SHORT COMMUNICATION

A Long Way to Diagnosis: Linear IgA Dermatosis Mimicking Prurigo Pigmentosa

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Linear IgA dermatosis (LAD) is a rare subepidermal autoimmune bullous disorder (AIBD) defined by predominant or exclusive deposits of immunoglobulin A along the basal membrane zone of the skin or mucous membranes (1–4). Prurigo pigmentosa (PP) is an inflammatory disease of the skin, increasingly recognized outside East Asia. It manifests as recurrent, intensely itchy papules and plaques that resolve with reticulated hyperpigmentation, most commonly involving the trunk in symmetrical distribution. Histopathological findings vary depending on the stage of the disease; however, in early stages, neutrophilic granulocytes may predominate within the infiltrate, mimicking prebullous stages of AIBD (5–7).

CASE REPORT

A 51-year-old Caucasian man presented with disseminated, severely itchy (NRS 7/10) skin lesions involving the trunk and scalp, persisting for 2 months. The patient was otherwise healthy, and no mucosal involvement was observed. Initially, numerous scattered, slightly infiltrated plaques and crusted erosions were confined to the back but gradually spread to the chest and arms. Notably, no blisters were present (**Fig. 1**A). Previous therapies included systemic anti-inflammatory treatment with prednisolone, topical clobetasol and an immunomodulatory regimen with doxycycline resulting in partial band temporary improvement. Lesions followed a chronic course and resolved with residual reticulated hyperpigmentation.

The first histopathological examination revealed ulceration consistent with a prurigo-like disorder (Fig. 1B–C). Both perilesional direct immunofluorescence (IF) and indirect IF on both monkey esophagus and human salt-split skin were negative. A regimen combining the above-mentioned anti-inflammatory treatment and bath-PUVA phototherapy led to temporary improvement.

Six months later, the patient experienced his first exacerbation (Fig. 1D) with multiple new itchy plaques and erosions on the upper body, which prompted new biopsies. Histopathological examination revealed papillary dermal edema and a superficial inflammatory infiltrate composed predominantly of neutrophils and eosinophils, with focal neutrophilic involvement of both the epithelium and papillary dermis. These findings supported a diagnosis of superficial eosinophil- and neutrophil-rich dermatitis. Considering clinical presentation and marked pruritus, the histopathological features were interpreted as consistent with atypical PP. Nevertheless, dermatitis herpetiformis remained within the differential diagnosis (Fig. 1E–H), despite persistently negative results by both direct and indirect IF analyses.

Treatment with minocycline led to near-complete remission, consistent with its known anti-neutrophilic effect in PP. However, approximately fifteen months after the initial presentation, the patient developed another exacerbation (**Fig. 2**A). For the first time, small tense blisters were observed. Histopathology demonstrated a subepidermal cleft with neutrophil- and eosinophil-rich infiltrates (Fig. 2B–C), while direct IF of a perilesional biopsy and indirect IF continued to show no immune deposits. Based on the clinical course and the previous partial response to minocycline, dapsone therapy was initiated, resulting in marked clinical improvement. Nevertheless, after the patient independently discontinued dapsone, a severe flare occurred, now characterized by widespread tense blisters and erosions, compatible with the classical clinical presentation of LAD (Fig. 2D).

A lesional biopsy now revealed subepidermal blistering with dense neutrophilic infiltration (Fig. 2E). For the first time, performed on 2 perilesional biopsies revealed discrete linear IgA deposition along the dermal–epidermal junction. No clear serrated staining pattern could be identified in either of the 2 biopsies. Indirect IF on salt-split skin showed linear IgA deposition at the roof of the artificial blister (Fig. 2F). Immunoblot analyses using the conditioned medium of cultured human keratinocytes demonstrated IgA reactivity with LAD-1, the 120 kDa soluble ectodomain of BP180 (Fig. 2G), while IgG antibodies were directed against the C-terminal fragment of BP180 (**Table I**). After reintroduction of dapsone, even without additional topical corticosteroids, the skin lesions healed within a few weeks and pruritus ceased. The patient has remained in remission to date.

