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Rituximab for Refractory Adult Linear Iga Bullous Dermatosis: A Case Report

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Abstract | **Case Report**

Linear IgA bullous dermatosis (LABD) is a rare autoimmune bullous disease characterized by linear IgA deposition along the basement membrane zone. While dapsone remains the cornerstone of treatment, a subset of adult patients develops severe, treatment-refractory disease requiring prolonged systemic corticosteroid therapy, and the role of rituximab in this setting remains poorly documented. We report the case of a 34-year-old woman with severe mucocutaneous LABD who failed to achieve disease control despite 18 months of treatment with dapsone, high-dose systemic corticosteroids, and doxycycline, resulting in persistent corticosteroid dependence and significant impairment in quality of life. Following confirmation of the diagnosis by repeat histopathology and direct immunofluorescence, rituximab was initiated using the pemphigus protocol (two 1-g infusions administered two weeks apart), followed by maintenance infusions every six months. A marked and sustained clinical improvement became evident from the second treatment cycle, allowing complete withdrawal of systemic corticosteroids and doxycycline, and at the latest follow-up the patient remained clinically stable on dapsone monotherapy, with only minimal residual cutaneous activity. This case highlights rituximab as a promising steroid-sparing rescue therapy for severe refractory adult LABD and contributes to the limited evidence supporting its use in this uncommon autoimmune bullous disease; further studies are needed to establish its optimal treatment regimen and long-term efficacy.

Keywords: Autoimmune bullous disease, corticosteroid dependence, dapsone, linear IgA bullous dermatosis, rituximab.

INTRODUCTION

Linear IgA bullous dermatosis (LABD) is a rare autoimmune bullous disease characterized by linear deposition of IgA autoantibodies along the basement membrane zone on direct immunofluorescence, the diagnostic hallmark of the disease (Fortuna & Marinkovich, 2012). LABD affects both children and adults, with adult-onset cases being predominantly idiopathic, although drug-induced forms, particularly those associated with vancomycin, have also been reported (Fortuna & Marinkovich, 2012; Paul *et al*., 1997). The principal target antigens are the 97-kDa (LAD-1) and 120-kDa (LABD97) cleavage fragments of BP180, although immunological heterogeneity has been increasingly recognized (Paul *et al*., 1997). The reported annual incidence ranges from 0.5 to 2.3 cases per million individuals (Bernard *et al*., 1995).

Clinically, LABD is characterized by tense vesicles and bullae arranged in annular or polycyclic configurations, classically described as the "crown of jewels" sign, and may involve both the skin and mucous membranes (Fortuna & Marinkovich, 2012; Genovese *et al*., 2019). Dapsone remains the first-line treatment and is frequently combined with topical or systemic corticosteroids in patients with extensive disease. Nevertheless, a small subset of adult patients develops persistent, corticosteroid-dependent disease despite conventional therapy, representing a major therapeutic challenge (Genovese *et al*., 2019; Khan *et al*., 2023).

Rituximab, an anti-CD20 monoclonal antibody widely used in pemphigus and other autoimmune bullous diseases, has recently emerged as a potential therapeutic option for severe refractory LABD, although evidence remains limited to isolated reports (Werth *et al*., 2021). We report the case of a 34-year-old woman with severe corticosteroid-dependent LABD who achieved sustained clinical improvement following rituximab therapy after failure of multiple conventional treatments.

CASE PRESENTATION

A 34-year-old woman with no significant past medical history presented in July 2022 with a generalized pruritic bullous eruption involving both the skin and mucous membranes. Physical examination revealed multiple tense vesicles and bullae arising on an erythematous background, associated with erosions and hemorrhagic crusts. The lesions predominantly involved the trunk, the proximal portions of the upper and lower extremities, the axillary and inguinal folds, as well as the oral and genital mucosae (Figure 1).

