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Successful Treatment of Refractory Epidermolysis Bullosa Pruriginosa with Dupilumab Injections

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A 39 year old male presented to dermatology evaluation of numerous pruritic pink papulonodular lesions on bilateral upper and lower extremities that had been present since childhood but were becoming more numerous with age. His father, son, paternal cousin, and paternal grandmother were afflicted with similar lesions. A biopsy yielded orthokeratotic hyperkeratosis, hypergranulosis, and acanthosis without atypia within the epidermal layer. The upper dermis showed fibrosis and perivascular mononuclear inflammatory infiltrate. There was focal separation visible at the dermo-epidermal junction (DEJ), suggesting epidermolysis bullosa pruriginosa (EBP) as a probable diagnosis. The patient's symptoms failed to respond to numerous attempted treatments, including clobetasol ointment, pimecrolimus cream, and oral dapsone. Finally, the patient was offered dupilumab, a monoclonal antibody that targets the alpha subunit of the IL-4 receptor (IL-4Rα), as an alternative treatment due to reported success treating prurigo nodularis.[1–4]. Four weeks after his first dupilumab injections, the patient reported near-complete resolution of his pruritis and notable reduction in lesion number. The patient continues to receive biweekly dupilumab injections and has maintained improvement of his symptoms without side effects. There has been only two reported cases of successful treatment of EBP with dupilumab, and the addition of our case highlights the need for further study of the efficacy of dupilumab for this rare condition, particularly for patients refractory to more conservative management. More broadly, this case supports the hypothesis that blockade of IL-4 may provide treatment for a wide variety of chronic pruritic conditions.

References:

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