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Case Report

A Severe Case of Pediatric Linear IgA Bullous Dermatosis

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KEYWORDS

Linear IgA bullous dermatosis, chronic bullous disease of childhood, pediatric dermatology

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ABSTRACT

Linear IgA bullous dermatosis (LABD) is a rare immune-mediated subepidermal blistering disease, most commonly affecting children between 4 and 5 years of age. Although generally self-limiting, some cases present diagnostic and therapeutic challenges.

We report an 8-year-old boy who presented at age 5 with disseminated pruritic bullous lesions. Initial direct immunofluorescence was inconclusive, and diagnosis was confirmed only three years later upon repeat biopsy demonstrating linear IgA deposition at the basement membrane zone. The clinical course was complicated by recurrent secondary infections, drug-induced hepatitis from dapsone overdose, cyclosporine-induced disease aggravation, and treatment non-adherence. Disease control was ultimately achieved with dapsone and corticosteroids.

This case highlights the importance of repeat biopsy when clinical suspicion persists despite inconclusive findings, careful drug monitoring, and comprehensive caregiver education. A multidisciplinary approach is essential for managing severe pediatric LABD.

1. Intoduction

Linear IgA bullous dermatosis (LABD), classically referred to as chronic bullous disease of childhood (1–3), is an immune-mediated vesicobullous disease that may affect both children and adults. Pediatric prevalence is unknown; however, it is considered a rare disease with a peak occurrence between 4 and 5 years of age and an estimated global incidence of 0.2 to 2.3 cases per million population annually (3–5). LABD is a subepidermal blistering disease characterized by a hallmark immunohistochemical finding: linear deposition of IgA autoantibodies along the basement membrane zone (BMZ) of the dermoepidermal junction, where IgG and C3 may also be detected, in conjunction with a neutrophilic dermal infiltrate (3, 6–8).

In children, LABD is mostly benign; however, in rare cases, it can cause severe mucosal involvement and be associated with a worse prognosis. The disease is considered idiopathic and multifactorial and has been associated with environmental triggers, such as drug exposure, as well as autoimmune diseases (e.g., rheumatoid arthritis and psoriasis). Antibiotics—especially vancomycin—are the most commonly implicated agents in drug-induced forms; however, many other drugs, including anti-inflammatory/analgesic agents (e.g., ketoprofen, diclofenac, and piroxicam) and antihypertensive medications (e.g., captopril and other ACE inhibitors), can also trigger the disease (5, 9–11). More recently, associations with specific human leukocyte antigen (HLA) haplotypes have suggested a genetic predisposition (12).

The diagnosis of LABD can be challenging, as there are no rapid tests or expedited diagnostic pathways to reliably distinguish it from bullous pemphigoid (BP) or dermatitis herpetiformis (DH). Bullous pemphigoid typically presents with tense bullae on erythematous or urticarial bases, whereas dermatitis herpetiformis is characterized by grouped vesicles and papules accompanied by intense pruritus. In contrast, LABD classically exhibits annular or polycyclic lesions forming the characteristic “cluster of jewels” configuration. Nevertheless, significant clinical overlap frequently delays diagnostic confirmation. Moreover, drug-induced LABD can manifest from 24 hours to 4 weeks after exposure to the causal agent, warranting consideration of LABD in the differential diagnosis when evaluating patients with bullous disease.

Herein, we report a challenging case of pediatric LABD complicated by recurrent secondary infections and treatment non-adherence, highlighting the diagnostic and therapeutic complexities of this condition.

2. Clinical Case

An eight-year-old boy first presented to the Pediatric Dermatology Division of Hospital Jesus in 2020, at the age of five, with disseminated papules. Over a 10-month period, these lesions progressed to severe pruritic bullous lesions with annular configurations, predominantly affecting the back, arms, and periaxillary regions (Figure 1A and 1B).

Initial laboratory evaluations, including complete blood count, bilirubin levels, and renal and liver function tests, were within normal ranges. Based on the clinical history and lesion characteristics, LABD was suspected. A skin biopsy with direct immunofluorescence (DIF) was performed; however, the results were inconclusive, supposedly due to intense secondary infection and/or technical difficulties. The first treatment regimen consisted of dapsone, the first-line therapy for pediatric LABD, combined with prednisolone for additional disease control, chlorpheniramine to alleviate pruritus, and vitamin D supplementation for documented deficiency. This regimen was administered for four months.

