

PAEDIATRIC DERMATOLOGY

LICHEN PLANUS PEMPHIGOIDES: A PEDIATRIC CASE

M Soua (1) - Y Soua (1) - H Belhadiali (1) - M Daldoul (1) - L Njim (2) - M Youssef (1) - J Zili (1)

Fattouma Bourguiba Hospital, Dermatology, Monastir, Tunisia (1) - Fattouma Bourguiba Hospital, Anatomopathology, Monastir, Tunisia (2)

Background: Lichen planus pemphigoides (LPP) is a rare autoimmune blistering disease, exceptionally described in children. It's characterized by an acute onset of lichen planus followed by the development of blisters on both involved and on uninvolved skin.

Observation: A 14 year-old child, with no medical history, consulted our department, for generalized itchy lesions evolving since few months. Physical examination revealed the presence of widespread and symmetric erythemato-violaceous maculo-papules on the trunk and extremities. Hyperkeratotic plaques with fissures were present on both palms and soles. Flask bullae were present on lichenoid area, but not on the uninvolved skin. The nails were spared, and there were no mucous membrane involvement. A cutaneous biopsy revealed a lichenoid lymphocytic infiltrate associated with sub epidermal blisters. Direct immunofluorescence (DIF) on the perilesional skin revealed linear deposits of IgG and C3 along the basement membrane zone. Routine laboratory tests were within normal range, and were negative for hepatitis A, B and C. Because of his young age, the patient was treated with topical corticosteroids and showed significant clinical improvement within a span of 10 days.

Key message: LPP is a controversial rare entity, which was first described by Kaposi in 1892. LPP usually occurs on adults. Only fifteen pediatric cases are reported. The pathogenies of LPP is incompletely elucidated. It is typically characterized by blisters appearing on active lichen planus lesions and on healthy skin. On our case, bullous lesions were only present on affected skin. The diagnosis of LPP is confirmed by histopathological examination revealing the concurrence of both lichen planus and sub epidermal bullae with positive DIF. The main differential diagnosis is bullous lichen planus, which is characterized by development of blisters only on lichenoid lesions with negative DIF. Treatment options include topical corticosteroids, oral corticosteroids and azathioprine, depending on the severity of symptoms.





