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Unmasking The Blisters: A Case Of Pediatric Linear IGA Bullous Dermatosis Mimicking Bullous Impetigo

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ABSTRACT: Introduction – Linear IgA bullous dermatosis (LABD) is one of many diseases that fall under the umbrella of bullous diseases. It is a rare and chronic autoimmune condition affecting both adult and pediatric populations. In children, it is also known as chronic bullous disease of childhood and presents with a typical cluster of jewel-like appearance.

Case presentation — We report a case of a 6-year-old boy who presented with vesicles and bullae over a course of 10 days. The patient was initially diagnosed with bullous impetigo and started on treatment with topical mupirocin and oral amoxicillin-clavulanate. Upon seeing no improvement, the patient returned and was diagnosed with linear IgA bullous dermatosis based on direct immunofluorescence testing (DIF). He was initiated on dapsone and topical corticosteroids, which resulted in gradual remission of the disease.

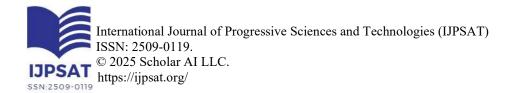
Conclusion – Given that LABD is a rare disease, it is often misdiagnosed. Its clinical features, although typical, are not specific to the disease. Diagnostic investigations include histopathological studies and direct immunofluorescence testing. Although the diagnosis of LABD is challenging, its treatment is relatively simple with dapsone.

Keywords: Linear IgA Bullous Dermatosis, Bullous Impetigo, Pediatric Blistering Disorders, Autoimmune Blistering Disease, Dapsone Therapy, Direct Immunofluorescence, Misdiagnosis

1.1 INTRODUCTION

LABD is a rare, blistering autoimmune condition commonly seen in pediatric age groups. It is characterized by the presence of linear IgA deposits in the basement membrane between epidermal and dermal junctions. Although a chronic disease, spontaneous remission may be achievable. It typically presents with vesicles and bullae in a cluster of jewel arrangement, along with central crusting on an inflammatory base [1]. Diagnostic tests include skin biopsy, histopathological studies, and DIF, which shows the deposition of IgA in a linear pattern in the basement membrane. Dapsone and corticosteroids are the treatment of choice. Linear IgA bullous dermatosis may often be misdiagnosed as other bullous diseases, especially bullous pemphigoid and dermatitis herpetiformis. Bullous impetigo is a superficial bacterial infection, predominantly seen in children. It is often caused by Staphylococcus aureus, and the hallmark sign is honey-crusted lesions. Bullous impetigo is managed with antibiotics, amoxicillinclavulanate being the medication of choice [2]. Despite the differences in their etiology and pathophysiology, both LABD and bullous impetigo present with crusting, blistering lesions, making it easy for one disease to be mistaken for the other. This signifies the importance of thorough investigations when diagnosing conditions with similar, non-specific clinical features.

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1.2 CASE PRESENTATION

A 6-year-old boy visited the dermatology outpatient department, presenting with a 10-day history of mild pruritus associated with multiple fluid-filled lesions on his trunk, face, and extremities. The lesions had an acute onset, initially presenting as erythematous macules that progressed to vesicles and bullae. The patient was initially prescribed topical mupirocin and oral amoxicillinclavulanate following a presumptive diagnosis of bullous impetigo. Due to a lack of improvement in symptoms, the patient returned to the clinic. His vitals were recorded to be within the normal range (temperature: 98.6°F, pulse rate: 90 bpm, respiratory rate: 22/min, and blood pressure: 100/65 mmHg). Dermatological examination revealed the presence of multiple tense bullae, clearturbid fluid, targetoid appearance in some lesions and post-bullous erosions with crusting. Nikolsky's sign was negative, and there was no oral or mucosal involvement. His past medical and family history was insignificant. Laboratory investigations revealed a normal CBC (Hemoglobin: 12.4 g/dL, Total Leukocyte Count: 8,500/mm, Neutrophils: 55%, Lymphocytes: 35%, Eosinophils: 6%, ESR: 25 mm/hr, CRP: 11.2 mg/L), Negative ANA and Anti-desmoglein 1 & 3 antibodies. DIF revealed linear deposition of IgA at the dermoepidermal junction. Further histopathological testing noted the presence of subepidermal blisters, neutrophilic infiltrates in dermal papillae but no acantholysis or epidermal necrosis. This led to the diagnosis of Linear IgA Bullous Dermatosis. The patient was prescribed oral dapsone 1 mg/kg/day (18 mg/day), and topical corticosteroid cream (mometasone furoate) to be applied twice daily. Additionally, antihistamines for pruritus and daily cleansing with saline were recommended. Within 7 days, significant clinical improvement of lesions was noted, and no new lesions were present at the 3-week follow-up. The patient was continued on dapsone for 6 weeks with dosage tapering. There was no relapse observed during the 3-month follow-up, and hematologic monitoring of dapsone was unremarkable.

