP (5, 6, 15).

Due to the lack of long-term follow-up, often seen in retrospective studies, it is difficult to draw an inference about the most effective treatment and long-term prognosis of LP in children. Topical corticosteroids remain the treatment of choice in most patients with

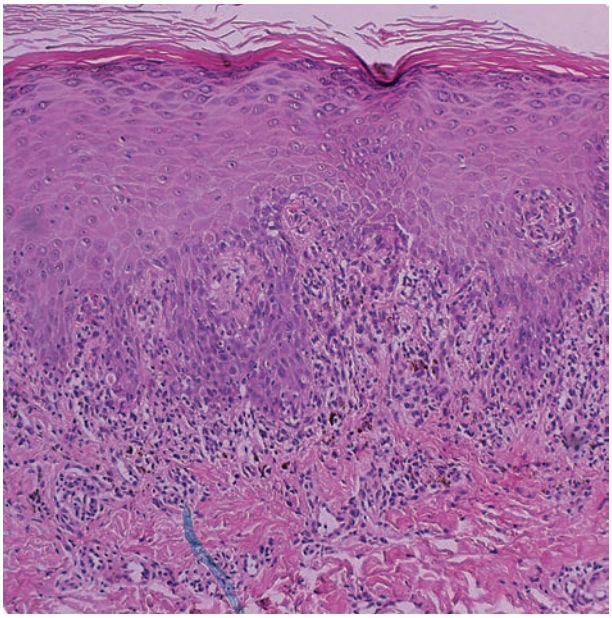

Figure 5. Orthohyperkeratosis, hypergranulosis, irregular acanthosis, vacuolization of the basal cell layer, pigmentary incontinence and a dense band-like infiltrate (H&E, x100)

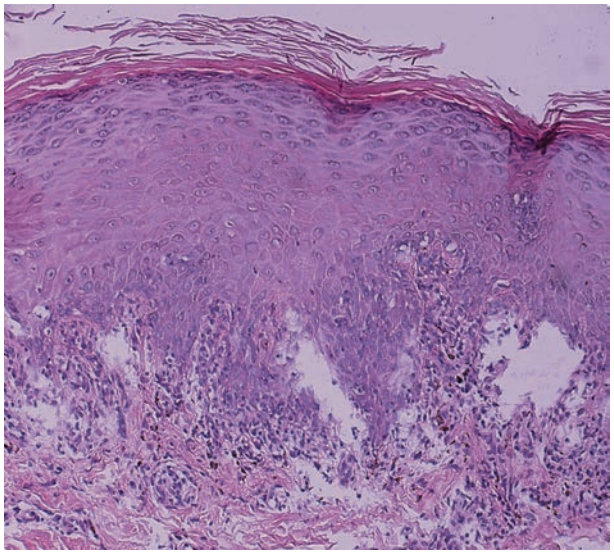

Figure 6. Subepidermal blister formation and dense band-like infiltrate with pigmentary incontinence (H&E, x100)

localized lesions (5, 16). The UVB phototherapy (8) is safe and effective in children with acute widespread LP. Oral corticosteroids (prednisolone 1 mg⁄kg/day) are given to patients with extensive/eruptive lesions (5, 6, 17). Dapsone is useful in patients with chronic, recurrent LP, and in disease control after the activity of disease is controlled by systemic corticosteroids (5, 18). Currently, the first-line treatment of mucosal lesions and localized cutaneous LP consists of potent topical corticosteroids, although recent clinical reports indicate that topical tacrolimus might be an effective treatment alternative (19, 20). Tazarotene gel 0.05% (5) is used topically on the periungual folds if few nails are involved. In contrast, due to the lack of controlled clinical trials, generalized eruptions or more severe types of cutaneous LP are still treated with a broad spectrum of various systemic agents, acitretin and oral corticosteroids (21). Furthermore, immunosuppressants e.g., mycophenolate mofetil (22), azathioprine (21), methotrexate (23), PUVA (24), low-molecular-weight heparin (25), and T-cell receptor mimic peptides (26) have been successfully used in adults. Still, it is important to keep in mind that the disease may have a spontaneous remission.

