International Journal of Research in Dermatology
Manikanta RSS et al. Int J Res Dermatol. 2025 May;11(3):273-276
http://www.ijord.com

DOI: https://dx.doi.org/10.18203/issn.2455-4529.IntJResDermatol20251054

Case Report

Epidermolysis bullosa acquisita in a 53-year-old female with extensive cutaneous and mucosal involvement

Reddy Siva Sai Manikanta*, K. V. T. Gopal, P. V. Krishnam Raju, N. Krishna Sagar

Department of Dermatology, Maharajah's Institute of Medical Sciences, Vizianagaram, Andhra Pradesh, India

Received: 07 March 2025
Accepted: 02 April 2025

***Correspondence:**
Dr. Reddy Siva Sai Manikanta,
E-mail: saimanikanta234@gmail.com

ABSTRACT

Epidermolysis bullosa acquisita (EBA) is a rare autoimmune blistering disorder characterized by subepidermal blisters, typically caused by an autoimmune response against type VII collagen, a key structural component of the dermal-epidermal junction. Here, we report a case of EBA in a 53-year-old female with extensive multiple bullae, erosions over extremities, trunk, genitaila. HPE revealed a sub-epidermal blister with fibrin, neutrophils, and some eosinophils, DIF showed IgG positivity (+3) in a linear pattern along the dermal-epidermal junction. enzyme immunoassay (EIA) for anti-collagen type VII antibodies was positive. Based on clinical features and investigations patient is diagnosed as Epidermolysis Bullosa Acquisita. She was treated with systemic steroids and topical steroids with gradual tapering. This case highlights the importance of early diagnosis and treatment in managing this condition effectively.

Keywords: Epidermolysis bullosa acquisita, Autoimmune blistering disorder, Sub-epidermal blister, Direct immunofluorescence, Mucosal involvement, Type VII collagen

INTRODUCTION

Epidermolysis bullosa acquisita (EBA) is a rare, autoimmune blistering disorder characterized by the formation of subepidermal blisters primarily affecting the skin and mucous membranes. It results from the production of autoantibodies against type VII collagen, a key structural protein found in the dermal-epidermal junction, which plays a vital role in anchoring the epidermis to the dermis.[1]

This autoimmune response leads to the disruption of the basement membrane zone, resulting in blister formation. The diagnosis of EBA can be challenging due to its similarity to other blistering disorders, and often requires a combination of clinical evaluation, histopathology, direct immunofluorescence, and specific antibody testing. We report a case of EBA in a middle-aged woman who presented with blisters and raw areas in oral mucosa and multiple sites over body.[2]

CASE REPORT

A 53-year-old female presented with a 3-month history of multiple fluid-filled lesions and erosions that initially appeared on the extremities, followed by the trunk and genital region. The lesions gradually ruptured, forming raw areas. Additionally, the patient reported fluid-filled lesions on her tongue for the past 10 days.

On examination, the patient had an intact blister over her left forearm (Figure 1), an intact blister with healing erosions over the right posterior arm, and active erosions on the inner aspects of both thighs. She also had vesicular lesions over the lateral aspect of her tongue and a hemorrhagic blister on the underside of the tongue (Figure 2).

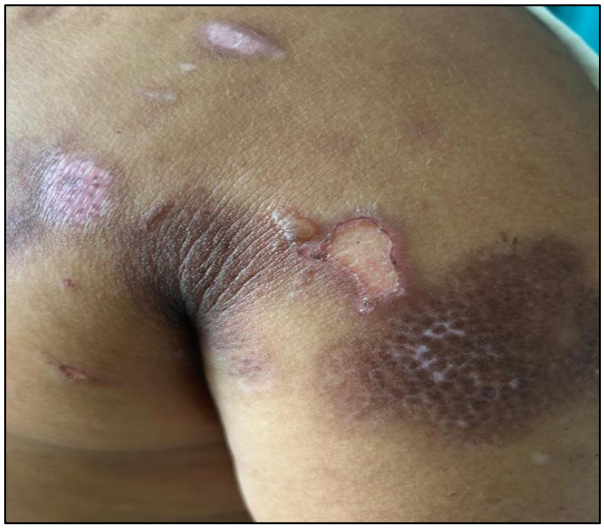


Figure 1: Blister over her left forearm.

