CASE REPORT

Dermatitis herpetiformis-like Linear IgA bullous disease in a Filipino

Lauren Camille B. Livelo, MD[1], Elisabeth Ryan, MD[a], Johannes F. Dayrit, MD, FPDS, FDSP[b], Ricky H. Hipolito, MD, FPDS[b]

ABSTRACT

Introduction: Linear IgA bullous disease (LABD) is a rare autoimmune blistering disease characterized by subepithelial bullae and linear IgA deposition along the basement membrane zone of the epidermis. Lesions present as pruritic papulovesicles and tense bullae which may coalesce forming annular or polycyclic urticarial plaques with blistering on the edge of the lesions forming the classic "string of pearls" sign. Lesions may affect the face, trunk, and extensor extremities. Incidence rates range from 0.5 to 2.3 cases per million individuals per year. Due to its rare occurrence, there are only a few documented reports on cases of LABD, particularly in the Filipino population.

Case Report: A 33 year-old Filipino female consulted because of a 3-week history of severely pruritic vesicles and crusts on the face, trunk, and arms. Patient noted no gastrointestinal symptoms on consultation. Skin punch biopsy revealed subepidermal blisters with collection of neutrophils at the dermal papillae. Direct immunofluorescence showed strong (+2) deposits of linear IgA at the dermo-epidermal junction in perilesional skin thus validating the diagnosis. The patient's serum was negative for IgA anti-tissue transglutaminase and IgA antiendomysial antibodies. Patient was treated with topical corticosteroids and Dapsone 50 mgs OD with remarkable improvement.

Conclusion: Linear IgA bullous disease has very few reported cases especially in the Philippine setting. Dapsone is considered the first-line treatment. The disease may persist for a decade or longer, and relapses may occur. Careful history-taking accompanied by the histological, immunofluorescence, and serological findings must be done to ensure proper treatment and good prognosis.

Keywords: Linear IgA bullous disease, LABD, autoimmune, dermatitis herpetiformis

INTRODUCTION

Linear IgA bullous dermatosis (LABD), also known as linear IgA disease (LAD), is an idiopathic or drug-induced autoimmune subepidermal blistering disease usually presenting with clear or hemorrhagic, tense, pruritic vesicles or bullae, sometimes with an erythematous or urticarial base. Some lesions may coalesce to form annular or polycyclic plaques with a characteristic blistering along the edge of the lesion, the supposed "string-of-pearls" sign. [1, 2, 3] Mucosal membranes may also be affected, with lesions more commonly found on the oral cavity and eyes in 70% of patients. [1, 2] It is a rare disease with an incidence rate ranging from less than 0.5 to 2.3 cases per million individuals per year, with slight female predominance. [1, 2] It affects people of all races and ages but often occurs at two peaks: 1. childhood-onset, or often termed chronic bullous disease of childhood (CBDC) which appears up until 5 or 6 years of age; and 2. adult-onset, which appears after puberty until later in life. [1, 2] In adults, the affected areas may include the scalp, face, trunk, hands, and feet. [4] The diagnosis of LABD relies

[a] Resident, [b] Consultant Research Institute for Tropical Medicine, Department of Dermatology

Corresponding Author: Lauren Camille B. Livelo, MD
lauren.livelo@gmail.com

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on the clinical presentation, histological picture, direct immunofluorescence, and serological findings. Given that LABD is a rare disease, there are very few studies and case reports available particularly in the Filipino population.

CASE REPORT

We present a case of a 33-year old Filipino female who consulted at our clinic due to a 3-week history of severely pruritic vesicles on erythematous base which coalesced to form crusted erosions with excoriations on the face, trunk, and arms (Fig. 1). There were no mucosal lesions. Her medical history did not include any comorbidities nor any previous infection. She denied any known skin conditions, possible allergens, or intake of any new medications. There were no noted gastrointestinal symptoms on consultation. The patient is a housewife. No similar lesions were found in any family member.

Skin punch biopsy was done, revealing a basket-woven stratum corneum and mild acanthosis of the epidermis and subepidermal blisters with collection of neutrophils at the dermal papillae (Fig. 2). The initial impression was dermatitis herpetiformis (DH), however, direct immunofluorescence showed strong (+2) deposits of linear IgA at the basement membrane zone of the epidermis in perilesional skin (Fig. 3), leading to the diagnosis of Linear IgA bullous disease. The patient's serum was negative for IgA anti-tissue transglutaminase and IgA antiendomysial antibodies. Patient was treated with topical corticosteroids and Dapsone 50 mgs OD with notable improvement of lesions.

DISCUSSION

Linear IgA bullous disease is a rare autoimmune subepidermal blistering skin disorder associated with linearly distributed IgA deposits along the basement membrane zone of the epidermis. [5, 6] LABD may be idiopathic or drug-induced in origin. The most commonly identified drug is vancomycin, but there are also reports of other drugs such as amiodarone, diclofenac, captopril, ceftriaxone, metronidazole, naproxen, peroxicam, and phenytoin. [2, 6, 7] Lesions in drug-induced LABD are more widespread and may mimic lesions of erythema multiforme, morbilliform drug eruptions, or toxic epidermal necrolysis.[8] A positive Nikolsky sign as well as larger erosions are seen in these cases.8 Histologic findings of LABD shows a subepithelial blister with neutrophilic-predominant infiltrates in the upper epidermis. [1, 2, 4] This closely resembles histological findings of dermatitis herpetiformis;[2]

