# Linear IgA disease in an adult with unusual clinical features

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## ABSTRACT

A 19-year-old male presented with complaints of fluid-filled lesions on the body of 2 weeks duration. On examination, he was found to have multiple tense bullae distributed on the flexures, face, and genitalia with associated oral ulcers and "cluster of jewels" sign. The diagnosis was confirmed by histopathology and direct immunofluorescence. There are very few reports of linear IgA earlier from India according to the literature available so far, that too from the Southern part of the country. The patient had bullous pemphigoid-like lesions typically seen in adults, but the distribution of lesions was akin to that of the "chronic bullous disease of childhood variant" found in children. This case has been reported for rarity in this region and also because the patient had atypical morphology and distribution of lesions.

**Key words:** Dapsone, immunofluorescence, linear IgA

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## INTRODUCTION

Linear IgA disease (LAD) is a blistering disorder characterized by linear deposits of IgA at the basement membrane zone (BMZ).[1] Though LAD is usually idiopathic, infections and drugs are also implicated as probable causes. Mucosal involvement is common occurring in 80% of adults. The diagnosis can be confirmed by direct immunofluorescence. Few earlier reports of LAD from India have been found in the literature. We herein present a case of adult LAD with atypical morphology and distribution of lesions.

## CASE REPORT

A 19-year-old male, resident of Jalandhar presented to the skin OPD with complaints of fluid-filled lesions in both the groins of 2 weeks duration. The blisters ruptured due to friction in 2-3 days time forming raw areas. These raw areas did not increase in size but were associated with discharge of clear fluid. Mild burning sensation was present on the lesions. Similar lesions also appeared on the arms, neck, and face over the duration of 3-4 days and followed the same course. He also developed multiple oral ulcers during the same period. No constitutional or systemic symptoms were present. Patient had an episode of enteric fever 3 months back which resolved in 2 weeks time following treatment with a course of tablet ciprofloxacin. There was no history of similar illness in the past or in related family members. There was also no history suggestive of diabetes, weight loss, or any other major illness in the past.

On examination, he had bilateral tender inguinal lymphadenopathy. Rest of the general examination was normal. Dermatological examination revealed multiple bullae distributed over crural regions, genitalia, perianal area, face, neck, and ears [Figures 1-3]. There was sparing of scalp, axilla, palms, and soles. The bullae were tense and contained clear fluid. Bulla spread sign and Nikolsky's sign were negative. Multiple erosions were interspersed among the bullae in above areas and few of these erosions had overlying honey colored crusts. He also had multiple urticarial lesions on the back [Figure 4] and a single annular plaque with a rim of vesicles on the dorsum of the right hand. Few oral erosions were also present.

Investigations revealed normal hematological and biochemical parameters. ELISA for HIV was negative. X-ray chest and USG abdomen were normal. Tzanck smear from the lesion showed no acantholysis and pus culture grew *Staphylococcus aureus*. Skin

biopsy revealed a subepidermal blister with inflammatory cells predominantly comprising neutrophils, with a few lymphocytes and eosinophils [Figure 5]. The dermis revealed superficial perivascular lymphomononuclear infiltrate. Direct immunoflourescence done on perilesional skin showed a linear homogenous deposition of IgA with the absence of IgG and C3 along the basement membrane zone [Figure 6]. The diagnosis of LAD was thus confirmed. The patient was started on Tab Dapsone 100 mg twice daily and 75% of lesions regressed in 72 h with complete remission within 2 weeks [Figure 7].