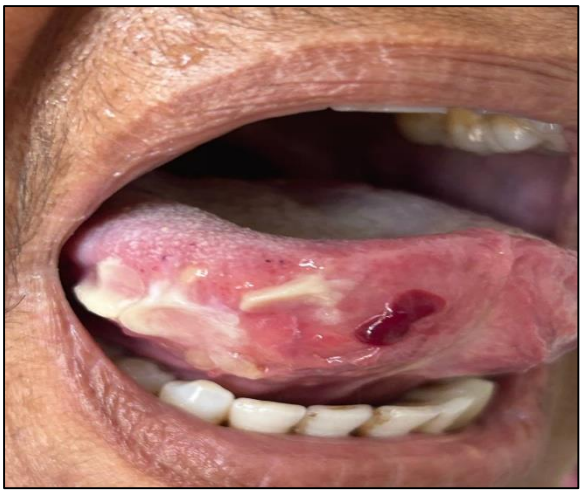


Figure 2: Vesicular lesions over the lateral aspect of her tongue and a hemorrhagic blister on the underside of the tongue.

The clinical differential diagnosis for this presentation included bullous pemphigoid, bullous systemic lupus erythematosus (SLE), Epidermolysis Bullosa Acquisita, and linear IgA disease. Given the patient's presentation of mucosal involvement along with widespread blistering, EBA was considered a likely diagnosis.

Two punch biopsies were performed for diagnostic purposes. One biopsy was taken from the lesion for histopathological examination, and the second was from the peri-lesional skin for direct immunofluorescence (DIF). Histopathology revealed a sub-epidermal blister with fibrin, neutrophils, and some eosinophils. The superficial dermis showed a mild perivascular inflammatory infiltrate composed of lymphocytes and a few eosinophils (Figure 3).

Direct immunofluorescence results revealed IgG positivity (+3) in a linear pattern along the dermal-epidermal junction (DEJ), exclusively on the floor of the blister. C3c positivity (+3) was also noted in a linear pattern along the DEJ, confirming the autoimmune nature of the blistering (Figure 4). Additionally, an enzyme immunoassay (EIA) for anti-collagen type VII antibodies was positive, further supporting the diagnosis of epidermolysis bullosa acquisita (Figure 5).

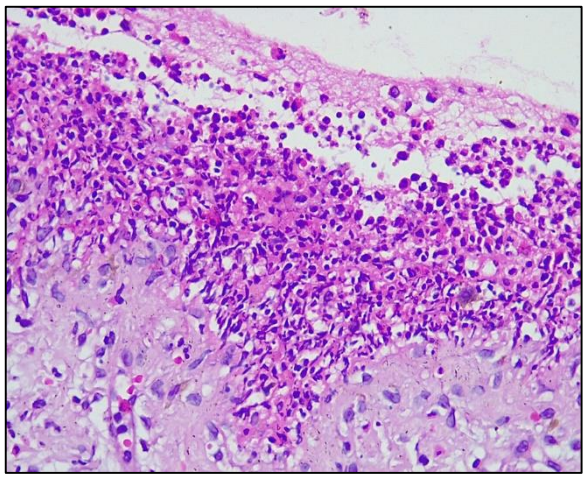


Figure 3: Superficial dermis showed a mild perivascular inflammatory infiltrate composed of lymphocytes and a few eosinophils.

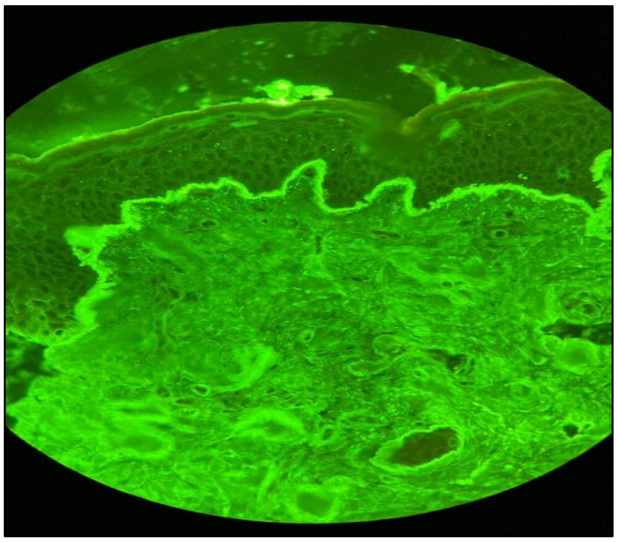
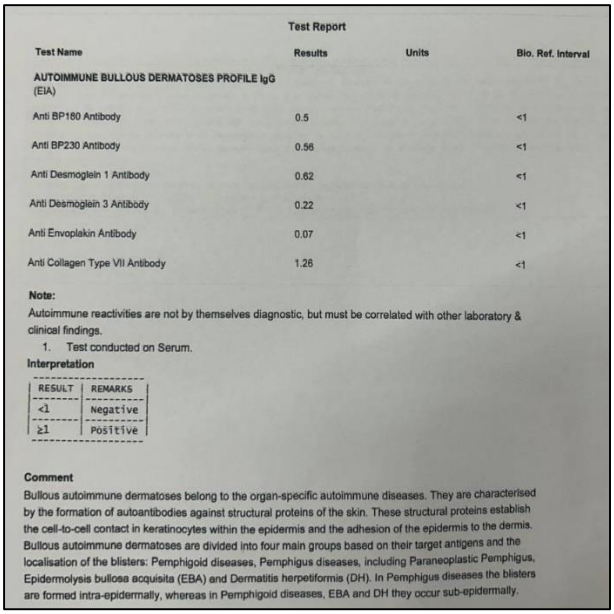