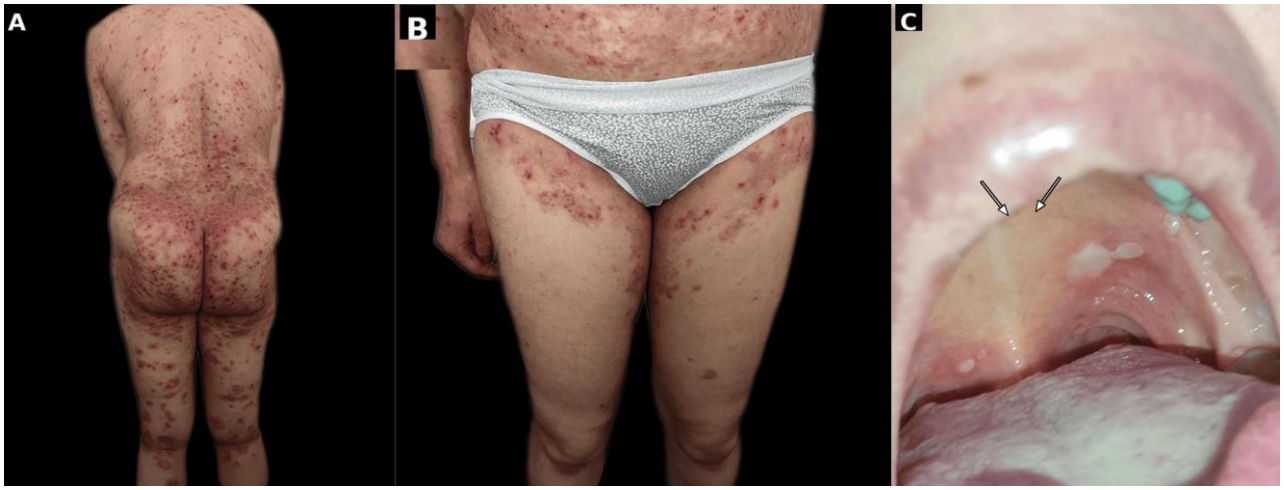

Figure 1: (A, B) Numerous excoriated, crusted papules and erosions with post-inflammatory hyperpigmentation involving the trunk, buttocks, groin, and thighs, seen from behind (A) and from the front (B). (C) Small vesicles and erosions on the palatal mucosa, reflecting mucosal involvement

A skin biopsy demonstrated a subepidermal blister with an intact roof, without acantholysis or keratinocyte necrosis, associated with a mixed inflammatory infiltrate containing scattered eosinophils. Direct immunofluorescence performed on salt-split skin revealed fine linear deposits of IgA, C3, and IgG along the basement membrane zone, with staining restricted to the epidermal side of the split, confirming the diagnosis of linear IgA bullous dermatosis (LABD). Indirect immunofluorescence detected circulating anti-basement membrane zone antibodies (titer: 1:40), whereas anti-intercellular substance antibodies were negative. An extended autoantibody panel, including antibodies against BP180, BP230, desmoglein 1, desmoglein 3, envoplakin, and type VII collagen, was negative, as were anti-endomysial and anti-tissue transglutaminase antibodies.

Treatment was initiated with dapsone at 2 mg/kg/day (125 mg/day). Because of persistent disease activity, systemic corticosteroids were introduced at 0.5 mg/kg/day and gradually increased to 1 mg/kg/day (70 mg/day), without achieving satisfactory disease control. Approximately one year later, doxycycline (200 mg/day) was added because of ongoing lesion formation but was subsequently discontinued owing to persistent epigastric pain despite concomitant proton pump inhibitor therapy. Despite 18 months of combination therapy with dapsone, high-dose corticosteroids, and doxycycline, the patient remained corticosteroid-dependent, with continuous development of new lesions and marked impairment of her quality of life.

Given the refractory course of the disease, rituximab was initiated according to the pemphigus protocol, consisting of two intravenous infusions of 1 g administered two weeks apart. At the second infusion, clinical examination revealed multiple small erosions covered by hemorrhagic crusts involving the axillae, inframammary folds, back, buttocks, pubic region, and inner thighs, together with diffuse post-inflammatory hyperpigmentation. No active vesicles or bullae were observed, the Nikolsky sign was negative, and the estimated body surface area involved was approximately 5% (Figure 2A–C). The patient reported noticeable clinical improvement after the first rituximab infusion, with approximately one new lesion per day compared with substantially greater disease activity before treatment initiation. Vaginal mucosal involvement persisted as dryness associated with dyspareunia. No infusion-related adverse events or infectious complications were observed during treatment.

Three additional maintenance infusions of rituximab (1 g each) were administered at 6, 12, and 18 months, for a total of five infusions. A marked clinical response became evident after the second treatment cycle, allowing gradual tapering and complete discontinuation of systemic corticosteroids, followed by doxycycline. At the latest follow-up, the patient remained clinically stable on dapsone monotherapy, with only minimal residual cutaneous activity and no further requirement for systemic corticosteroid therapy (Table 1).