## DISCUSSION

LAD is an autoimmune subepidermal vesiculobullous disease that may be idiopathic or drug induced. It is more frequent in China, Malaysia, Sri Lanka, and Thailand.[2] A strong association between the disease and autoimmune haplotypes HLA-B8, CW7, and DR3 has been reported. Vancomycin is the most common drug incriminated while others include captopril, penicillin, ceftriaxone, sulpfonamides, furosemide, lithium, phenytoin, carbamazepine, glibenclamide, atorvastatin, and non-steroidal anti-inflammatory drugs.[3,4] Preceding illnesses such as typhoid, brucella, tuberculosis, varicella, herpes zoster, certain gynecologic infections and upper respiratory infections have all been reported in association with linear IgA dermatosis.[5] Our patient had an episode of enteric fever 3 months prior to the onset of skin lesions, but such a longlag period makes it difficult to proclaim it as etiology. LAD also has been rarely associated with lymphoma, membranous glomerulonephropathy, ulcerative colitis, and solid tumors.[6]

Antibody deposition leads to complement activation and neutrophil chemotaxis, which results in loss of adhesion at

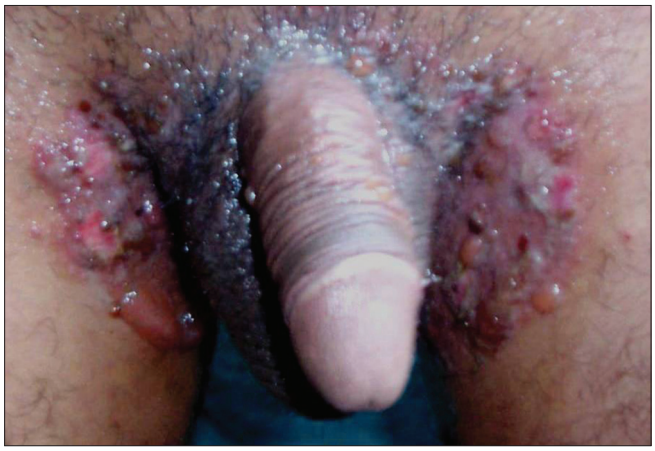

**Figure 1:** Bullae and erosions on genitalia and crural regions

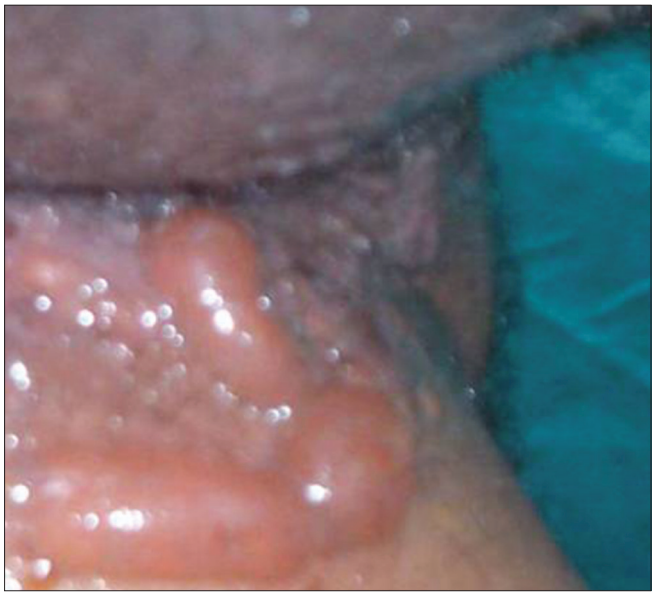

**Figure 3:** Cluster of jewels' appearance on the right crural region

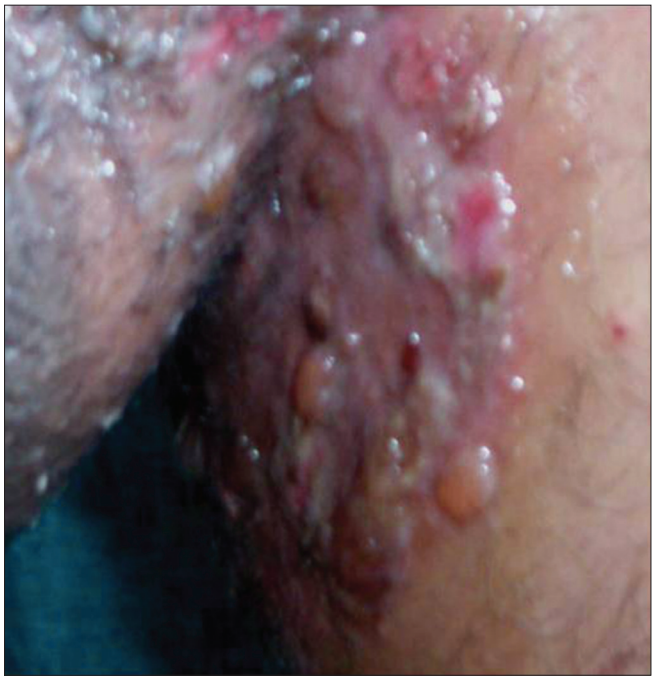

**Figure 2:** Bullae on the left crural region

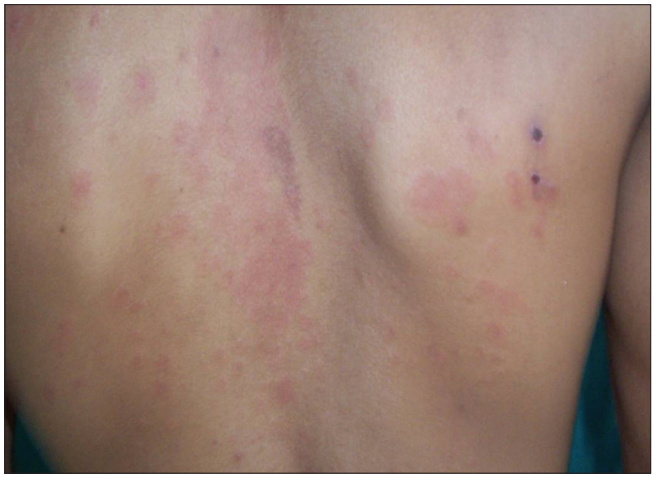

**Figure 4:** Urticarial lesions on the back

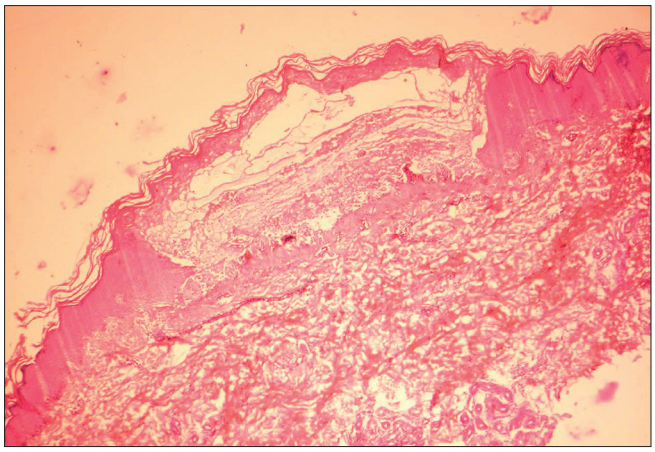

**Figure 5:** Histopathology of skin lesions revealing subepidermal split. H and E, ×10 magnification

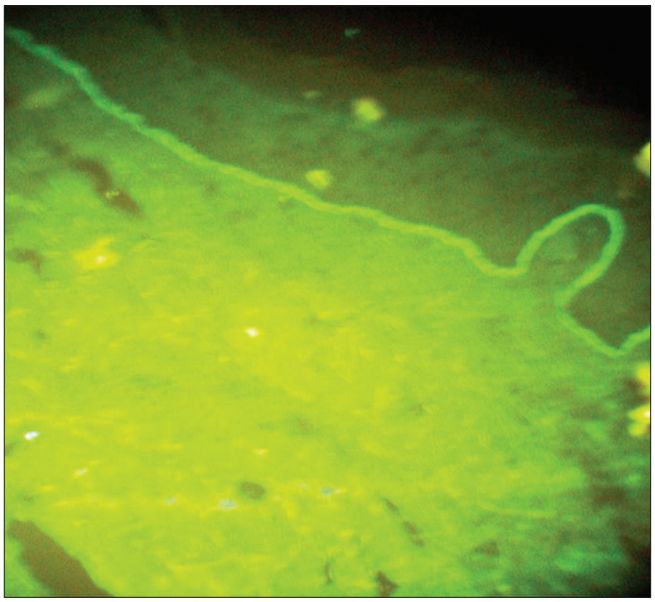

**Figure 6:** Direct immunofluorescence revealing linear IgA deposition

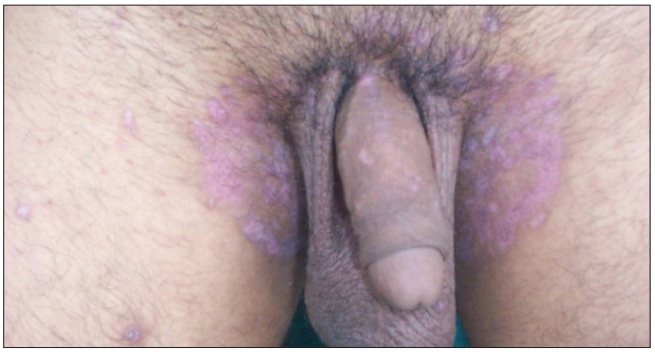