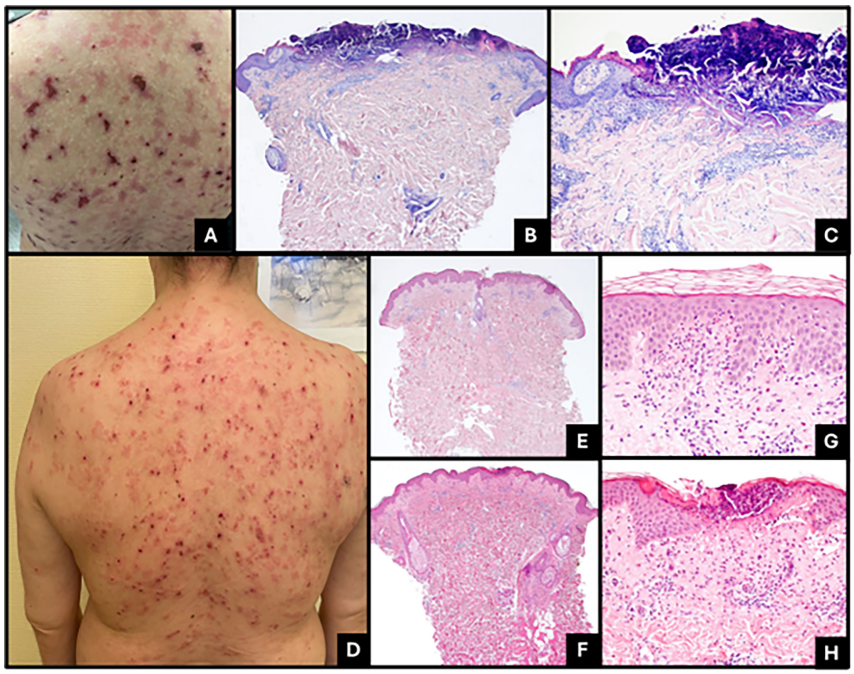

Fig. 1. Clinical and histopathological findings at initial presentation and during the first exacerbation. Multiple erythematous, flat and eroded papules and plaques on the upper body (A). Biopsy specimen showing a superficial ulceration and an inflammatory infiltrate located in the upper dermis (haematoxylin-eosin, 40×) (B). Higher magnification illustrating the shallow ulceration covered by a crust (haematoxylin-eosin, 100×) (C). Numerous erythematous papules, plaques and erosions with crusts on the back (D). Biopsy specimens showing papillary dermal edema and a superficial inflammatory infiltrate in the upper dermis (haematoxylin-eosin, 40×) (E, F). Higher magnification illustrating alignment of neutrophils and eosinophils along the dermoepidermal junction (haematoxylin-eosin, 200×) (G), and focal epidermal erosion with an overlying neutrophilic crust (haematoxylin-eosin, 200×) (H).

