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Lymphoplasmacytic plaque in children: Case report and literature review

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Abstract

We report a case of benign lymphoplasmacytic plaque (LPP) in a child. These asymptomatic erythematous papulonodular lesions are an emerging clinicopathological entity. Herein, we describe a previously unreported site for LPP lesions, namely, the volar wrist and the distal ipsilateral palm.

KEYWORDS
lymphoplasmacytic plaque, pseudolymphoma

1 | INTRODUCTION

Lymphoplasmacytic plaque in children (LPP) is a rare cutaneous disease. It has been recently proposed as an emerging clinicopathologic entity characterized by a dense dermal lymphoplasmacytic infiltrate with or without epithelioid granulomas and is considered a plasma cell pseudolymphomatous process of unknown origin.[1] These lesions are clinically determined by reddish-brown papules and plaques, which are typically found on the lower limbs, especially on the anterior shin.

2 | CASE REPORT

A 4-year-old boy presented with skin lesions on his right wrist of 4 months' duration. There were erythematous papules that coalesced in a linear pattern on the volar distal right wrist, along with some papular lesions grouped into clusters on the dorsal and palmar aspects of the ipsilateral hand. These lesions were asymptomatic and stable since onset. There were no other lesions, and the patient was otherwise healthy (Figure 1).

Histopathology of lesional skin revealed a dense lymphoplasmacytic infiltrate occupying the entire dermis, including some epithelioid granulomas without necrosis (Figure 2). CD138 immunostaining confirmed the prominent plasmacytic nature of the dermal infiltrate, which extended perivascularly into the deep dermis and the superficial layer of the subcutis (Figure 3). Lack of extension and organization of the lymphoplasmacytic infiltrate around adnexal structures in the dermis, such as the eccrine glands, excluded the diagnosis of lichen striatus, which is the clinical differential diagnosis.

Additional immunohistochemistry showed a polyclonal pattern of immunoglobulin light chain expression in the plasma cell population. Most of the plasma cells expressed IgG, and only a minority of plasma cells expressed IgG4. Serum immunoglobulin levels were normal. Finally, in situ hybridization was performed to assess the expression of Ig kappa and lambda light chains; it confirmed the absence of dysglobulinemia.

PAS, Giemsa, Ziehl-Neelsen, and Warthin-Starry stains were negative. In addition, Treponema pallidum, Borrelia burgdorferi, and human herpesvirus 8 virus were not detected in immunohistochemical studies of tissues. Serological tests for Treponema pallidum and Borrelia burgdorferi were also negative.

Based on the clinical features and histopathology, lymphoplasmacytic plaque was diagnosed.

3 | DISCUSSION

Initially described in the literature as "pretibial lymphoplasmacytic plaque,"[1] LPP is a benign chronic dermatosis without systemic involvement that primarily affects children.

Hence, there is no systemic plasmacytosis in these children. The lesions are typically stable and asymptomatic, but are sometimes slightly pruritic. Most patients are young, healthy Caucasian

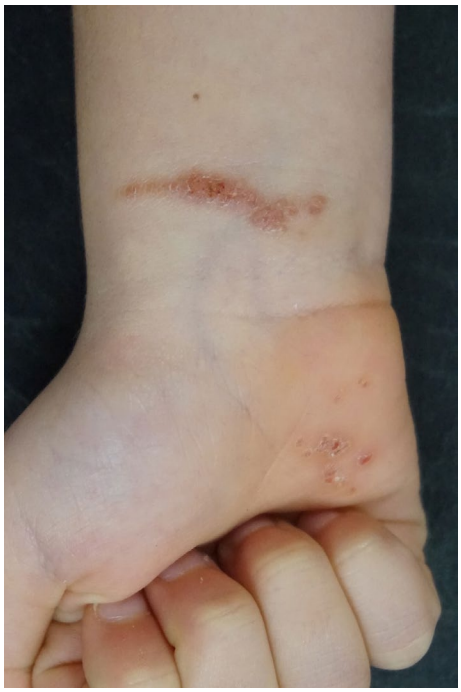

FIGURE 1 Clinical characteristics of lymphoplasmacytic plaque in a 4-y-old boy. Erythematous papules converging in a linear pattern were found on the volar wrist

on the wrist and ipsilateral hand, as our 4-year-old patient presented, have not been previously described. The diagnosis of LPP is based on clinicopathologic correlation.[3] The characteristic clinical presentation reveals erythematous, brownish papules, and plaques, often grouped in a linear pattern. Histologically, a dermal lymphohistiocytic infiltrate with numerous polyclonal plasma cells is commonly found in all LPP lesions.[2] These histologic features of LPP are similar to those of acral pseudolymphomatous angiokeratoma of children, also known as APACHE.[2] This suggests that LPP and APACHE might belong to the same spectrum of diseases, namely, pseudolymphomas. The differences between the two are clinical. While LPP commonly presents as a reddish-brown plaque, APACHE most often presents as grouped red papules separated from one another. In addition, APACHE has a predilection for the extremities, especially the legs and hands.