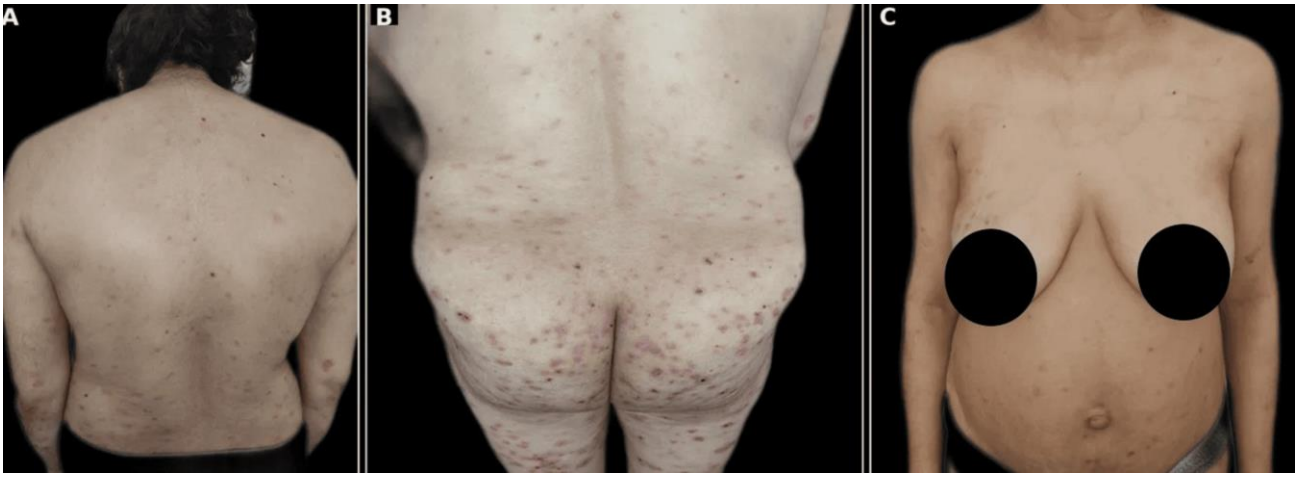

Figure 2: Cutaneous improvement after the first cycle of rituximab, with markedly fewer and less inflammatory lesions on the trunk (A, B) and back (C) compared with baseline

Table 1: Timeline of diagnostic work-up, treatment, and clinical course

Time point	Treatment / Investigation	Clinical course
July 2022	Initial skin biopsy and direct immunofluorescence; dapsone initiated (2 mg/kg/day, 125 mg/day)	Persistent disease activity
6 months after	Repeat skin biopsies and direct immunofluorescence confirmed LABD	Diagnosis established
Following weeks	Systemic corticosteroids initiated at 0.5 mg/kg/day	Persistent lesion formation
Approximately 7 months later	Corticosteroid dose increased to 1 mg/kg/day (70 mg/day)	No satisfactory clinical response
Approximately 1 year after diagnosis	Doxycycline (200 mg/day) added	Persistent disease activity; discontinued because of epigastric pain despite proton pump inhibitor therapy
After 18 months of combination therapy	Dapsone + high-dose corticosteroids + doxycycline	No remission; corticosteroid-dependent disease with continuous development of new lesions
Rituximab induction (Day 1 and Day 15)	Two intravenous infusions of rituximab (1 g each) according to the pemphigus protocol	Early clinical improvement with marked reduction in new lesion formation
Months 6, 12, and 18	Three maintenance rituximab infusions (1 g each)	Progressive clinical improvement; corticosteroids gradually tapered
Latest follow-up	Dapsone monotherapy	Sustained disease control with minimal residual cutaneous activity; systemic corticosteroids and doxycycline successfully discontinued

DISCUSSION

Linear IgA bullous dermatosis (LABD) is a rare autoimmune bullous disease that predominantly affects children but may also occur in adults, in whom it is generally idiopathic and often follows a more persistent clinical course. Although dapsone remains the mainstay of treatment, either alone or in combination with systemic corticosteroids, a subset of patients develops refractory disease characterized by prolonged corticosteroid dependence and recurrent relapses despite multiple therapeutic interventions (Genovese *et al*., 2019; Khan *et al*., 2023).

Our case illustrates two major challenges in the management of adult LABD: diagnostic uncertainty and therapeutic refractoriness. Despite an initial clinicopathological evaluation, repeated histopathological and immunopathological examinations were required before establishing the diagnosis. This emphasizes the importance of careful clinicopathological correlation and repeat direct immunofluorescence when the initial findings are not fully consistent with the clinical presentation. The coexistence of IgA, IgG, and C3 deposits along the basement membrane zone further highlights the immunological heterogeneity of LABD and the potential overlap with other autoimmune subepidermal bullous diseases.

