

Case Report

A middle-aged male with bullous lesions and co-morbidities

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ABSTRACT

Epidermolysis bullosa acquisita (EBA) is a rare acquired type of mechano bullous disease affecting the dermal-epidermal junction (DEJ) of trauma prone acral surfaces. It manifests as tense vesicles, bullae, and milia and typically heals as atrophic hypo or hyperpigmented scars. Classic noninflammatory mechano bullous EBA typically presents at a mean age of 48 years. A 57 years old male patient, presented with itchy fluid filled lesions over the face since, 2 months. On cutaneous examination, discrete and grouped papulo-vesicles on an erythematous base and areas of erosions present over both eyelids, erythematous annular plaque with vesicles at the border present over the left cheek. Blood investigations revealed an increased total blood count, total bilirubin, direct bilirubin, and positive HCV antibodies. Histopathology showed a sub-epidermal blister and surrounding dermis shows inflammatory infiltrate composed of eosinophils and neutrophils. Direct immunofluorescence revealed linear deposits of IgG and C3 at the DEJ. The patient was started on dapsone.

Keywords: Epidermolysis bullosa acquisita, Blistering disorders, DEJ

INTRODUCTION

Epidermolysis bullosa acquisita (EBA) is an acquired mechano-bullous disorder that results from the presence of IgG antibodies against collagen VII, the main anchoring filament in the lamina densa and sublamina densa region of the basement membrane zone of the dermal-epidermal junction (DEJ). EBA has four subtypes 'classical' mechanobullous, bullous pemphigoid-like, cicatricial pemphigoid-like, and IgA bullous dermatosis.^{1,2} This disease commonly presents in adulthood (mean age of 48 years), and rare in adolescence or young adulthood. This case report is unique given the patient's age of onset, atypical clinical presentation and positive HCV antibodies. In addition, the patient also responded well to dapsone. The objective of presenting this case report is to bring the atypical nature of this disease to light.

CASE REPORT

A 57 years old male patient, driver by occupation presented to the clinic with chief complaints of itchy fluid filled lesions over the face since, 2 months, which rupture in a few days to leave behind raw areas. No h/o lesions in the oral cavity, photosensitivity, hair dye application. Patient is a known case of diabetes mellitus on tab. Teneligliptin 20 mg, hypertension on tab. Amlodipine 5 mg (since many years, no recent change of medications).

Patient also had an HCV positive status on treatment since, 1 month with tab ribavin (ribavirin 200 mg) and tab ledihev (ledipasvir 90 mg and sofosbuvir 400 mg), diagnosed 1 month back when patient had consulted a physician for symptoms of fatigue, decreased appetite and indigestion.

Cutaneous examination

Discrete and grouped papulo-vesicles on an erythematous base and areas of erosions present over both eyelids. Erythematous annular plaque with vesicles at the border present over the left cheek. Multiple tender indurated plaques with pointing pustules present over the trunk and extremities, oral mucosae showed few erosions and areas of crusting whereas genital mucosae were normal, muddy conjunctiva was present, palms and soles were normal.



Figure 1: Discrete and grouped papulo-vesicles on an erythematous base and areas of erosions present over both eyelids. Erythematous annular plaque with vesicles at the border present over the left cheek.



Figure 2: Close up of the lesions over the eyelids and left cheek.



Figure 3: Close up view of the lesions over the left cheek.



Figure 4: Multiple vesicles and crusted erosions with areas of depigmented macules and patches present over left side of lower face.

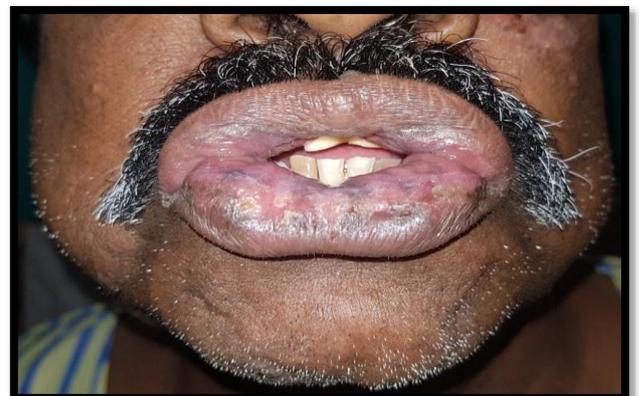


Figure 5: Crusted erosions present over the inner aspect of lower lips.