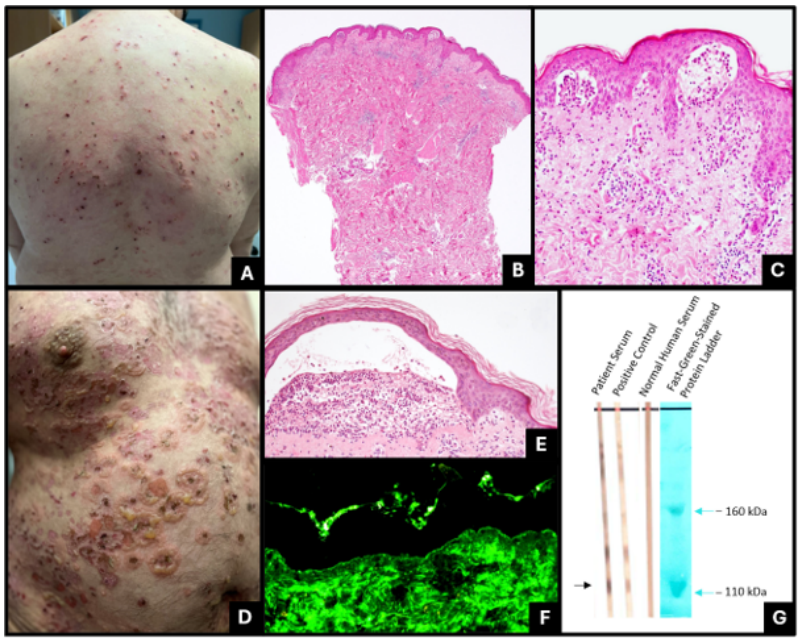

Fig. 2. Clinical and histopathological findings at relapse after loss of response to minocycline and at the relapse after discontinuation of dapsone. Persistent erythematous papules and erosions, with a few newly appearing small tense vesicles on the upper back (A). Biopsy specimen showing papillary dermal edema and a superficial inflammatory infiltrate located in the upper dermis (haematoxylin-eosin, 40×) (B). Higher magnification illustrating focal subepidermal cleft formation and the mixed neutrophilic and eosinophilic composition of the infiltrate along the dermoepidermal junction (haematoxylin-eosin, 300×) (C). Extensive erythematous erosions and vesiculobullous lesions with serous content, partially arranged in a "string-of-pearls" configuration on the abdomen (D). Biopsy specimen illustrating a subepidermal cleft and a dense neutrophilic infiltrate (haematoxylin-eosin, 300×) (E). Indirect immunofluorescence on salt-split skin demonstrating linear IgA deposition at the roof of the artificial blister (F). Immunoblot analysis showing IgA reactivity of the patient's serum with LAD-1, the 120 kDa soluble ectodomain of BP180. From left to right: lane 1 Patient Serum; lane 2 Positive Control (a serum of a patient with linear IgA); lane 3 Normal Human Serum; lane 4 Fast-Green-Stained Protein Ladder with molecular weight marker (G).

Table I. Serological findings during disease course

Date	BP180-NC16A ELISA (IgG)	Anti-gliadin-IgA ELISA	Anti-tissue transglutaminase -IgA ELISA	IF monkey esophagus	IF Salt-split skin	IB LAD-1	IB with recombinant C-terminal fragment of BP180
07/2021	Negative			Negative	Negative		
02/2022		Negative	Negative	Negative	Negative		
12/2022	Negative	Negative	Negative	Negative	Negative		
11/2023	Negative			Negative	Positive	Positive IgA	Positive IgG

BP180-NC16A: Bullous pemphigoid antigen, 180 kDa, non-collagenous domain 16A; ELISA: Enzyme-linked immunosorbent assay; IB: Immunoblot; IF: Indirect immunofluorescence; LAD-1: Linear IgA dermatosis antigen 1.

DISCUSSION

Prodromal AIBD without evident blistering, negative direct IF, and no detectable serum autoantibodies pose a considerable diagnostic challenge. In cases with ambiguous clinical morphology, physicians must remain alert and critically reassess initial but potentially non-contributory immunopathologic results, rather than relying on them uncritically.

In our case, the initial clinicopathologic constellation suggested PP. This working diagnosis appeared even more plausible given the clinical improvement under anti-inflammatory and anti-neutrophilic therapy. However, the subsequent exacerbations and chronic–recurrent course eventually raised suspicion that an AIBD might in fact be present.

The diagnosis of an AIBD was ultimately established only after 5 perilesional biopsies, when direct IF finally revealed linear IgA deposition along the cutaneous basal membrane zone, coinciding with the first clinical appearance of tense blisters. Direct IF is the diagnostic gold standard, and according to the European S2k guideline (4), the diagnosis of LAD requires both compatible clinical features and a positive direct IF result. When direct IF is negative, a repeated biopsy is strongly recommended, as false negatives may occur if samples are taken from central lesional area or if immune deposits have degraded over time. As Shimanovich et al. reported in a large study on mucous membrane pemphigoid, negative direct IF findings do not necessarily exclude AIBD and should prompt further biopsy-based diagnostic evaluation (8). If direct IF remains repeatedly negative or cannot be performed, the diagnosis may still be established by combining clinical features with serology (9). In the present case, the detection of circulating IgA labelling the roof of salt-split skin by indirect IF and IgA reactivity against LAD-1 confirmed the diagnosis of LAD. In addition, concomitant circulating IgG autoantibodies were detected, a phenomenon described in about 20–25% of LAD cases (10).

A comparable situation, in which PP simulated an AIBD, was described by Saito et al., who reported a 24-year-old Japanese woman initially diagnosed with PP before dermatitis herpetiformis was established. Only after repeated biopsies performed during a relapse of lesions following dapsone dose reduction, after an initial therapeutic response, did direct IF reveal IgA deposits confirming dermatitis herpetiformis (6). PP typically manifests in young adults between 20 and 30 years of age, with a marked female predominance (2–3:1). The characteristic lesions consist of intensely pruritic urticarial papules and plaques that resolve with reticulated hyperpigmentation. In early disease stages, PP can histopathologically resemble AIBD; however, direct IF is consistently negative (5).

In summary, this case highlights the broad clinical spectrum of LAD, which may manifest for extended periods without overt blistering, and the diagnostic challenges posed by false-negative immunofluorescence results. Physicians should therefore maintain a high index of clinical suspicion and repeat perilesional IF studies whenever the clinical picture does not fully align with initial findings.

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Data Availability: The data that support the findings of this study are available from the corresponding author upon reasonable request.

The authors have no conflicts of interest to declare.

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