SKIN

BRIEF ARTICLE

Eosinophil-rich Linear IgA Bullous Dermatosis: A Case Report of a Rare Entity

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ABSTRACT

Linear IgA bullous dermatosis (LABD) is an uncommon autoimmune bullous disease characterized by a subepidermal neutrophilic infiltrate. It classically presents as widespread tense vesicles in an annular pattern and is defined by the presence of linear deposition of IgA at the dermal-epidermal junction. Eosinophil-rich LABD has been rarely reported in the literature, including in association with COVID-19 booster vaccination, parvovirus B19 infection, hematologic malignancies, and drug-induced LABD. The varied clinical and histopathologic findings often make it difficult to differentiate LABD from other blistering diseases such as bullous pemphigoid and dermatitis herpetiformis. For this reason, immunofluorescence and serologic testing are critical for identifying the underlying diagnosis.

CASE REPORT

We present a 65-year-old male with a history of end-stage renal disease due to hypertension (on peritoneal dialysis), hyperlipidemia, and a newly discovered bladder mass who presented for a several-month history of a widespread pruritic rash. Physical examination revealed erythematous papules and plaques with hemorrhagic crust scattered on the trunk and arms. No intraoral lesions were observed. Lesional hematoxylin and eosin (H&E)-stained biopsy from the flank revealed a subepidermal blister with eosinophils (**Figure 1A and B**), favoring bullous pemphigoid. However, direct immunofluorescence (DIF) was negative for immunoglobulin (Ig) G, IgM, IgA, C3 and fibrinogen. The patient's skin disease progressed despite empiric treatment including oral doxycycline and topical clobetasol, leading to hospitalization.

Upon re-evaluation, the patient had diffuse erythematous annular and polycyclic plaques with hemorrhagic crust and tense vesicles (**Figure 2**). A repeat biopsy from the patient's left arm again showed a subepidermal blister with numerous eosinophils and some neutrophils. Serologic testing for antibodies against bullous pemphigoid (BP) 180 and 230 antigens was negative. DIF revealed the presence of 1+ linear IgA along the dermo-epidermal junction, leading to a diagnosis of linear IgA bullous dermatosis or LABD (**Figure 1C**). The patient was started on a prednisone

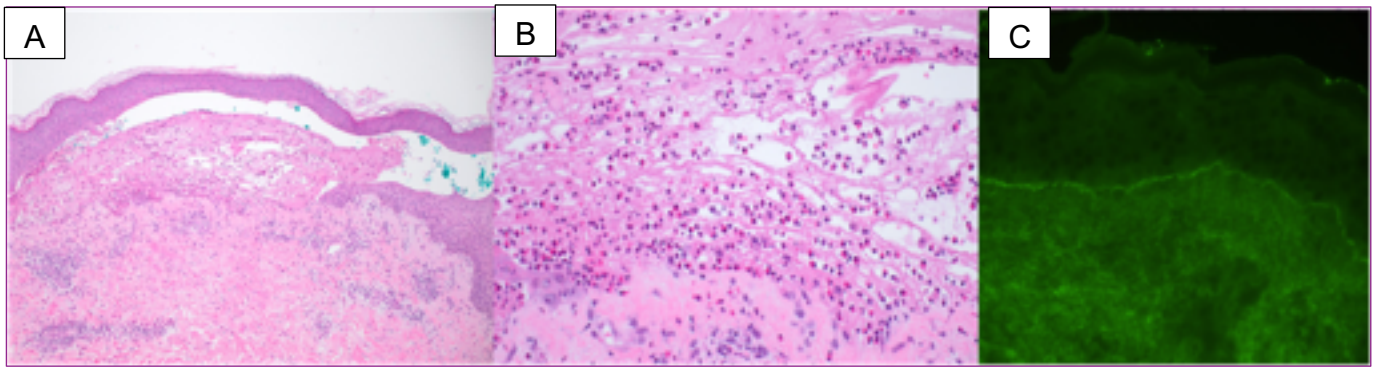

Figure 1. (A) Sections of the punch biopsy showed subepidermal bulla formation with superficial dermal inflammation and inflammatory cells within the blister cavity (H&E, 40x) (B) High-power view demonstrated that many of the inflammatory cells within the blister cavity as well as in the superficial dermis were eosinophils (H&E, 400x). (C) Direct immunofluorescence studies demonstrated 1+ linear IgA deposition at the basement membrane zone (400x). There was no deposition of IgG, IgM, C3, or fibrinogen.