Blood investigations were done and total WBC count was found to be 16,300 cells/ cum and LFT was normal

except for high total bilirubin (1.23 mg/dl) and direct bilirubin (0.67 mg/dl). RFT, urine routine, FBS, PPBS, HbA1C were within normal limits. HCV antibodies were found to be reactive (31.0).

Two 3 mm punch biopsies were taken from vesicular lesions over the patient's left cheek for haematoxylin and eosin (H and E) and direct immunofluorescence (DIF). H and E showed a sub-epidermal blister and inflammatory infiltrate composed of eosinophils and neutrophils in the surrounding dermis. Blister cavity showed haemorrhage and few scattered inflammatory cells composed of eosinophils and few neutrophils (Figure 5 and 6).

DIF revealed linear deposits of IgG (++) and C3 (++) at the dermo-epidermal junction (Figure 7) and salt-split skin technique revealed floor pattern of deposits of C3 (Figure 8). Both findings were reported to be in keeping with epidermolysis bullosa acquisita.

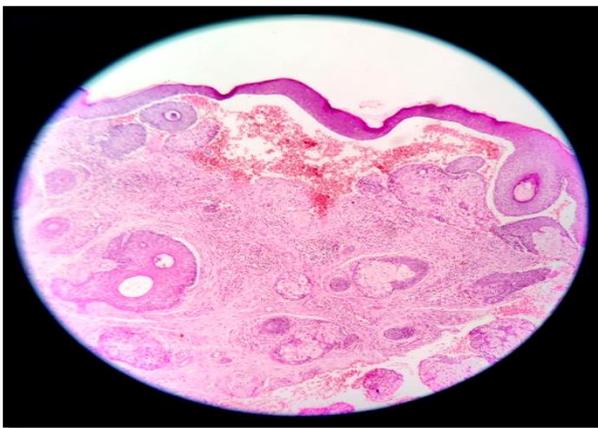


Figure 6: Sub-epidermal blister seen. Blister cavity shows haemorrhage and a few scattered inflammatory cells. Dermis shows scattered areas of inflammatory infiltrate (10X).

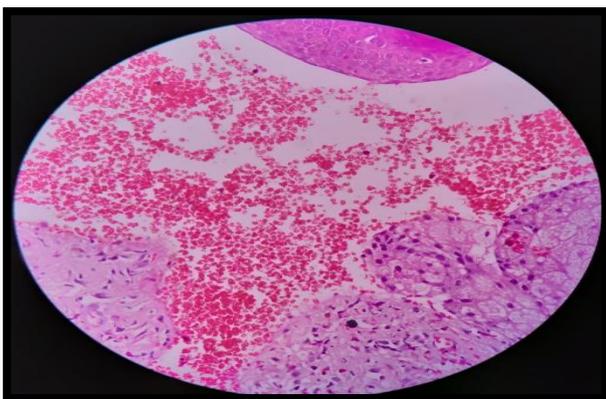


Figure 7: Blister cavity shows haemorrhage and a few scattered inflammatory cells composed of eosinophils and a few neutrophils (40X) view).

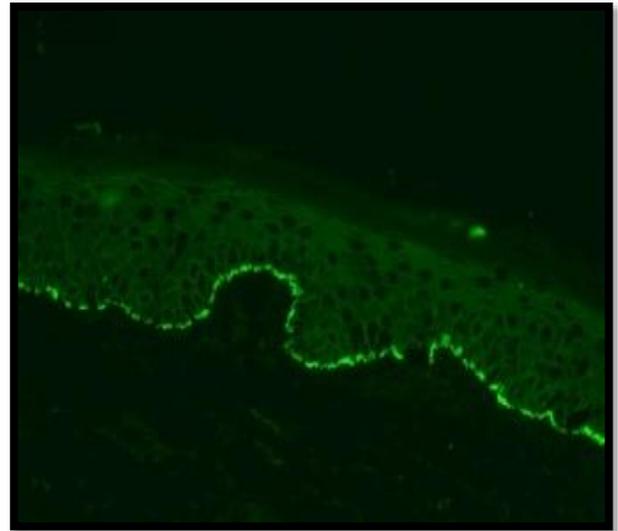


Figure 8: DIF-linear deposits of IgG (++) and C3 (++) at the dermo-epidermal junction.