In February 2022, the patient was hospitalized for 14 days due to secondary skin infections and drug-induced hepatitis caused by maternal doubling of the prescribed dapsone dosage. During hospitalization, the dapsone dose was adjusted, oxacillin was initiated, and prednisolone was continued. Although temporary improvement was observed, blistering lesions persisted. One month after discharge, another secondary infection required treatment with sulfamethoxazole for 14 days, in combination with prednisolone and dapsone.

Attempts to taper corticosteroids in May 2022 resulted in exacerbation of the lesions. Subsequent maternal nonadherence further exacerbated disease activity, by August 2022, with the emergence of scalp lesions. Cyclosporine was initiated but led to significant disease aggravation. Therapy was reverted to dapsone and prednisolone, with terbinafine added to treat concurrent tinea capitis.

By May 2023, further efforts to taper corticosteroids led to recurrence of bullous lesions, prompting initiation

of mycophenolate mofetil. Later that year, the patient developed febrile lymphadenopathy, abdominal pain, and infected skin lesions, leading to another hospitalization. Management included intravenous immunoglobulin and broad-spectrum antibiotics (clindamycin, vancomycin, and fluconazole) for the treatment of sepsis.

By November 2023, at eight years of age, the patient demonstrated stable clinical improvement on a regimen of dapsone, corticosteroids, sulfamethoxazole, and omeprazole. Bullous lesions were limited to the perioral and genital regions and were effectively managed with topical corticosteroids. A new skin biopsy with DIF exhibited linear IgA deposition along the basement membrane zone (BMZ), confirming LABD diagnosis (Figure 2). Long-term dapsone therapy proved successful (Figure 1 C and D).

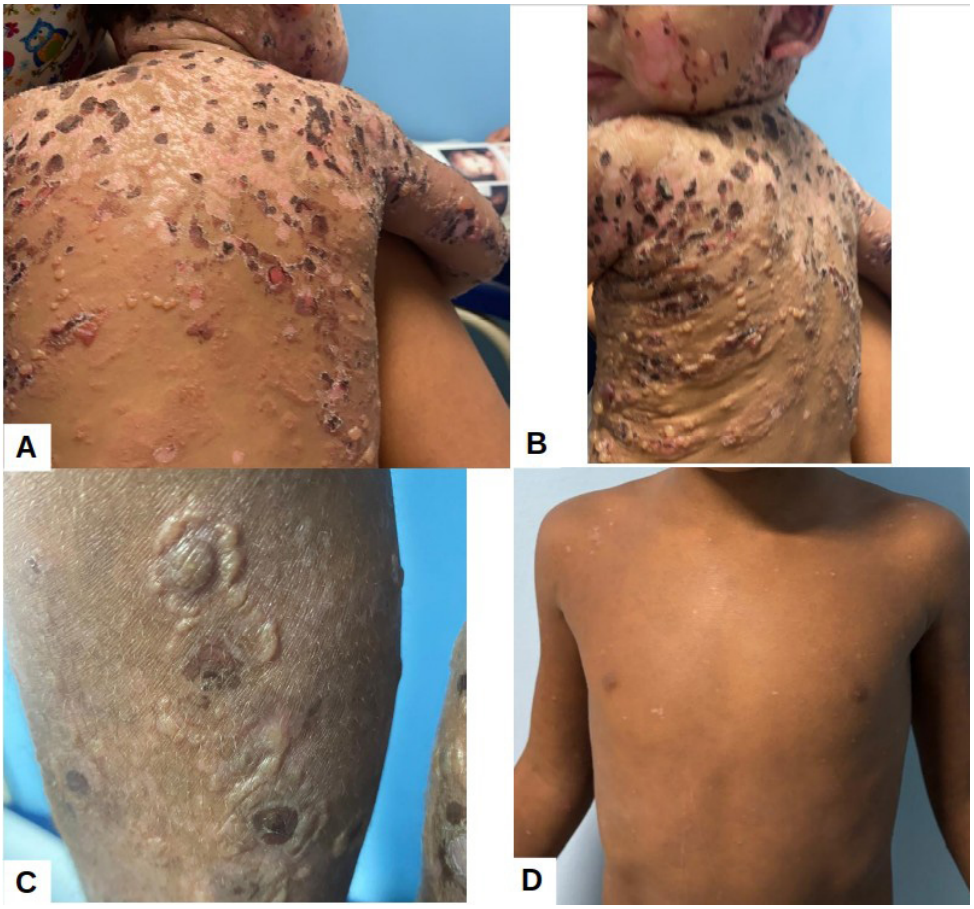

Figure 1. A, B: *Disseminated pruritic bullous lesions with annular configuration involving the back, arm, and periaxillary regions at initial presentation, showing the characteristic "cluster of jewels" pattern of pediatric linear IgA bullous dermatosis.* **C, D:** *Marked clinical improvement after long-term treatment with dapsone and corticosteroids, with resolution of widespread bullous lesions and residual limited involvement.*