In the presence of a diffuse lymphoplasmacytic infiltrate, an infectious cause should be excluded. The patient should be tested for syphilis and leishmaniasis. If epithelioid granulomas are found within the lymphoplasmacytic infiltrate, mycobacteria and fungal infections must also be ruled out. Special staining, such as PAS (fungal infection), Giemsa (leishmania), Ziehl (mycobacteria), and Warthin-Starry (spirochetes), should be performed, along with blood serologies. Fresh tissue cultures or PCR-based studies can be added if

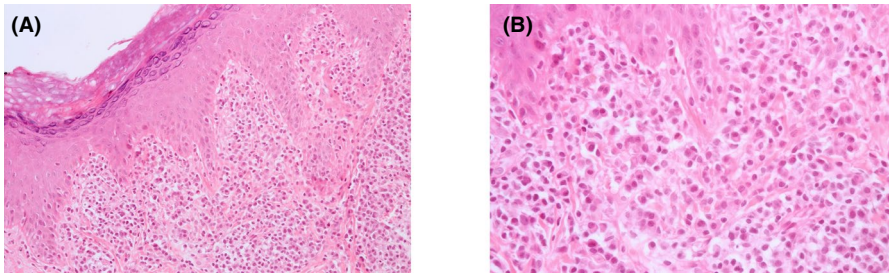

FIGURE 2 Histopathology revealed a dense lymphoplasmacytic infiltrate at the dermoepidermal junction (H&E, ×200) (A). Characterization of the dermal infiltrate with admixed plasma cells at a higher magnification (H&E, ×400) (B)

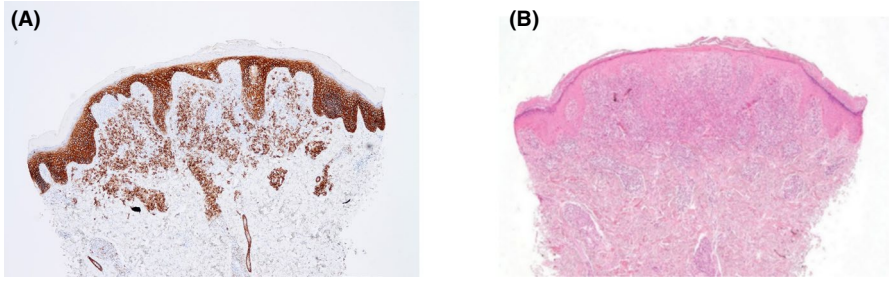

FIGURE 3 Prominent polyclonal plasmacytic nature of the dermal infiltrate demonstrated by immunohistochemical staining for the plasma cell marker CD138 (CD138, ×100) (A). Typical histological features with a dense lymphoplasmacytic infiltrate occupying the entire dermis (H&E, ×100). (B)

girls; the disorder affects a ratio of one male to five females. This cutaneous disorder is very rare, with only 16 cases reported in the literature to date; four of the cases were patients over 18 years of age.[2] As the initial description notes, LPP has a predilection for the lower legs, particularly the pretibial area. However, there are recent reports of LPP on the upper limbs, buttocks, and trunk. Only four cases of upper limb involvement have been described, including three on the arm and one on the middle finger.[2] Lesions

necessary.[2] It is also essential to exclude a monoclonal plasma cell population by immunohistochemistry and FISH analysis.[2] Indeed, the differential diagnosis of lymphoplasmacytic infiltrates includes plasmacytoma, lymphocytoma cutis, cutaneous marginal zone lymphoma, and primary cutaneous plasmacytosis. In these cases, in situ hybridization reveals a light chain-restricted plasma cell population with a monoclonal immunoglobulin heavy chain rearrangement. IgG4-related disease must also be considered a differential

diagnosis. The low number of plasma cells expressing IgG4 argues against an IgG4-related disease in this case.

The pathogenesis of these lymphoplasmacytic infiltrates is still unknown, and its course seems to be chronic with clinical stability. No cases of spontaneous resolution have been reported in the literature to date. The longest reported duration of LPP lesions was for lymphoplasmacytic plaques on the thigh and lower leg of two Caucasian girls; these plaques lasted for more than 10 years, with minimal changes in clinical appearance and no systemic involvement.[2,3]

The proposed treatments include diverse conservative approaches and are all anecdotal in nature. Topical or injected steroids can produce slight improvement but are not curative.[1-3] Pulsed dye laser treatment can reduce the intensity of the color to a moderate degree.[1] Only surgical excision of the lesions have led to a complete cure.[1] Except for purely cosmetic reasons, there are no indications for surgery since there is no risk of progression or long-term consequences.

4 | CONCLUSION

Lymphoplasmacytic plaque in children is an emerging clinicopathological entity whose criteria continue to evolve. LPP is currently considered a plasma cell-rich pseudolymphoma. A recently published analysis of the distribution of infiltrating immune cells in LPP describes a distinct compartmentalization of the immune cells, which is a characteristic feature of the disease.[4] Immunohistochemistry reveals CD 20 + B cells confined to the upper dermis, while CD3+ T cells are distributed mainly in the upper to middle dermis. Mature plasma cells are present throughout the dermis. This distribution of immune cells may be a clue toward understanding its pathogenesis and etiology, potentially leading to the classification of LPP as a distinct dermatosis in the future. The clinical presentation of papules converging in a linear plaque or clusters in children should suggest the diagnosis of benign lymphoplasmacytic plaque. Biopsy and ancillary microbiological molecular genetic studies are essential to exclude other diagnoses.

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