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Original Research Paper

Dermatology

CASE REPORT-A 73 YEAR OLD FEMALE WITH LINEAR IgA/IgG BULLOUS DERMATOSIS (LAGBD)

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ABSTRACT

Immunobullous disorders are a heterogeneous group of disorders having varied clinical presentations.Linear IgA/IgG bullous dermatosis (LAGBD) is an immunobullous disorder with an overlap spanning the clinical and immunological spectrum between Bullous pemphigoid[BP] and Linear IgA disease[LAD].It is a rare condition. There are a few reports of patients showing a mixed linear deposition of IgG and IgA along the dermoepidermal junction on DIF.A 73 year old female patient which was provisionally diagnosed as Bullous Pemphigoid and who did nor respond to oral corticosteroids was finally diagnosed as LAGBD based on equivocal linear staining of both IgA and IgG on Direct Immunofluorescence(DIF) and significant response to Dapsone.DIF test is very helpful in the diagnosis of autoimmune bullous disorders.It helps to differentiate histologically similar conditions which differ in their treatment protocols and prognosis.A holistic approach of correlation of clinical, histological and immunofluorescence data is necessary for a definite diagnosis. Dapsone may play an important role in the management of Immunobullous disorders.

KEYWORDS : LAGBD, DIF, BP, LAD, Dapsone

INTRODUCTION

Immunobullous disorders are a heterogeneous group of disorders having varied presentations.

These disorders may overlap in morphology and immunopathology.

DIF, Histopathology, response to treatment in addition to clinical presentation are important in establishing the diagnosis.A holistic approach is required for arriving at the correct diagnosis.

Linear IgA/IgG bullous dermatosis (LAGBD) is an immunological overlap spanning the clinical and immunological spectrum between Bullous pemphigoid [BP] and Linear IgA disease [LAD].LAD is characterized by linear deposition of IgA while Bullous Pemphigoid (BP) has linear deposits of IgG along the epidermal basement membrane zone which is seen on Immunofluorescence. A few cases known as Linear IgA/IgG bullous disease(LAGBD)/Mixed Immunobullous disease show equivocal linear staining with both IgA and IgG and show clinical features of both diseases[1].

Case Report:

A 73 year old female presented to our hospital with chief complaints of raw areas, fluid filled blisters all over the body associated with itching and burning sensation since 1 month . c/o painful oral lesions associated with difficulty in swallowing since 4 weeks.

History of present Illness-Patient was apparently asymptomatic 1 month back when she developed fluid filled blisters first on chest then progressed to involve back, trunk, lower limbs associated with itching and burning sensation.

Blisters ruptured on their own to form raw areas. History of painful oral lesions associated with difficulty in swallowing. Past history : known case of hypertension since 10 years, she is on telmisartan 40 mg.

Personal and Family History : Not Significant.

Vitals are stable. General and Systemic Examination-No abnormality detected On Cutaneous Examination,Bilaterally symmetrical multiple erosions ,with few of them covered with crusts on chest, trunk, back, upper 1/3rd of both shoulders, lower limbs,itchy vesicles and a few bullae which were symmetrically distributed over the body Few vesicles are seen on the chest.

Multiple small erosions over the buccal mucosa Ocular and nasal mucosa were normal.Hair and nails were normal.No urticarial lesions(as seen in BP).Nikolsky's sign negative.The characteristic String of Pearls appearance(as seen in LAD) over skin was absent.

Patient was diagnosed as Bullous Pemphigoid outside and was advised low dose oral steroids (0.5mg/kg) before presenting here but had not shown any improvement even after using medication for 1 week.

A punch biopsy was done for the patient and she was started on high dose corticosteroids(1mg/kg). Direct Imm unofluorescence (DIF) of perilesional skin was done.

After 1 week patient came to OPD with increase in number of lesions.

Histopathology showed subepidermal bulla with predominantly neutrophilic infiltration(neutrophils in LAD) and a few eosinophils(eosinophils in BP). Direct Immunofluorescence(DIF) revealed Linear deposition of IgG [+++], IgA [+++], and C3[++], along the basement membrane zone.The patient who earlier did not show clinical improvement with oral Prednisolone(given in BP)-1 mg/kg/day showed good response with oral Dapsone-100 mg qd(given in LAD)

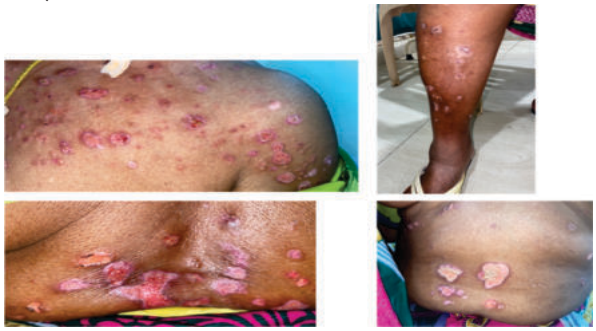

Figure 1-Bilaterally symmetrical multiple erosions ,with few of

them covered with crusts on chest, trunk, back, upper 1/3rd of both shoulders, lower limbs

Erosions seen on buccal mucosa.
Other mucosa normal.
Hair and nails- normal.