Figure 4: IgG positivity (+3) in a linear pattern, C3c positivity (+3) was also noted in a linear pattern along the DEJ.

The patient was treated with a combination of systemic and topical therapies. She received intravenous Dexamethasone 8 mg for 4 weeks with gradual tapering, Oral Azathioprine 50 mg BD was prescribed for 6 weeks, along with oral levocetirizine 10 mg OD.

Additionally, topical clobetasol propionate 0.05% with fusidic acid was applied over the bullae to manage local inflammation and prevent secondary infection. After

three weeks of treatment, the patient showed significant improvement, with reduced blister formation and healing of erosions.



Test Report

Test Name	Results	Units	Bio. Ref. Interval
AUTOIMMUNE BULLOUS DERMATOSES PROFILE IgG (EIA)			
Anti BP180 Antibody	0.5		<1
Anti BP230 Antibody	0.56		<1
Anti Desmoglein 1 Antibody	0.62		<1
Anti Desmoglein 3 Antibody	0.22		<1
Anti Envoplakin Antibody	0.07		<1
Anti Collagen Type VII Antibody	1.26		<1

Note:
Autoimmune reactivities are not by themselves diagnostic, but must be correlated with other laboratory & clinical findings.
1. Test conducted on Serum.

Interpretation

RESULT	REMARKS
<1	Negative
≥1	Positive

Comment
Bullous autoimmune dermatoses belong to the organ-specific autoimmune diseases. They are characterised by the formation of autoantibodies against structural proteins of the skin. These structural proteins establish the cell-to-cell contact in keratinocytes within the epidermis and the adhesion of the epidermis to the dermis. Bullous autoimmune dermatoses are divided into four main groups based on their target antigens and the localisation of the blisters: Pemphigoid diseases, Pemphigus diseases, including Paraneoplastic Pemphigus, Epidermolysis bullosa acquisita (EBA) and Dermatitis herpetiformis (DH). In Pemphigus diseases the blisters are formed intra-epidermally, whereas in Pemphigoid diseases, EBA and DH they occur sub-epidermally.

Figure 5: Anti-collagen type VII antibodies was positive.

DISCUSSION

Epidermolysis Bullosa Acquisita (EBA) is a rare autoimmune blistering disorder that presents with distinct clinical and histopathological features. However, its clinical presentation, especially in the mechanobullous variant, can overlap with several other blistering diseases, making accurate diagnosis challenging.[3]

The differential diagnosis for EBA involves conditions that share similar blistering patterns, including dystrophic epidermolysis bullosa (DEB) and porphyria cutanea tarda (PCT) in the mechanobullous subtype, as well as other pemphigoid diseases in the inflammatory subtype. our patient, a 53-year-old female, exhibited the hallmark symptoms of EBA, including multiple fluid-filled lesions, erosions, and mucosal involvement, which are typical manifestations of the disease.[4]

In patients presenting with the mechanobullous phenotype, distinguishing EBA from DEB is critical. DEB, unlike EBA, has a positive family history, as it is an inherited disorder, whereas EBA is acquired. Direct immunofluorescence (IF) microscopy can help differentiate the two conditions, as DEB typically shows a different pattern of autoantibody deposition compared to EBA.[5]

Porphyria cutanea tarda (PCT) also shares clinical features such as skin fragility and blistering, but it can be distinguished by the presence of porphyrins in the urine, which is not a feature of EBA. Additionally, the pattern of immunofluorescence would differ between PCT and EBA. In our patient the histopathological features, along with direct immunofluorescence findings of linear IgG and C3c deposition at the dermal-epidermal junction, provided strong evidence to differentiate EBA from other conditions with similar clinical presentations.[6]