Bullous eruptions in LP were first described in 1892 by Kaposi (27), and since then, two distinct forms of LP with bullae have been described. Bullous LP is characterized by blister formation on LP lesions,

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caused directly by upper dermal inflammation and liquefactive degeneration of the basal cell layer. By contrast, *lichen planus pemphigoides* (LPP) is marked by bullous eruptions on both LP lesions and normal skin. On direct immunofluorescence (DIF), histopathology of LPP reveals sub-epidermal blisters with linear deposition of IgG and/or C3 along the dermal–epidermal junction. In addition, circulating autoantibodies against epidermal basement membrane zone (BMZ) components are often found using indirect immunofluorescence (IIF) (28). In bullous LP, DIF does not show deposition of antibodies along BMZ, but IIF, using patient serum and autologous perilesional skin, may show deposition of IgG and IgA in the stratum granulosum and stratum spinosum (29 - 31). Our patient had a typical clinical presentation, and representative histopathology of bullous LP (9, 32). Negative DIF test excluded lichen planus pemphigoides.

The long-term prognosis of childhood LP is uncertain. It is limited by a small number of reported cases. Handa and Sahoo analyzed records of 87 children with LP, and most patients with classical LP showed clearance of lesions within 6 months, except for postinflammatory pigmentation that persisted in all patients (6). Uncommon variants, such as mucosal (buccal) LP, LPH, and lichen planopilaris, required a prolonged therapy up to 1 year (6). In the study of Sharma and Maheshwari, the majority of patients (97.8%) cleared in less than 1 year (9). Only one child had recurrent relapses during the follow-up period of 6 months (9).

In our population, bullous lichen planus in children seems to be very rare, since in the last 20 years we had only one patient with this presentation.

Conclusion

LP, especially bullous LP, is a rare disease during childhood. There are few studies about LP in childhood, its therapy and outcome. We presented a 5-year-old boy with bullous LP, who responded well to a short course of systemic corticosteroid therapy, and ultraviolet B phototherapy. We believe that any information about treatment outcome of bullous LP during childhood should be widely reported in order to achieve consensus on the treatment protocol for this uncommon form of the disease.

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Abbreviations

LP - Lichen planus
LPP – *Lichen planus pemphigoides*
HCV - Hepatitis C virus
HBV - Hepatitis B virus
BLP – Bullous lichen planus
DIF – Direct immunofluorescence test
UVB – Ultraviolet B phototherapy
BMZ – Basement membrane zone
IIF – Indirect immunofluorescence

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Bulozni lichen planus u dečijem uzrastu

Sažetak

Uvod: U Evropi, samo 1% obolelih od lichen planusa su deca. Bulozna forma se viđa kod 1-16% obolele dece.
Prikaz obolelog: Prikazujemo dečaka uzrasta 5 godina obolelog od lichen planusa, kod koga je na papulama došlo do razvoja bula. U prethodnih 12 meseci nije bio vakcinisan protiv hepatitisa B. Rutinske laboratorijske analize bile su normalne. Hepatitis B površinski (eng. *surface*) antigen i antitela protiv Hepatitisa C virusa su bili negativni. Papule su bile diseminovane na trupu i ekstremitetima, sa tendencijom da se slivaju u plakove. Na šakama i stopalima su postojale vezikule i bule na papulama i malim plakovima.
Histopatološka analiza: Histopatologija je potvrdila dijagnozu buloznog lichen planusa. Negativni test direktne imunofluorescencije je isključio lichen planus pemphigoides.
Lečenje: Dečak je lečen prednizonom u dozi 1 mg/kg (doza je postepeno snižavana do isključenja nakon 1,5 mesec), UVB fototerapijom, lokalno fluocinolon acetonidom i pimekrolimus 1% kremom do potpune remisije posle 2,5 meseca. Dva meseca po završetku terapije nije bilo znakova relapsa.
Zaključak: U našem slučaju primena opšte kortikosteroidne terapije i UVB fototerapije pokazale su se uspešnim i bezbednim. Ovo je jedini slučaj buloznog lichen planusa, u dečjem uzrastu koji je lečen u našoj klinici za proteklih 20 godina. Dugoročna prognoza lichen planusa, u dečjem uzrastu nije poznata. Ne postoji konsenzus o terapiji lichen planusa, u dečjem uzrastu.

Ključne reči

Predškolsko dete; Vezikulobulozne bolesti kože; Lihen planus; Prednizon; Fototerapija

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