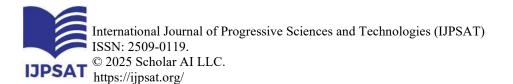
1.3 DISCUSSION

Linear IgA Bullous Dermatosis (LABD) is a rare autoimmune blistering disorder seen more in children than adults, typically between 6 months and 10 years old. The usual presentation is tense vesicles and bullae, typically arranged in a "string of pearls" or annular formation which often gets mistaken for infections such as bullous impetigo, Dermatitis herpetiformis, Bullous pemphigoid, or Epidermolysis bullosa acquisita deposition of IgG, IgM, or C3 due to superficial crusting and the acute onset of lesions [2]. The case presented is of a 6-year-old boy who developed tense bullae and vesicles on the trunk and extremities, sparing the mucosa. LABD is quite often misdiagnosed as Bullous impetigo mainly due to their similar clinical features. Both diseases commonly present with sudden onset bullae, erosions, and crusting, primarily on the trunk and extremities. In the early stages, LABD bullae can resemble flaccid pustules of bullous impetigo, leading to misdiagnosis. The absence of mucosal involvement further increases the confusion regarding the diagnosis [3]. In our case, empirical antibiotic treatment showed no signs of improvement, which led to the realization that the disease was misdiagnosed as bullous impetigo. The correct diagnosis was made through histopathologic exam and direct immunofluorescence (DIF). The diagnostic hallmark of LABD is linear deposition of IgA at the dermoepidermal junction, seen in DIF, which is essential to differentiate LABD from other blistering diseases. Histopathological exam showed subepidermal blisters with neutrophilic infiltration, which supports the diagnosis. [4] Negative anti-desmoglein 1 and 3 antibodies aided in excluding pemphigus foliaceus and vulgaris. Dapsone is the first-line treatment for LABD, showing rapid clinical improvement. Its anti-neutrophillic action aids the resolution of the inflammatory cascade, which is responsible for the blister formation. Additional topical corticosteroids were prescribed to control inflammation and pruritus. Regular hematologic monitoring is essential during dapsone therapy, as there is a risk of hemolysis. LABD in the pediatric population has a good prognosis due to the efficacy of dapsone and the absence of adverse effects. Early recognition and confirmation are important to avoid unnecessary antibiotic usage and prolonged disease course. [5]

CONCLUSION

Our case highlights the challenges faced in diagnosing LABD, mainly in children, where it can closely resemble bullous impetigo. The sudden onset of bullous lesions, crusting, and the absence of systemic symptoms can lead to a misdiagnosis. In our patient, the lack of mucosal involvement and initial resemblance to flaccid pustules of bullous impetigo further delayed the accurate diagnosis. LABD should be considered in the differential diagnosis of bullous skin diseases in children, especially when there is poor response to antibiotic therapy. This case helps us understand the importance of the early recognition of LABD from infectious dermatoses.

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Delay in the right treatment prolongs the patient's discomfort and also exposes the child to unnecessary antibiotics. Timely immunopathological confirmation is necessary for the right diagnosis and management.

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