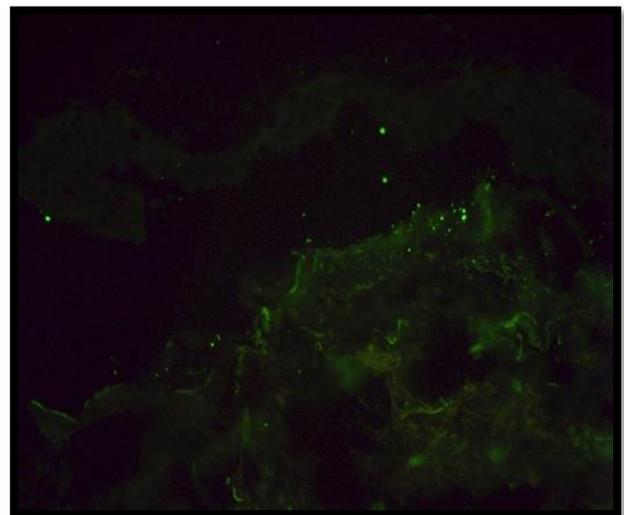


Figure 9: Salt-split skin technique-floor pattern of deposits of C3.

Final diagnosis

Epidermolysis bullosa acquisita, HCV positive status on antiviral drugs, furunculosis (MRSA), diabetes mellitus and hypertension on treatment.

Treatment

Inj. methylprednisolone 80 mg IM stat, tab dapsone 100 mg 0-0-1x3 weeks, injection ceftriaxone 1g IV BDx10 days, tab hydroxyzine hydrochloride 25 mg 0-0-1x3 weeks, framycetin cream BDx1 week, cap B-complex 0-0-1x3 weeks, tab teneligliptin 20 mg 1-0-0x3 weeks tab amlodipine 5 mg 1-0-0x3 weeks, tab ledihev (ledipasvir 90 mg and sofosbuvir 400 mg) 0-0-1x3 weeks.

Follow up after 3 weeks



Figure 10: Hyperpigmented patches present over the upper eyelids and left cheek.



Figure 11: Close up view of the healed lesions on the left cheek.

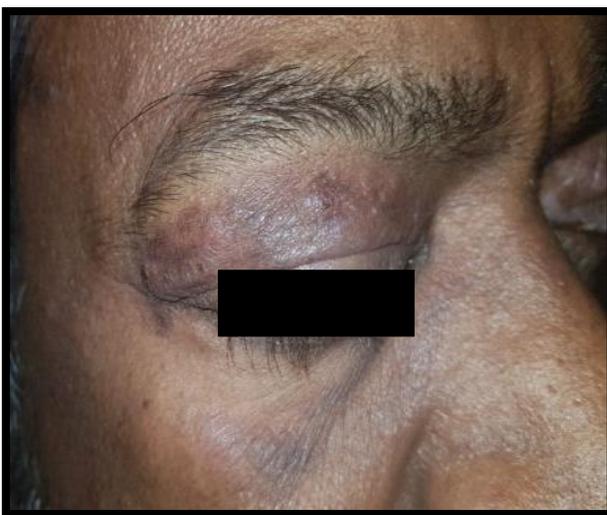


Figure 12: Close up view of the lesion on the right eye lid.

DISCUSSION

Epidermolysis bullosa acquisita (EBA) is a chronic autoimmune subepidermal blistering disease of the skin and mucous membranes. Caused by antibodies targeting type VII collagen, the major component of anchoring fibrils that connect the basement membrane to dermal structures. The classic presentation is characterized by blisters, mild mucosal involvement, and healing with dense scars primarily at trauma-prone areas.^{1,3,4}

Clinical presentations of EBA

Classic presentation: Non-inflammatory bullous disease with an acral distribution that heals with scarring and milia formation.

Bullous pemphigoid (BP): As a widespread, inflammatory vesiculobullous eruption involving the trunk, skin folds and extremities.

Cicatricial pemphigoid (CP): Erosions and scars on the mucosal surfaces of the mouth, upper oesophagus, conjunctiva, anus, vagina with or without similar lesions on the glabrous skin.

Presentation reminiscent of Brunsting-Perry pemphigoid: Subepidermal bullae, residual scars, IgG deposits at the DEJ, and minimal or no mucosal involvement, predominant head and neck distribution.