**Figure 7:** Complete healing of lesions on crural and genitalia

the dermal–epidermal junction and blister formation. IgA autoantibodies from LAD sera react with antigens of 97 and 120 kDa, both of which are fragments of the extracellular domain of bullous pemphigoid antigen 180 (type XVII collagen).[7] Antibodies have also been found to act against bullous pemphigoid antigen 230 and collagen VII rarely.

LAD has a bimodal age of onset: Childhood type (originally known as chronic bullous disease of childhood (CBDC)) appearing before the age of 5 and the adult linear IgA disease appearing after the age of 40 years with a slight female predilection. Cutaneous lesions in LAD are heterogeneous and may mimic other bullous diseases. In children, the onset is usually acute and presents as papules, urticarial plaques and polycyclic lesions with blisters situated mainly on the face, abdomen, and perineum.[8] The adult variant is characterized by vesicles and bullae occurring less symmetrically than those seen in dermatitis herpetiformis, but may be distributed in similar locations. Ocular and oral lesions may be present in 50%. Our patient in spite of the older age had blisters distributed in a manner akin to that seen in CBDC. However, bullous pemphigoid-like lesions were also present which does not occur in children. The tendency for new blisters to arise in a ring around an old one is called the “string of beads” sign which is usually present in children, but was present in our case also. Two other clinical phenotypes namely drug induced and resembling cicatricial pemphigoid have also been described.

