

Intro/Background

- Epidermolysis bullosa pruriginosa (EBP) is a rare clinical variant of epidermolysis bullosa characterized by intensely pruritic nodules and plaques located on the extremities.
- While a majority of EBP cases have been described as a subtype of dystrophic epidermolysis bullosa, EBP as a subtype of epidermolysis bullosa simplex has been described.
- Current therapeutic options for EBP have primarily aimed at reducing pruritus, with variable success.
- Six cases of dystrophic EBP treated with dupilumab have previously been reported.¹⁻⁵
- However, simplex variant EBP treated with dupilumab has not yet been documented. We present a novel case of simplex variant EBP demonstrating rapid improvement with dupilumab.

Case History and Treatment Course

A 48-year-old Caucasian man presented with pruritic lesions on the bilateral lower extremities and abdomen. The lesions had been present since childhood, interfered with sleep, and did not improve with topical therapies. On the bilateral lower extremities were numerous coalescing vesicles and bullae with overlying hemorrhagic crust. There were also coalescing pink papulonodules, several with surrounding collarettes of scale (Figure 1). A biopsy taken from a papulonodule on the right thigh demonstrated noninflammatory subepidermal fissuring with a perivascular infiltrate of lymphocytes and eosinophils. High molecular weight keratin staining demonstrated labeling at the base of the blister, indicative of splitting within basal keratinocytes characteristic of simplex variant EB (Figure 2A and 2B).

Within 3 months of therapy with dupilumab, he reported improvement of pruritus and no development of new blisters or erosions (Figure 3). He continues to have near-complete relief of itching and has not developed new lesions at 3-month follow-up.



Figure 1. Initial presentation of epidermolysis bullosa pruriginosa. Numerous pink lichenified nodules and plaques with scattered bullae, vesicles and erosions.

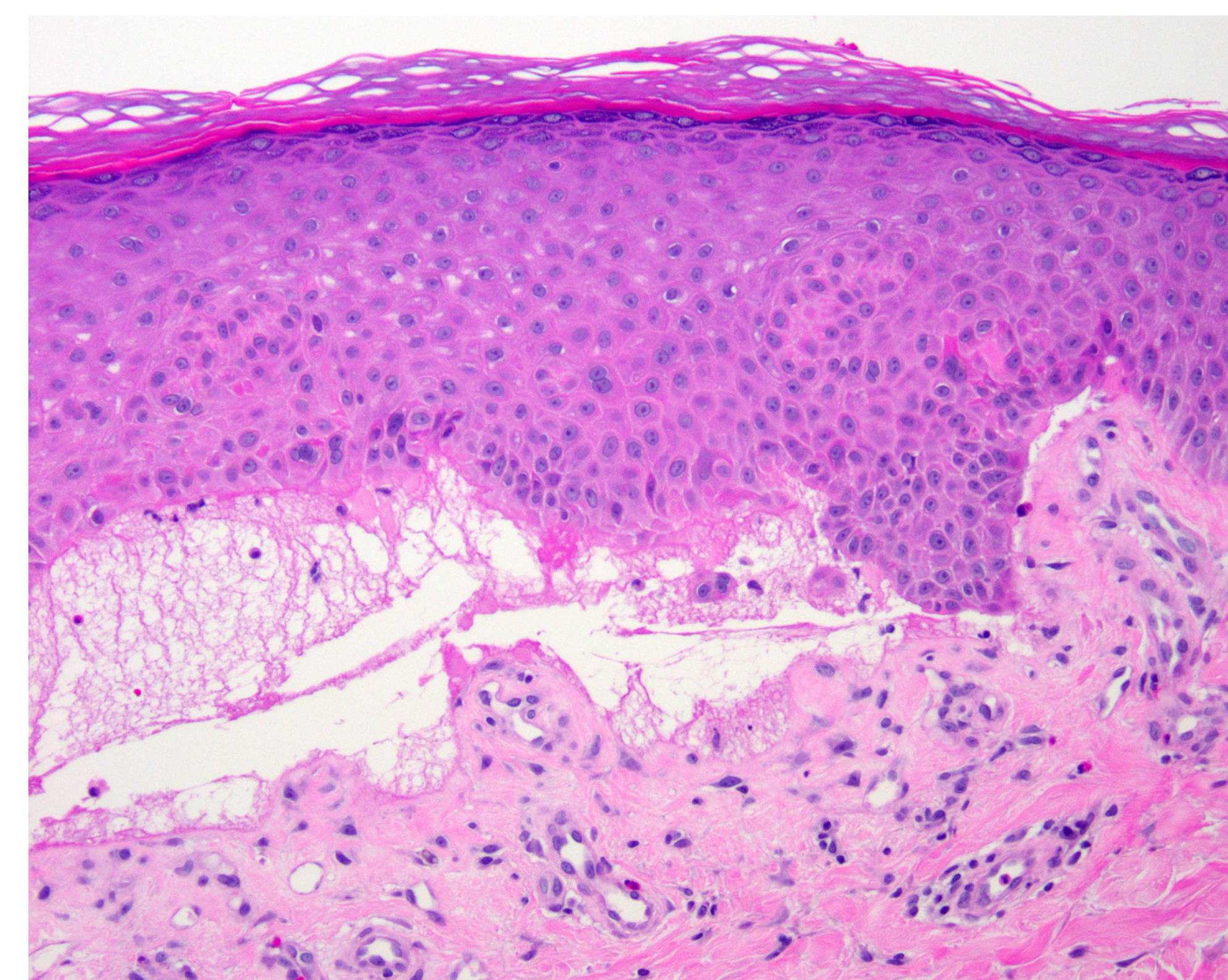


Figure 2A. Histopathologic examination showing noninflammatory subepidermal fissuring with a perivascular infiltrate of lymphocytes and eosinophils (hematoxylin & eosin, 100x).