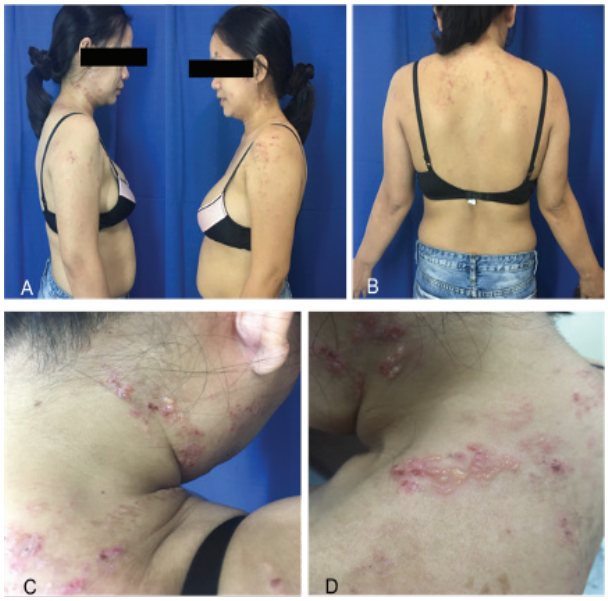

Figure 1. A) Physical examination of the lesions shows symmetrical appearance of vesicles on erythematous base and crusted erosions with excoriations on the face, arms, and (B) trunk. (C) and (D) shows close-up of the vesicles and crusted erosions.

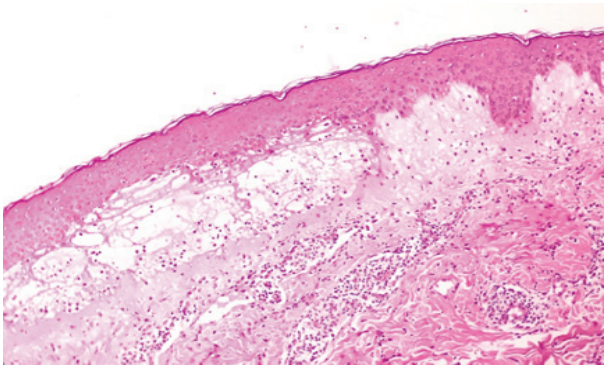

Figure 2. Biopsy of the lesions on the upper back. Epidermis shows basket-woven stratum corneum and mild acanthosis with subepidermal blisters with collection of neutrophils at the dermal papillae. (H&E stain, 100x)

therefore, it is recommended to perform direct immunofluorescence (DIF) on perilesional skin and/or indirect immunofluorescence (IIF) of the patient's serum to validate the presence of IgA autoantibodies. DIF results would show linear IgA deposition at the basement membrane zone. [1,2,4] Other immunoreactants such as IgG, IgM, and C3 may also appear on DIF. [2, 4] IIF shows typically low circulating antibody titers (1:2 to 1:64) and is positive in less than 50%. [2]

Most cases of LABD were controlled with dapsone and topical corticosteroids. [1,2] Dapsone is considered the first-line therapy for LABD.[4] Before starting dapsone, complete blood count, liver and renal function tests, G6PD deficiency, and urinalysis should be tested. Dapsone should be started at a dose of 25-150mg/day.[2] The duration of treatment for LABD is variable. LABD typically persists for months to several years prior to spontaneous resolution. However, the disease may persist for a decade or longer, and relapses may occur. [9]

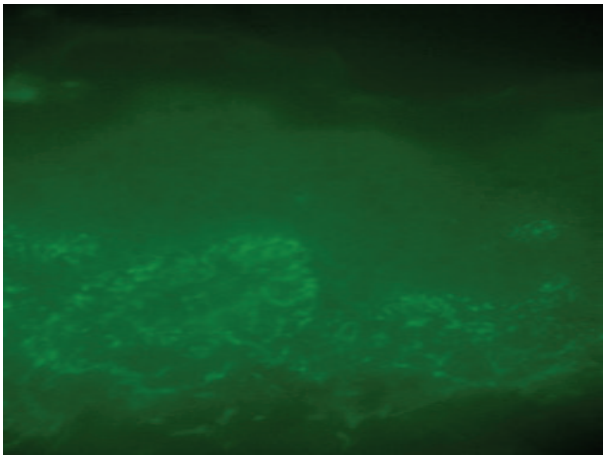

Figure 3. Direct Immunofluorescence showed strong (+2) linear deposits of IgA at the basement membrane zone of the epidermis in perilesional skin.

Table 1. Comparison between features of LABD and DH.

	Linear IgA Bullous Disease	Dermatitis Herpetiformis
Clinical features	Annular or grouped papules, vesicles, and bullae on extensors, including elbows, knees, and buttocks. Pruritus is less severe than in DH	Erythematous papules, urticarial-like plaques, or vesicles on elbows, knees, buttocks, shoulders, sacral area Associated with gluten-sensitive enteropathy
Histopathology	Sub-epidermal bulla with collections of neutrophils along the basement membrane, often accumulating at the papillary tips	Sub-epidermal cavity with neutrophils in the dermal papillae, edema of the papillary dermis, eosinophils may be present
Direct Immunofluorescence	Linear deposition of IgA at the dermal-epidermal basement membrane	Granular deposition of IgA in dermal papillae

CONCLUSION

Linear IgA bullous disease is a rare blistering skin disorder with very few reported cases especially in the Philippine setting. Diagnosis is achieved through meticulous history-taking in conjunction with clinical, histological, immunofluorescence, and serological findings to ensure proper treatment and good prognosis.

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