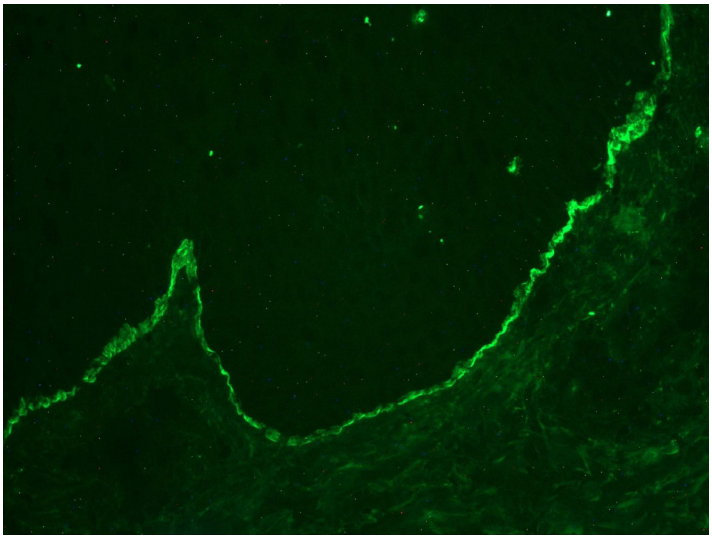

Figure 2. *Direct immunofluorescence: finely linear fluorescence for IgA at the basement membrane.*

3. Discussion

LABD clinical and histopathological features overlap with dermatitis herpetiformis, bullous lupus erythematosus, drug eruption, and rare forms of bullous pemphigoid, and the precise diagnosis requires a skin biopsy with DIF, which demonstrates linear IgA deposition at the BMZ. Occasionally, IgG and C3 deposits may also be observed. In cases where initial DIF findings are inconclusive despite strong clinical suspicion, repeat biopsy is warranted, as demonstrated in our patient, whose diagnosis was only confirmed upon a second DIF performed three years after initial presentation.

Prompt recognition of LABD is critical for ensuring accurate diagnosis and timely treatment. Pruritic bullous lesions with annular configurations and the characteristic "cluster of jewels" pattern should alert pediatricians to consider LABD, particularly when drug-induced causes are suspected or exacerbations occur following medication use.

Dapsone remains the first-line treatment for pediatric LABD, with favorable outcomes at doses of 50–100 mg/day or 0.5 to 3mg/kg/day (12, 13). However, regular monitoring of hepatic function is essential, as illustrated by our case, in which maternal overdose of dapsone resulted in drug-induced hepatitis requiring hospitalization.

Cyclosporine has been proposed for refractory cases; however, it has also been reported to induce LABD (6, 12, 14). In our patient, cyclosporine initiation led to significant disease aggravation, prompting its discontinuation. The precise mechanisms underlying this paradoxical response remain unclear but are likely related to complex interactions between the host immune or genetic background and drug-induced immune modulation, underscores the importance of close clinical monitoring when introducing immunosuppressive agents in LABD.

Although pediatric LABD is generally considered a self-limiting condition, with spontaneous resolution typically occurring within four years (4), our case demonstrates that a subset of patients may experience a protracted and complicated course. Factors contributing to disease severity in this case included recurrent secondary infections, treatment non-adherence, and challenges in corticosteroid tapering.

This case illustrates many of these diagnostic and therapeutic challenges, including inconclusive early biopsy, overlapping clinical features, and refractory disease course, emphasizing the need for vigilance in prolonged or refractory bullous eruptions.

4. Conclusions

This case underscores the complexity of managing severe LABD, particularly in pediatric patients. Key challenges include recurrent secondary infections, treatment non-adherence, and the delicate balance between tapering corticosteroids and minimizing adverse effects while maintaining adequate disease control.

Effective management requires a multidisciplinary approach, strict adherence to prescribed therapy, and comprehensive caregiver education to reduce complications and optimize clinical outcomes. This case also reinforces the importance of timely biopsy, careful drug monitoring, and long-term follow-up to ensure sustained remission.

Authorship information

All authors contributed to all stages of the manuscript and had effective participation in the research guidance, contributed to the intellectual participation in the propaedeutic and/or therapeutic conduct of the studied cases, to the critical review of the literature and approved the final version of the manuscript.

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