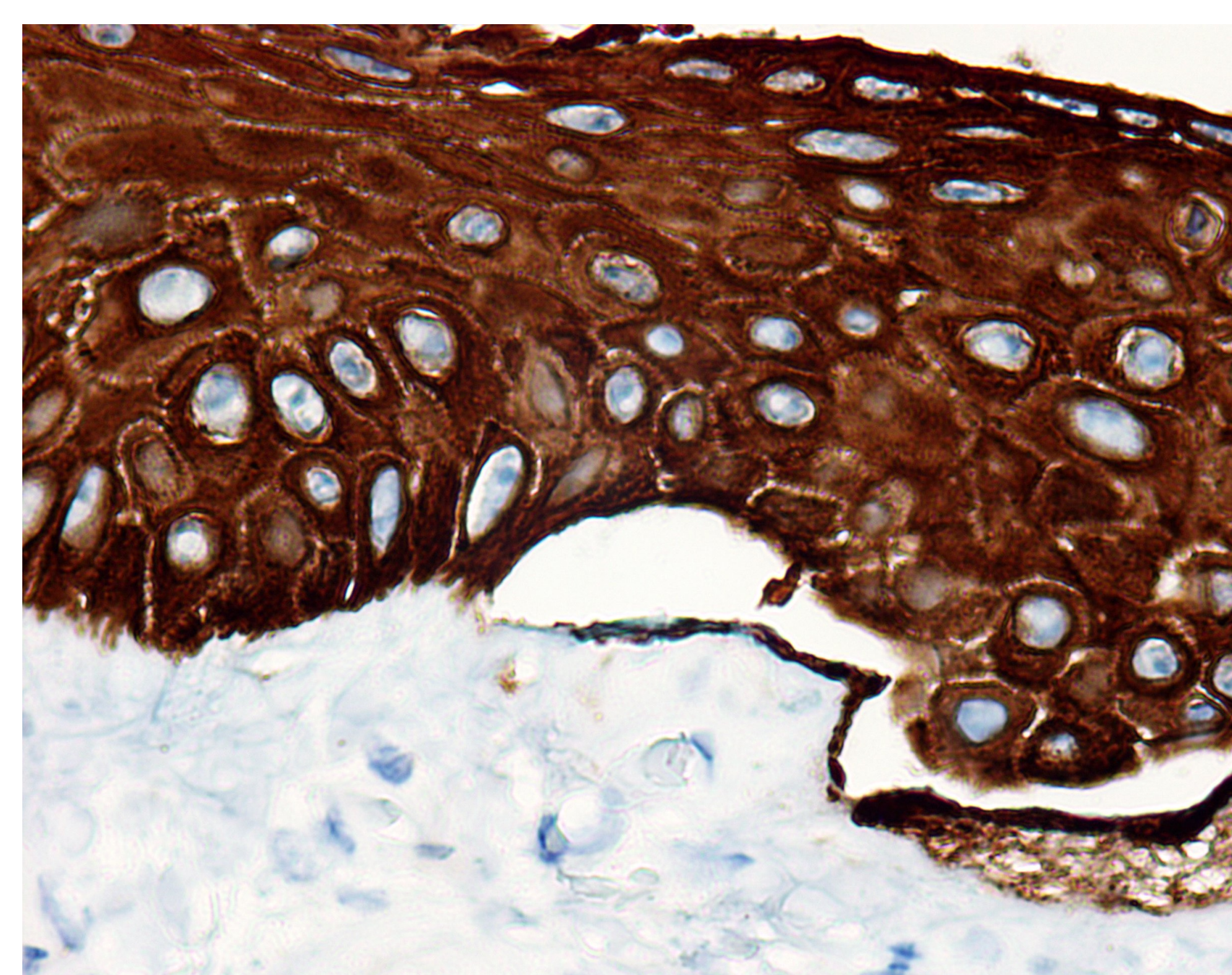


Figure 2B. Immunohistochemical analysis highlighting high molecular weight keratin at the base of the blister (cytokeratin 34 beta E12, 400x).

Discussion

Although the mechanism of dupilumab's efficacy in EBP is unknown, it has been proposed that by binding to the alpha subunit of the IL-4 receptor, it blocks sensitization of sensory neurons to pruritogens.⁶ This may contribute to breaking of the itch-scratch cycle, leading to flattening of pruritic papules, and allowing for skin restoration.



Figure 3. Improved lichenified nodules and plaques. No new vesicles or bullae.

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Disclosures: The authors state no relevant conflicts of interest.