

# A Rare Case of Chronic Bullous Dermatosi s of Childhood

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## Abstract

The childhood type of linear IgA dermatosis is also called as chronic bullous dermatosis of childhood. Linear immunoglobulin A (IgA) skin disease is a Subepithelial bullous autoimmune disease that can be idiopathic or drug-induced. In our case, it is actually idiopathic as there is no history of drug intake. Children and adults are affected, and the former condition is historically called chronic bullous dermatosis in children.

**Keywords:** Dermatosi s • Idiopathic • Chronic • Autoimmune disease

## Introduction

There are two different types of linear IgA dermatosis, the adult type and the childhood type. The childhood type of linear IgA dermatosis is also called as chronic bullous dermatosis of childhood. It is a rare disorder which manifests in the prepubertal age group. Hereby we present a case of chronic bullous dermatosis of childhood where we, the department of pathology, received a lesional biopsy from a five year old male child who visited the dermatology department.

## Case report

A 5 year old male child presented with complaints of blisters and crusted lesions all over the body including the scalp region for one month associated with severe itching, burning sensation and bloody discharge from the lesions and fever. There is no history of trauma, atopy, photosensitivity, drug history, any inherited disorder or family history. On examination, multiple tense fluid-filled bullae and vesicles were seen all over the body arranged in an annular pattern, peripheral arrangement, and also with areas of hypopigmentation. Multiple crusted lesions were also noted all over the body including the scalp. The clinical diagnosis was given as chronic bullous dermatosis of childhood and a 3.5 mm punch biopsy was taken from the lesion over the left lower back and sent for histopathological examination. We received a single skin-covered punch biopsy specimen measuring 0.5 cc, where the H&E sections showed epidermis with subepidermal bullae along with tiny intraepidermal bullae containing predominantly polymorphs, few eosinophils, and lymphocytes. The superficial dermis showed perivascular collections of polymorphs, eosinophils, and lymphocytes. Pilosebaceous units and deep adnexal structures show mild periadnexal inflammatory infiltrate and the impression was consistent with a clinical diagnosis of chronic bullous dermatosis of childhood (Figures 1-7).

## Discussion

Chronic childhood bullous dermatosis is a childhood chronic acquired epidermal bullous disease characterized by uniform linear deposition of IgA on the basement membrane of the epidermis. It is the



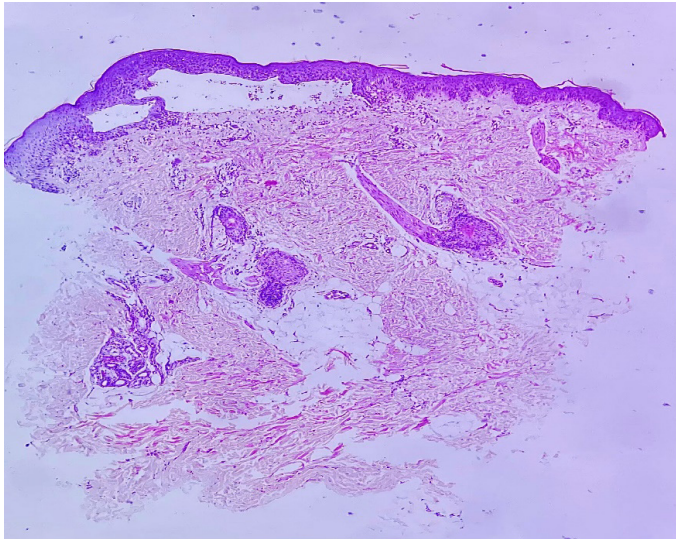
Figure 1. Clinical picture showing lesions over the back.



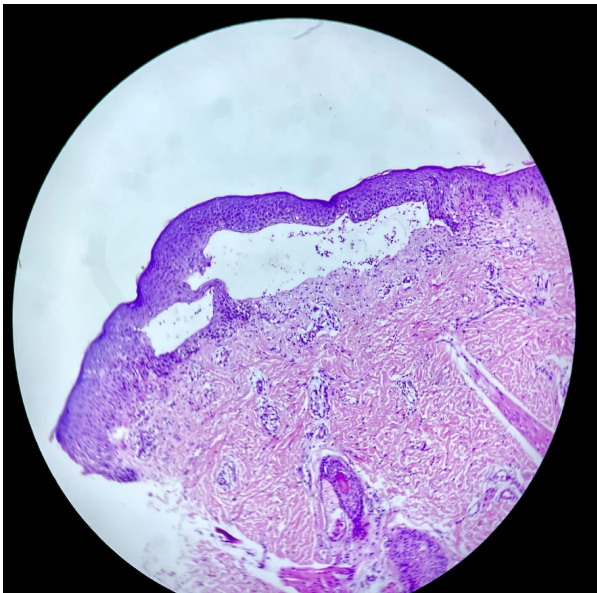
Figure 2. Clinical image showing lesions over the back , shoulders and arms.



Figure 3. Clinical image showing lesions over the cheeks and eyes.