Histologically LAD shows subepidermal blisters with an infiltrate primarily consisting of neutrophils. Direct immunofluorescence of perilesional skin usually shows deposition of IgA along the BMZ in a homogenous linear pattern, which was true in our patient too. Granular linear deposition at the BMZ along with IgG and C3 deposition are also found rarely.[9] Histopathologically, two varieties of LAD namely lamina lucida and sublamina densa have been described with the sublamina densa variant resembling epidermolysis bullosa acquisita.

The differential diagnosis of this condition includes other subepidermal bullous disorders like dermatitis herpetiformis, epidermolysis bullosa acquisita, bullous pemphigoid, and bullous systemic lupus erythematosus. The strong linear homogenous deposition of IgA along with the absence of IgG and C3, the absence of papillary microabscesses, and eosinophils in the infiltrate, along with the clinical picture and response to Dapsone helped us in ruling out these differentials and confirm the diagnosis of LAD.

Dapsone is the drug of choice for patients with Linear IgA dermatosis. The response is rapid and most lesions resolve within 48-72 h as seen in our case also. Flucloxacillin and sulfamethoxypyridazine have also been tried in few cases as second line treatment with good results.[10] Tetracycline, erythromycin, niacinamide, colchicines, systemic corticosteroids, mycophenolate mofetil, methotrexate, cyclosporine,

azathioprine, interferon-alpha, thalidomide, and intravenous immunoglobulins are the other successful modes of therapy mentioned in the literature.[11,12] There are very few reports of LAD earlier from India according to the literature available so far, that too from the southern part of the country.[13] This case has been presented for its rarity in the region as well as for the fact that the patient had bullous pemphigoid-like lesions typically seen in adults, but the distribution of lesions was akin to that of the CBDC variant found in children.

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