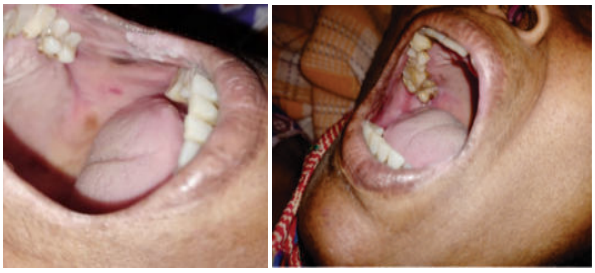

Figure 2- Erosions Over Buccal Mucosa

Wbc count is raised [20,000/cumm] and ESR is elevated[100 mm/hr] and rest parameters are within normal limits

Histopathological Examination :- There is a subepidermal bullae which is chiefly composed of neutrophils.Upper dermis shows perivascular lymphocytic infiltrate

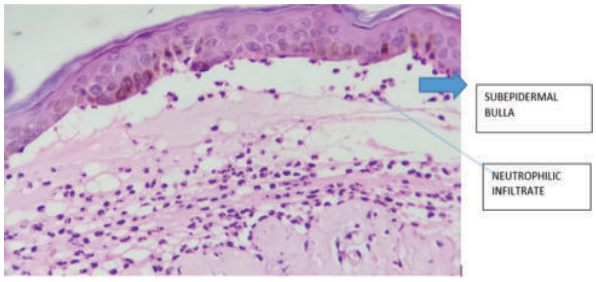

Figure 3- Subepidermal Bulla With Neutrophilic Infiltrate

Direct Immunofluorescence(dif):

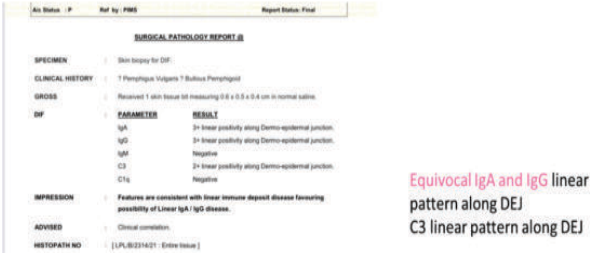

Figure 4- Dif With Equivocal Staining

Final Diagnosis:
Linear Iga/igg Bullous Dermatosis.

Treatment : Patient was kept on Tab DAPSONE 100 mg qd and topical steroids. The patient showed marked improvement within a week.

DISCUSSION:

Linear IgA/IgG bullous dermatosis [LAGBD]is an immunologic overlap spanning the clinical and immunological spectrum between Bullous pemphigoid [BP] and linear IgA bullous disease [LABD].

As we have seen in this where patient was started on steroid thinking BP ,but patient has not shown improvement, later DIF helped in aiding the final diagnosis of LAGBD.

As patient has shown good response to dapsone, this clinical mixed spectrum is more towards LABD.

Table 1- Differences Between Bullous Pemphigoid And Linear Iga/igg Bullous Dermatosis

	Bullous Pemphigoid	Linear Iga/Igg Bullous Dermatosis	Linear Iga Bullous Dermatosis
Auto-Antibodies	Bp180 Bp230	Bp180 Bp230 Laminin-332	97kda And 120kda Fraction Of BP 180 BP-180 BP230 Anti - Type7 Collagen
Clinical Findings	Prurities, Tense Blisters And Vesicle, Erosions And Crusting, Urticarial Plaques, Oral Involvement And Other Mucous Membrane Rarely Involved	Prurities, Tense Blisters And Vesicle, Erosions And Crusting, Annular Vesicular Pattern, Ocular And Oral Mucosa Involved	Prurities, Tense Blisters And Vesicle, Erosions And Crusting, Annular Or Herpetiform Vesicular Pattern, Oral, Ocular, Nasal, Esophageal Involvement In 80% Cases
Histopathological Examination [Subepidermal Blisters]	Eosinophilic And Mononuclear Cell Infiltrate	Predominant Neutrophilic Infiltrate	Neutrophilic Infiltrate

There are a few reports of patients showing a mixed linear deposition of IgG and IgA along the dermoepidermal junction on DIF.It is not well established whether to term these as linear IgA disease, a distinct entity or a variant of bullous pemphigoid creating a diagnostic conundrum[3].Such cases may be a part of LAD-BP spectrum and usually respond well to oral dapsone (inhibits neutrophil chemotaxis[4]).DIF helps to identify such cases and aids in the management.

Clinical picture of immunobullous disorders present in a protean manner and is often perplexing[2].A holistic approach is required for coming to the correct diagnosis.

DIF test provides a major contribution for the diagnosis of autoimmune bullous disorders.

It helps differentiate histologically similar conditions which differ in their treatment protocols and prognosis.

Correlation of clinical, histological, immunofluorescence data is necessary for definite diagnosis.

The role of Dapsone in the management of immunobullous disorders needs to be further evaluated as Dapsone might become a vital drug in the armamentarium of doctors for treatment of bullous disorders[4].

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