The inflammatory variant of EBA poses another diagnostic challenge, as it resembles other pemphigoid diseases such as bullous pemphigoid (BP), mucous membrane pemphigoid (MMP), and linear IgA disease (LAD). BP is one of the most common conditions in the differential diagnosis of EBA, and it can be differentiated by reactivity with BP180 on immunofluorescence testing. MMP and LAD also show similar mucosal involvement and blistering, which can further complicate the diagnosis.[7]

The key to diagnosing EBA lies in identifying antibodies against type VII collagen. Direct immunofluorescence and enzyme immunoassay for anti-collagen type VII antibodies are critical diagnostic tools.[8] A positive result strongly supports the diagnosis of EBA. Both the mechanobullous and inflammatory variants of EBA can present with varying degrees of severity, and in some cases, both forms may coexist in the same patient.

This dynamic nature of the disease can lead to a shifting clinical presentation, making it even more important to regularly reassess the patient's symptoms and course of disease. Moreover, both adult and pediatric populations can be affected, with mucosal involvement seen in approximately 50% of patient.[9] In the present case a positive enzyme immunoassay for anti-collagen type VII antibodies further confirmed the diagnosis of EBA, as these autoantibodies are specific to this condition.[10]

Early recognition of EBA and distinguishing it from other similar conditions is critical for appropriate treatment. Immunosuppressive therapies, including corticosteroids and azathioprine, have shown efficacy in managing EBA, as seen in our case. The patient in this case was treated with a combination of systemic and topical therapies, including intravenous dexamethasone, oral azathioprine, and topical corticosteroids, which resulted in significant improvement within a week.

CONCLUSION

Epidermolysis bullosa acquisita (EBA) remains a challenging diagnosis due to its variable presentation and overlap with other blistering diseases. Early identification through a comprehensive diagnostic approach-incorporating histopathology, direct immunofluorescence, and anti-collagen type VII antibody testing can significantly improve patient outcomes.

The case presented here underscores the importance of considering EBA in patients with subepidermal blistering and mucosal involvement, particularly when other

autoimmune blistering disorders are suspected. Timely intervention with appropriate immunosuppressive therapy can lead to marked clinical improvement and prevent further morbidity. As our understanding of EBA continues to evolve, it is crucial to remain vigilant for this rare but impactful condition, ensuring that patients receive the best possible care to enhance their quality of life.

ACKNOWLEDGEMENTS

Authors would like to thank Dr Sudhir MD Pathology.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Salah E. TEN mimics: Classification and practical approach to toxic epidermal necrolysis-like dermatoses. Indian J Dermatol Venereol Leprol. 2023;89(3):337-46.
2. Kridin K, Kneiber D, Kowalski EH, Valdebran M, Amber KT. Epidermolysis bullosa acquisita: A comprehensive review. Autoimmunity Rev. 2019;18(8):786-95.
3. Das JK, Sengupta S, Gangopadhyay AK. Epidermolysis bullosa acquisita. Indian J Dermatol Venereol Leprol. 2006;72:86.
4. Kumar V, Garg VK. A rare presentation of epidermolysis bullosa acquisita in a 60-year-old male. J Dermatol Clinical Res. 2017;4:672-7.
5. Miyamoto D, Gordilho JO, Santi CG, Porro AM. Epidermolysis bullosa acquisita. Anais Brasileiros de Dermatol. 2022;97(4):409-23.
6. Chhabra S, Minz RW, Saikia B. Immunofluorescence in dermatology. Indian J Dermatol Venereol Leprol. 2012;78:677.
7. Hsi AC, Rosman IS. Histopathology of cutaneous inflammatory disorders in children. Pediatr Develop Pathol. 2018;21(2):115-49.
8. Singh P, Varma S. Immunofluorescence and management of epidermolysis bullosa acquisita: A case study from India. Indian J Dermatol Venereol Leprol. 2014;6:285.
9. Feranandes CZ, Bhat R. Epidermolysis bullosa acquisita-a case report. Indian J Dermatol. 2002;47(2):96-7.
10. Bhattacharjee O, Ayyangar U, Kurbet AS, Ashok D, Raghavan S. Unraveling the ECM-immune cell crosstalk in skin diseases. Frontiers Cell Devel Biol. 2019;7:68.

Cite this article as: Manikanta RSS, Gopal KVT, Raju PVK PV, Sagar NK. Epidermolysis bullosa acquisita in a 53-year-old female with extensive cutaneous and mucosal involvement. Int J Res Dermatol 2025;11:273-6.