e-ISSN 3050-8460
Journal of Dermatology Research

ATHENAEUM SCIENTIFIC PUBLISHERS

Case Report

Successful Treatment of Epidermolysis Bullosa Acquisita with Dupilumab

Katie M Hodges[1], Ahmed M Sikder[1], Edward M Klepper[1*], David Terrano[2], Howard N Robinson[1]

[1]Bernstein and Robinson Dermatology P.A., Bel Air, Maryland, USA

[2]Bethesda Dermatopathology Laboratory, Silver Spring, Maryland, USA

***Correspondence author:** Edward M. Klepper, MS, Bernstein and Robinson Dermatology P.A., Bel Air, Maryland, USA; Email: edwardklepper@gmail.com

Citation: Hodges KM, et al. Successful Treatment of Epidermolysis Bullosa Acquisita with Dupilumab. J Dermatol Res. 2025;6(3):1-4.

https://doi.org/10.46889/JDR.2025.6306

Received Date: 07-10-2025

Accepted Date: 03-11-2025

Published Date: 10-11-2025

Abstract

Epidermolysis Bullosa Acquisita (EBA) is a rare, autoimmune blistering disease which can impact both the skin and mucous membranes. It is not inherited and typically impacts older adults. Not all patients with EBA respond positively to treatment. We are reporting what we believe to be the third case of EBA treated successfully with dupilumab.

Keywords: Bullous; Immunotherapy; Epidermolysis Bullosa Acquisita; EBA; PD-1; PDL-1; CTLA-4; Dupilumab

Introduction

Epidermolysis Bullosa Acquisita (EBA) is a rare, autoimmune blistering disorder that can affect both the skin and mucous membranes and is due to autoantibodies to type VII collagen. Type VII collagen is an integral component of the anchoring fibrils within the dermal-epidermal junction. Damage to the anchoring fibrils results in the characteristic epidermal detachment. The blisters are aggravated due to friction [1]. EBA lacks a cure and treatment is tailored towards supportive and symptomatic measures. Treatments include wound care, avoidance of friction,and topical/oral steroids [1]. Here we present a case of drug-induced EBA successfully treated with dupilumab. In this case, EBA was most likely triggered by ipilimumab (Yervoy), a CTLA-4 inhibitor, for treatment of esophageal cancer.

Case Report

We describe a 75-year-old established male patient who presented with a pruritic rash on his trunk and extremities that started approximately one year after he began dual immunotherapy with nivolumab every 2 weeks and ipilimumab every six weeks for esophageal cancer. The patient was prescribed topical triamcinolone 0.1% ointment twice a day for two weeks. The patient was also recommended to moisturize with over-the-counter Eucerin Eczema Relief Cream, CeraVe Itch Relief Cream with Pramoxine HCl and Sarna Lotion with Menthol to manage symptoms pending biopsy results.

The initial biopsies for direct Immunofluorescence (DIF) were positive for Bullous Pemphigoid (BP). Treatment was initiated with oral doxycycline 100 mg twice a day for three weeks, then tapered down to once a day for one month. The patient reported improvement over the course of the doxycycline regimen. Shortly after his final ipilimumab infusion, the patient presented on 5/23/24 with a bullous pemphigoid exacerbation. Retrospectively, the flares appeared to correlate with the ipilimumab infusions in particular. The patient was prescribed topical clobetasol 0.05% ointment once a day, along with over-the-counter moisturizers and oral fexofenadine 180 mg a day. The symptoms did not improve for two months, therefore the patient was initiated on subcutaneous dupilumab. A loading dose of two 300 mg/2 ml subcutaneous injections were administered and then a 300 mg/2 ml subcutaneous injection was given every 2 weeks. The patient was being followed once a month and exhibited gradual and significant improvement up until 10/29/24, when his flares were completely resolved. The patient was instructed to discontinue the dupilumab and just maintain the topical clobetasol twice a day for two weeks, then twice a week as needed for any new areas.

Three months later, the patient presented with another exacerbation which appeared after he started taking daily intravenous long-acting morphine (Fig. 1). The topical clobetasol did not provide relief for the patient, but he was able to manage symptoms somewhat by moisturizing. Skin biopsies were sent for Hemotoxin and Eosin (H&E) staining and outside DIF consultation. The H&E showed eosinophil interface dermatitis, consistent with a subepidermal bullous dermatosis (Fig. 2). DIF studies were once again positive - this time with Epidermolysis Bullosa Acquisita (EBA) in the differential with bullous pemphigoid. Subsequent salt-split results, showing IgG and C3 only on the dermal side of the blister confirming diagnosis of EBA. Following diagnosis, the patient was restarted on subcutaneous dupilumab 300 mg/2 ml subcutaneous injections every 2 weeks. The patient was seen for a follow-up 3/7/25, at which point he had received two dupilumab injections. At this visit, he was completely clear of EBA lesions and was switched to once monthly dupilumab 300 mg/2 ml subcutaneous injections for maintenance (Fig. 3).