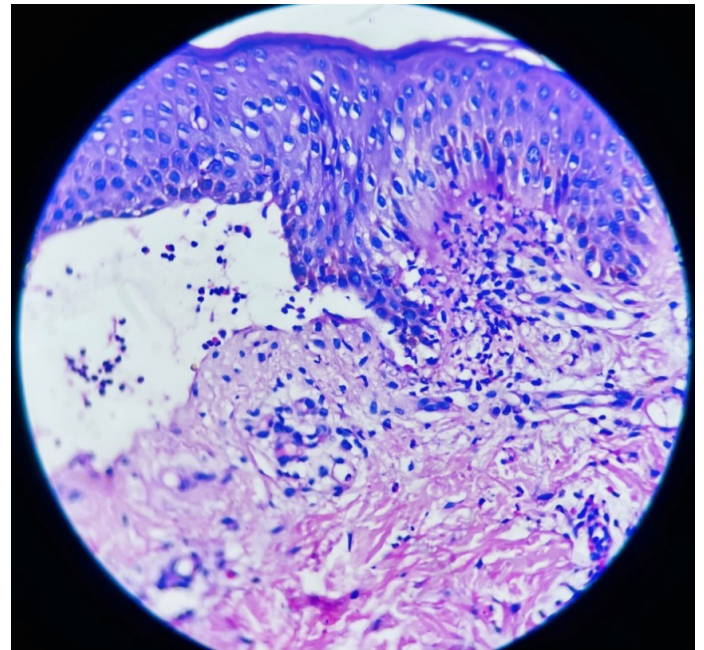


**Figure 4.** Hematoxylin& eosin, scanner view showing epidermis with subepidermal bullae along with a tiny intraepidermal bullae.

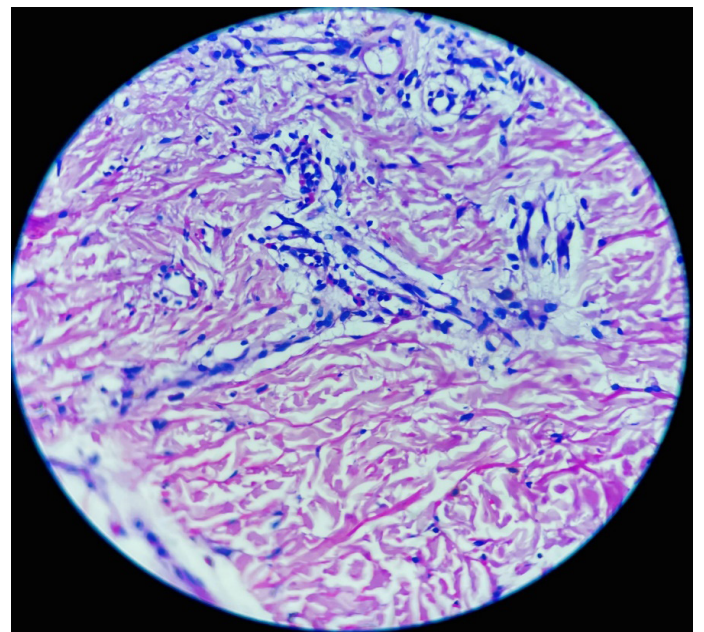


**Figure 5.** Hematoxylin& eosin, low power view showing epidermis with subepidermal bullae along with a tiny intraepidermal bullae.

most common acquired immune bullous disease in children and usually develops before the age of five. Remission is most common between the ages of 6 and 8. Linear Immunoglobulin A (IgA) skin disease is a subepithelial bullous autoimmune disease that can be idiopathic or drug-induced. In our case, it is idiopathic as there is no history of drug intake. Children and adults are affected, and the former condition is historically called chronic bullous dermatosis in children. The clinical manifestations are heterogeneous and resemble other bullous diseases such as bullous pemphigoid and dermatitis herpetiformis [1]. Clinically, it is primarily characterized by monotypic, large tense blisters, often forming "rosette patterns" or "jewel-like" clusters, predominantly in the lower body, pelvis, and lower extremities [2]. Mucosal involvement is important because it is associated with scarring [3]. Histopathological examination of the skin of the vesicles reveals mild inflammatory cell infiltration of the subepithelial vesicles and upper dermis. Direct Immunofluorescence (IF) examination of the peri-lesion area showed linear deposition of IgA at the dermo-epidermal junction and no IgG deposition [1]. Bullous linear IgA disease may be associated with underlying diseases such as inflammatory bowel disease, solid and lymphatic malignancies, and rheumatoid arthritis. Unlike dermatitis herpetiformis, in which IgA's direct immunofluorescent deposits are granular, it is not associated with gluten-sensitive enteropathy. Chronic childhood bullous dermatosis, unlike adult linear IgA dermatosis, is not associated



**Figure 6.** Hematoxylin& eosin, high power view showing intraepidermal bullae containing predominantly polymorphs, few eosinophils and lymphocytes.



**Figure 7.** Hematoxylin& eosin, high power view showing perivascular and periadnexal inflammatory cell infiltrates.

with HLA-B8 [2]. Come to treat the condition and stop the problematic medicine if necessary. Dapsone or Dapsone and corticosteroids-dexamethasone 2 mg/day, enteric sodium mycophenolate for refractory cases [3-5].

## Conclusion

Since chronic bullous dermatosis of childhood, also called linear IgA disease is a rare disorder but can be easily diagnosed with histopathology findings as well as immunofluorescence technique by demonstrating IgA levels in the basement membrane, it is important to diagnose the condition to treat the children as early as possible to facilitate faster treatment and complete remission as it responds well to treatment.

## References

1. Mark, T.H. "Linear IgA Dermatitis" Medscape. (2020)
2. Han, J.H., et.al." A Case of Chronic Bullous Disease of Childhood That Was Reactive to the Antigen of 120 kDa (LAD-1)" *Ann. Dermatol.* 23.2 (2011): 209-212.
3. Sweren, R.J., & Burnett, J.W. "Benign chronic bullous dermatosis of

- childhood: a review". 29.4 (1982): 350–356.
4. Hamodat, M. " Skin non tumor- vesiculobullous and acantholytic reaction patterns -Linear IgA disease". *Pathol. Outl.* 2011
  5. Amanda, O. " Linear IgA bullous disease" Gus Mitchell. November 2021.