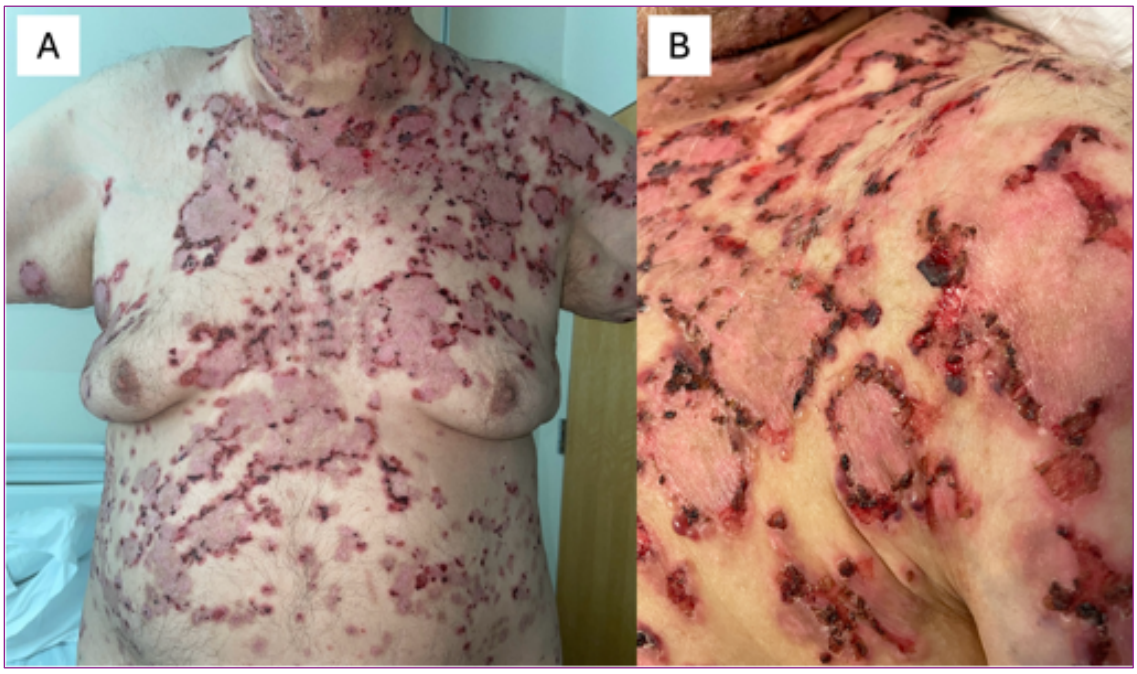

Figure 2. (A, B) Two months after initial presentation showed a progressed extensive eruption of erythematous annular plaques with peripheral vesicles and hemorrhagic crusted erosions.

taper at 1 milligram per kilogram and oral dapsone. Unfortunately, the patient passed away before the scheduled one-month follow-up. The exact cause of death was unknown.

LABD is a rare autoimmune blistering disease typically presenting with widespread tense annular or arciform vesicles, resembling a “string of pearls” or “crown of jewels.” Histologically, LABD is generally characterized by the presence of a subepidermal blister with neutrophils and linear deposition of IgA at the dermo-epidermal junction. The diagnosis of LABD relies on both clinical and histological

assessments. Despite the unusual eosinophilia, the diagnosis was made in this case based on the clinical morphology, subepidermal blister, linear IgA deposition on DIF, and absence of BPAG1 and 2 antibodies.

There are three prior non-drug cases of eosinophil-rich LABD reported in literature. These include associations with a vaccine, an infection, and a malignancy, and possibly up to 21% of LABD from drug-related cases have it.[1-5] Nahm et al. and García-Gi et al. described cases of LABD with an eosinophilic infiltrate occurring in association with the recombinant COVID-19 booster and parvovirus B19, respectively.[1-2] A retrospective study in France found that 21% of drug-induced LABD cases feature eosinophilic infiltrates, with vancomycin as the suspected drug for more than half of their subjects.[3]

A further interesting aspect of this case was the temporal association of the discovery of his bladder mass with the onset of his LABD. Although malignancy-associated LABD has indeed been documented, the primary association is with hematologic malignancies.[4-5] One reported patient with angioimmunoblastic T-cell lymphoma developed LABD that involved an eosinophilic infiltrate, along with neutrophils, within blister cavities.[4] No cases of LABD associated with bladder malignancy have been reported in the literature.

Blistering diseases, in general, can pose a challenge to providers due to similar clinical morphology and overlapping histologic features. This can be especially difficult when the pathology deviates from classical histologic features, as observed in our patient. Because numerous factors can trigger LABD, conducting a thorough patient history is crucial for identifying the etiology of this disease. Non-diagnostic biopsies in patients with suspected blistering diseases may require a repeat biopsy. Direct immunofluorescence and serologic testing for known antibodies or for indirect immunofluorescence are also valuable tools in distinguishing various blistering diseases and should be incorporated in the diagnostic workup.

Conflict of Interest Disclosures: None

Funding: None

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