Management of refractory LABD remains particularly challenging because current evidence is largely limited to case reports and small case series. Besides dapsone and systemic corticosteroids, several

therapeutic options, including doxycycline, colchicine, sulfapyridine, mycophenolate mofetil, azathioprine, and other immunosuppressive agents, have been used with variable success in refractory disease (Genovese *et al*., 2019; Khan *et al*., 2023). Nevertheless, our patient remained corticosteroid-dependent after 18 months of combination therapy with dapsone, systemic corticosteroids, and doxycycline, resulting in persistent disease activity and a significant impairment in quality of life.

Rituximab, a chimeric anti-CD20 monoclonal antibody, has revolutionized the treatment of pemphigus and has increasingly been used off-label in other autoimmune bullous diseases because of its corticosteroid-sparing effect and durable efficacy (Werth *et al*., 2021). However, experience with rituximab in LABD remains extremely limited. Pinard *et al*. (2019) first reported two patients with severe refractory LABD who achieved durable remission only after repeated rituximab cycles administered over 14 and 20 months, respectively. Subsequently, Kaya İslamoğlu and Akyürek (2019) described successful treatment of recalcitrant LABD with rituximab after failure of conventional therapy. Nedosekin *et al*. (2021) reported a patient with overlapping IgG/IgA-mediated bullous dermatosis who also responded favorably to rituximab, further illustrating the immunological overlap that may exist among autoimmune subepidermal blistering diseases. More recently, Dhillon *et al*. (2023) described another successful use of rituximab in a patient with refractory LABD who was unable to tolerate first-line therapies. These observations, together with our case, consistently support the efficacy of rituximab in carefully selected patients with severe refractory disease.

The mechanism underlying rituximab efficacy in LABD remains incompletely understood. By depleting CD20-positive B lymphocytes, rituximab reduces the generation of autoreactive B-cell clones and consequently decreases autoantibody production. However, because long-lived plasma cells do not express CD20, IgA-producing plasma cells are not directly targeted, which may explain the heterogeneous therapeutic responses reported among IgA-mediated autoimmune diseases (Lamberts *et al*., 2018). Individual variations in immune pathways and autoantibody profiles probably further contribute to this variability.

Compared with previously published reports, our patient experienced relatively early clinical improvement after the induction regimen, while scheduled maintenance infusions every six months allowed complete withdrawal of systemic corticosteroids and sustained disease control with dapsone monotherapy. Although no standardized rituximab protocol currently exists for LABD, our observation suggests that maintenance therapy may contribute to long-term disease control in selected patients.

Our case further expands the limited but growing body of evidence supporting rituximab as an effective rescue therapy for severe corticosteroid-dependent LABD refractory to conventional treatment. In addition, it highlights the importance of repeated clinicopathological evaluation in diagnostically challenging cases, as immunopathological findings may evolve over time. Larger multicenter studies are warranted to establish standardized treatment protocols, identify predictors of therapeutic response, and better define the long-term efficacy and safety of rituximab in this rare autoimmune bullous disease (Table 2).

Table 2: Published cases of refractory linear IgA bullous dermatosis successfully treated with rituximab

Author (Year)	Age/Sex	Previous therapies	Rituximab regimen	Clinical outcome
Pinard *et al*. (2019)	Adult	Dapsone, corticosteroids, immunosuppressive agents	Repeated rituximab cycles	Durable remission after 14–20 months
Kaya İslamoğlu & Akyürek (2019)	Adult	Conventional therapies	Rituximab	Marked clinical improvement
Nedosekin *et al*. (2021)	Adult	Multiple previous therapies	Rituximab	Sustained remission
Dhillon *et al*. (2023)	Adult	Failure/intolerance of first-line therapies	Rituximab	Favorable clinical response
Present case	34-year-old woman	Dapsone, systemic corticosteroids, doxycycline	1 g × 2 (2 weeks apart), followed by maintenance infusions at months 6, 12, and 18	Marked clinical improvement, complete corticosteroid withdrawal, maintained on dapsone monotherapy

CONCLUSIONS

This case highlights rituximab as a promising rescue therapy for severe corticosteroid-dependent linear IgA bullous dermatosis refractory to conventional treatment. It also underscores the importance of repeated clinicopathological evaluation in diagnostically challenging cases, as immunopathological findings may evolve over time and delay definitive diagnosis. Although current evidence remains limited to isolated case reports, our observation adds to the limited but growing body of evidence supporting the use of rituximab in carefully selected patients with refractory LABD. Larger multicenter studies are warranted to establish standardized treatment protocols, identify predictors of therapeutic response, and better define the

long-term efficacy and safety of rituximab in this rare autoimmune bullous disease.

Additional Information

Author Contributions: All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

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Disclosures

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