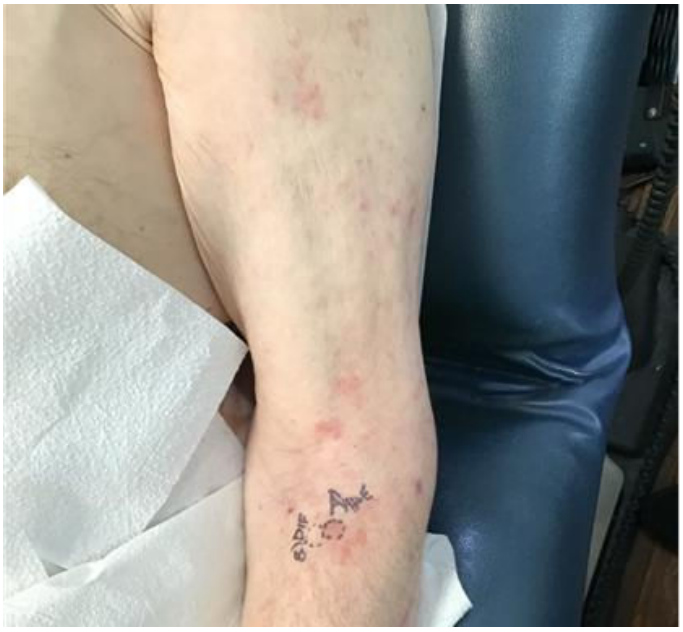

Figure 1: Patient presented with erythematous papules, macules and patches on the trunk and extremities.

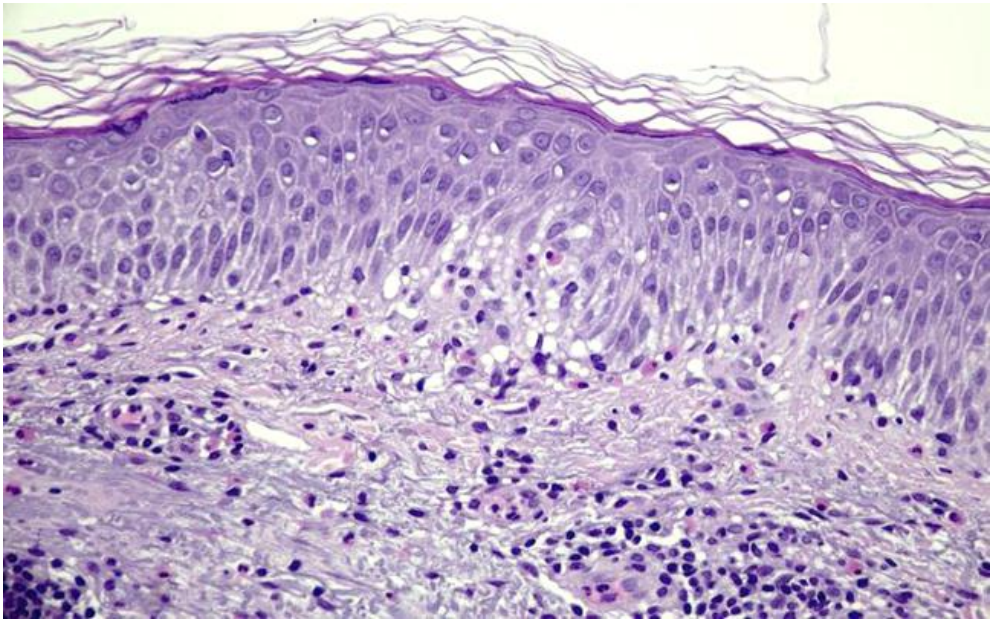

Figure 2: 400x magnification of histologic sections demonstrating the eosinophilic inflammation at the dermo-epidermal junction.

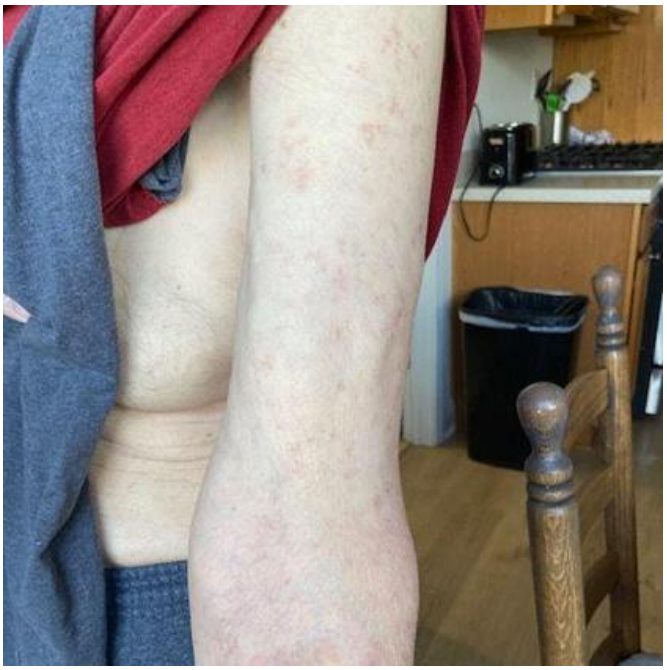

Figure 3: After 2 weeks on dupilumab, the patient had faded erythematous macules and patches on the trunk and extremities with a reduction in pruritus.

