CASE REPORT

LINEAR IgA DISEASE: A RARE ENTITY

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ABSTRACT: Linear IgA Disease (LAD) is chronic, acquired, sub epidermal vesiculo bullous disease of children and adults with cutaneous and mucosal involvement characterized by deposition of IgA basement membrane antibodies. LAD most commonly occurs after puberty having two peak age distributions, teenagers and young adults and second peak at 60 years.

KEYWORDS: Linear IgA Disease, IgA basement membrane antibodies.

INTRODUCTION: Linear IgA (LAD) in adults was often misdiagnosed either as dermatitis herpetiformis or as bullous pemphigoid until 1975, when Chorzelski and Jablonska first suggested that LAD was a distinct entity characterized by linear deposition of IgA (particularly IgA₁) along the basement membrane zone (BMZ).

On the other hand, chronic bullous disease of childhood (CBDC) was recognized earlier as sub epidermal vesiculobullous disease in preschool children characterized by tense blisters, often in an annular arrangement, predominately in flexural areas, especially the lower trunk, thigh and groin. Soon, it was realized that children with CBDC and adults with LAD are simply expression of the same disease in different age groups. Separation of LAD into childhood and adult form depends on whether the onset is before or after puberty.

CASE REPORT: A 12 years old Male patient, from a rural place in Maharashtra presented with chief complaints of severe itching all over body since 20 days and fluid filled lesions all over body since 4 days. It was associated with high grade fever with chills and rigors. Next day, patient's mother noticed reddish lesion with raised border over nape of neck, back and trunk.

Over the next two days the lesion spread to all over body and patient developed tense fluid filled blisters containing clear fluid over the left hand. The fluid filled lesions both increased in number and size over the period of 1 day and spread to upper extremity, lower extremity, chest and back.

Patient presented to a local ayurvedic practitioner and received treatment, oral and injectable (details not known) for a period of 4 days but got no relief. This episode lasted of 8-9 days and lesions healed spontaneously with crust formation and post inflammatory hypopigmentation leaving behind no sequelae.

This was followed by appearance of new lesions (2nd episode) 3 days without any h/o fever but h/s/o weakness, lethargy and pain abdomen. Pain abdomen was mild, continuous in nature in the infra umbilical region, relieved by passing stool. History of burning sensation after rupturing of lesions.

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There was no H/S/O of involvement of perianal and perioral region. No H/S/O fluid filled lesions followed by trauma or blood in stools. No H/S/O joint pain or malaise associated with fluid filled lesion.

No H/S/O yellowish crusting after rupturing of lesion or upper respiratory tract infection prior to fluid filled lesion. No H/S/O fluid filled lesions over sun exposed parts. No H/S/O appearance of new lesions around crusting.

On examination, he was afebrile with pulse of 108/min and Blood pressure of 110/72mm of Hg. Rest general examination was normal. On skin examination, patient had multiple vesiculo bullous tense blisters present over erythematous annular urticated plaques on chest, back, B/L upper and lower extremities, both on flexor and extensor aspect. Erosions topped with crust over buttocks, feet, trunk and back (Fig 1).

Multiple post inflammatory hypo pigmented patches present all over body (Fig.2). Nikolsky sign negative.

Fig. 1: Multiple vesiculobullous tense blisters present over erythematous annular urticated plaques.

Fig. 1

Fig. 2: Multiple postinflammatory hypopigmented patches.

Fig. 2

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Laboratory investigations were as given in Table no.1

Hb	11.6 gm/dl
TLC	6000
DLC	N70, L26, M2, E2
Platelets	2.45 lac
Serum Na	124
Serum K	5.4
ESR	35 mm
G6PD (quantitative)	257 (WNL)
Urine RM	WNL
Stool microscopy	WNL
Stool culture	No. organism grown
Tzanck smear	Acantholytic cells present
Table no. 1	

USG abdomen and pelvis- was within normal limits and Mantoux test – negative.

Direct immunofluorescence of unaffected skin showed linear deposition of IgA along basement membrane (Fig. 3).

Patient was started with Dapsone (200mg/day) and oral prednisolone and had shown good results within 2 weeks (Fig. 4).

DISCUSSION: By definition, Linear IgA disease is chronic, acquired, sub epidermal blistering disease of children and adults with cutaneous and mucosal involvement in 80%, characterized by IgA basement membrane antibodies. LAD most commonly occurs after puberty having two peak age distributions, teenagers and young adults and second peak at 60 years although the true incidence of disease is not known. It is separate from dermatitis herpetiformis on the basis of immunopathology and lack of association with Gluten sensitive enteropathy.

Patient presents with combination of annular and grouped papules, vesicles and bullae that are distributed symmetrically on trunk and extremities including elbows, knees and buttocks. Lesions are very pruritic. Clusters of blisters with new blisters forming around old blisters – String of pearls sign are seen.

Pathogenesis lies in the circulating antibodies against various epidermal basement membrane antigens like BP180⁽³⁾ and Collagen XVII have been found. The HLA B8 DR3, known as autoimmune extended haplotypes, is associated with LAD.^(4,5,6)

Histological features are not specific for this condition. Sub epidermal vesicles may contain Eosinophils (suggestive of bullous pemphigoid) or neutrophils (suggestive of dermatitis

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herpetiformis). Microabscesses (polymorphonuclear and eosinophils) at tips of dermal papillae. Dermal infiltration of neutrophils and eosinophils.

Direct immunofluorescence of unaffected skin shows linear deposition of IgA along basement membrane. Direct salt splitting of biopsies or raising suction blisters in patients can show autoantibodies associated with upper aspect of artificial blisters (as seen in bullous pemphigoid) or the dermal aspect (as seen in epidermolysis bullosa acquisita) or both.

Fig. 3: Linear IgA deposits on immunofluorescence

Fig. 4: Post treatment

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There is no definitive treatment; therapy is aimed at controlling the disease while awaiting spontaneous remission. Topical steroid may be used for mild cases and oral lesion. Dapsone (50–200 mg; 1–2mg/kg per day in children) is commonly used alone with prednisolone (0.5–1mg/kg per day).^(1,2,7) Sulfamethoxypyridazine (0.5–1.5g/day) is also an effective treatment in children as it causes less hemolysis.⁽⁸⁾

Recent trials have shown complete remission by flucoxacillin within 3 to 4 months from starting the therapy. Reports have also shown the benefit of colchicines (0.5–2mg/day), cyclosporine, tetracycline, nicotinamide, mycophenolate and erythromycin to be effective.

SUMMARY: Linear IgA Disease (LAD) is chronic, acquired, subepidermal blistering disease of children and adults with cutaneous and mucosal involvement characterized by IgA basement membrane antibodies, occurring most commonly occurs after puberty having two peak age distributions, teenagers and young adults and second peak at 60 years.

It characterized by tense fluid filled blisters and direct immunofluorescence showing deposition of Linear IgA in the basement membrane and having remission on its own after 3-6 years in 30% - 60% of patients. High index of suspicion is necessary to diagnose the disease.

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