Reminiscent of linear IgA bullous dermatosis or chronic bullous disease of childhood: Subepidermal bullous eruption, a neutrophilic infiltrate and linear IgA deposits at the BMZ on DIF, tense vesicles arranged in an annular fashion and involvement of mucous membranes.^{3,4}

The differential diagnosis for our patient included were bullous pemphigoid, epidermolysis bullosa acquisita, bullous lichen planus, bullous systemic lupus erythematosus, porphyria cutanea tarda. Classic noninflammatory mechano bullous EBA typically presents at a mean age of 48 years.¹

In this case, our patient presented with discrete and grouped papulo-vesicles on an erythematous base and areas of erosions present over both eyelids, erythematous annular plaque with vesicles at the border present over the left cheek. Patients with both inflammatory and noninflammatory EBA are also prone to erosions and adhesions, particularly on mucosal surfaces, which in our patient was not significantly involved except for few erosions and crusting on the inner side of the lower lip.¹

EBA has also been associated with many systemic diseases, most commonly inflammatory bowel disease, with 25% of patients having concomitant Crohn disease.⁴ Our patient denied any systemic symptoms, including gastrointestinal or extra-intestinal symptoms of

inflammatory bowel disease. However, our patient had an HCV positive status.

This case serves as an important reminder to interpret pathological results in the context of the clinical findings. Classic DIF findings show linear IgG deposits and complement at the DEJ. Our patient's DIF revealed linear deposits of IgG and C3 at the DEJ. When aiming to characterize histological features of EBA, Callot-Mellot et al found that while 100% of patients had IgG at the DEJ, a significant number were also found to have IgA (50%) and IgM (33%) deposited at the DEJ.⁶ IgA and IgM at the DEJ can aid in differentiating EBA from bullous pemphigoid, which characteristically demonstrates only IgG and C3 at the DEJ. When it comes to treatment options for EBA, data are limited. Often empiric treatment is tried with patients and the disease tends to be fairly refractory to treatment but the inflammatory forms respond well to treatment.⁷

Long-term systemic glucocorticoids have proven to be less effective for EBA than for other blistering disorders. Some improvements have been documented with immunosuppressive agents.⁸ We initiated treatment with dapsone, since treatment recommendations for most patients consist of colchicine or dapsone, in monotherapy or in combination.⁸ For patients with EBA refractory to colchicine and dapsone, rituximab, an anti-CD20 monoclonal antibody, is a promising option but the high costs represent a major barrier in the use of these agents for patients with EBA.⁹ A curative treatment for EBA does not currently exist. Hence, the goal of treatment is long-term remission of the disease. It usually has a prolonged course.

CONCLUSION

EBA, and in other rare diseases in general, current knowledge could be enriched with the discovery of additional clinical phenotypes that at the moment may be under-diagnosed or under-reported in the literature.

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Ethical approval: Not required

REFERENCES

1. Cyr J, Liu A, Ghazarian D, Siddha S. Case Report of a 21 years old man with epidermolysis bullosa acquisita. *J Cutaneous Med Surg.* 2018;22(3):356-8.
2. Das JK, Sengupta S, Gangopadhyay AK. Epidermolysis bullosa acquisita. *Indian J Dermatol Venereol Leprol.* 2006;72:86.
3. Woodley DT, Chen M. Epidermolysis Bullosa Acquisita. Wolf K, Goldsmith LA, Katz S I, Gilchrist B A, Paller A S, Leffell D J. *Fitzpatrick's Dermatology in General Medicine.* 7th ed. Mc Graw Hill Publishers; 2008: 494-499.
4. Wojnarowska F, Venning VA. Immunobullous diseases. Burns T, Breathnach S, Cox N, Griffiths C. *Rook's textbook of Dermatology.* 8th ed. Wiley-Blackwell publishers; 2010: 40.51-40.56.
5. Chan LS, Woodley DT. Pemphigoid: bullous and cicatricial. In: Lichtenstein LM, Fauci AS, eds. *Current Therapy in Allergy, Immunology and Rheumatology.* St Louis, MO: Elsevier; 1996: 5-93.
6. Mellot CC, Bodemer C, Caux F. Epidermolysis bullosa acquisita in childhood. *Arch Dermatol.* 1997;133(9):1122-6.
7. Lehman JS, Camilleri MJ, Gibson LE. Epidermolysis bullosa acquisita: concise review and practical considerations. *Int J Dermatol.* 2009;48:227-35.
8. Gurcan HM, Ahmed AR. Current concepts in the treatment of epidermolysis bullosa acquisita. *Expert Opin Pharmacother.* 2011;12:1259-68.
9. Bevans SL, Sami N. The use of rituximab in treatment of epidermolysis bullosa acquisita: three new cases and a review of the literature. *Dermatol Ther.* 2018;31:12726.

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