Discussion

Currently it has been well-established that immunotherapies such as PDL1, PD1 and CTLA-4 inhibitors have been responsible for triggering bullous pemphigoid [2]. In our particular patient, after exacerbation of his condition when off dupilumab, repeat biopsies did confirm that the patient had EBA. In addition, there was an eosinophilic infiltrate.

As of July 2025 dupilumab has been approved for the treatment of bullous pemphigoid. Dupilumab targets type 2 inflammatory mediators, interleukin-4 and interleukin-13, which recruit eosinophils and IgE [3]. The conditions that dupilumab are indicated for involve type 2 inflammatory mediators. In EBA, the immune response is regulated by both Th1 and Th2 cells [4]. Although the mechanism of action is not completely clear, one could assume that dupilumab's reduction of type 2 inflammatory mediators contributes to the improvement in EBA lesions. Of note, there is a bullous pemphigoid-like variant of EBA which does seem to fit this clinical picture [5,6]. The bullous pemphigoid-like EBA variant is characterized by eosinophils, monocytes and lymphocytes within the dermis [6].

Epidermolysis bullosa acquisita typically presents classically as bullae and erosions at the site of friction or injury. The BP-variant of EBA can look similar to the other inflammatory EBA variants such as mucous membrane pemphigoid-like EBA, linear IgA bullous dermatoses-like EBA and Brunsting-Perry cicatricial pemphigoid-like EBA [1].

Two case reports have been published thus far demonstrating the efficacious treatment of EBA with dupilumab.The first is of a 51-year-old female with a five year history of EBA confirmed by DIF. The patient failed treatment with oral prednisolone, oral azathioprine, intravenous rituximab infusions and topical steroids. Dupilumab injections in conjunction with short-term topical mometasone furoate resulted in significant improvement within 3 months [4]. The second case is of a 37-year-old female who exhibited 44 days of symptoms, after which diagnosis of EBA was established by DIF. The patient failed treatment with oral methylprednisolone and mycophenolate. Dupilumab injections resulted in clearance within 10 weeks, with a small relapse able to be controlled with oral prednisone and low-dose dapsone [7].

Conclusion

This is an interesting case of dupilumab being used in immunobullous disorders. It was effective for bullous pemphigoid and now EBA. There is a variant of bullous pemphigoid-like EBA which would potentially explain dupilumab's efficacy in this case. Further studies are needed to determine the effectiveness of dupilumab for other types of EBA. Other studies are needed to determine the most efficacious frequency, whether it is every other week, once a month or every two to three months.

Conflicts of Interest
The authors declare no conflict of interest in this paper.

Funding
None

Authors' Contributions
All authors contributed to conceptualization, treatment execution, manuscript writing and final approval.

References
1. Xiao A, Sathe NC, Tsuchiya A. Epidermolysis bullosa acquisita. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing. 2025. [Last accessed on: November 03, 2025] https://www.ncbi.nlm.nih.gov/books/NBK554512/
2. Nikolaou V, Tsimpidakis A, Stratigos A. Cutaneous adverse reactions of immunotherapy in patients with advanced melanoma. Cancers (Basel). 2023;15(7):2084.
3. US Food and Drug Administration. DUPIXENT (dupilumab) injection, for subcutaneous use: US prescribing information. Silver Spring (MD): US FDA. 2017. [Last accessed on: November 03, 2025] https://www.accessdata.fda.gov/drugsatfda_docs/label/2024/761055s064lbl.pdf
4. Diehl R, Kiritsi D, Schauer F. Treatment of epidermolysis bullosa acquisita with dupilumab. J Dtsch Dermatol Ges. 2024;22(10):1417-9.
5. Kasperkiewicz M, Sadik CD, Bieber K, Ibrahim SM, Manz RA, Schmidt E, et al. Epidermolysis bullosa acquisita: From pathophysiology to novel therapeutic options. J Invest Dermatol. 2016;136(1):24-33.
6. Kridin K. Accessible diagnostic methods to differentiate between epidermolysis bullosa acquisita and other subepidermal autoimmune bullous diseases. Indian J Dermatol. 2018;63(5):445-8.
7. Xing X, Liu Y, Chen L, Dan Y, Ren J. Treatment of refractory, inflammatory epidermolysis bullosa acquisita with a combination of dupilumab and systemic glucocorticoid. J Dermatol. 2025;52(4):e308-10.