Case of Bullous Pemphigoid Masquerading as Adult-Onset Atopic Dermatitis

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Abstract

The majority of dermatitis cases in adults result from chronic or relapsing atopic dermatitis in childhood. Adult-onset atopic dermatitis, also known as idiopathic chronic eczematous eruption of aging (CEEA), is a phenomenon seen in adults 50 years and older with no prior history of atopic dermatitis. CEEA is often a diagnosis of exclusion after ruling out more serious causes of dermatitis including bullous pemphigoid (BP), allergic conditions, and hematologic malignancies. This report details the case of a 67-year-old woman with no history of atopy who presented with a persistent, eczematous dermatitis not responsive to traditional therapy, consistent with CEEA, but was later identified as BP.

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Author Contributions
Mai Nojima, MD, and Iesha L Ticknor, MD, prepared manuscript draft and participated in patient care. Payam Sazegar, MD, was attending physician and provided conceptualization and review/editing of manuscript.

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Relevancy Statement
This case report contributed to practice change by highlighting the importance of considering serious conditions like bullous pemphigoid in the differential diagnosis for patients with treatment-resistant dermatitis.

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Introduction

The majority of dermatitis cases in adults result from chronic or relapsing atopic dermatitis in childhood.[1] However, CEEA is a phenomenon seen in adults over 50 years old with no prior history of atopic dermatitis.[2] Here we describe the case of a 67-year-old woman with no atopic history presenting with a pruritic dermatitis. The initial presentation was clinically consistent with CEEA resistant to standard treatment but subsequent skin biopsy and serological testing revealed a diagnosis of BP, a more serious diagnosis with an estimated 1-year mortality rate over 20%.[3] The patient provided informed consent for this case report on October 9, 2023.

Case Patient Presentation

A 67-year-old woman with hypertension (on hydrochlorothiazide) and hyperlipidemia (on atorvastatin) presented to her primary care clinic with a 3-month history of pruritic, non-bullous rash on her back that gradually progressed over 1 month to cover her extremities. She reported being too embarrassed to go out in public due to the appearance of her skin. She also reported no known triggers such as new foods, shampoos, medications, supplements, or laundry detergent. There were no close contacts with a similar dermatitis. She has no history of eczema or drug allergies. Examination revealed excoriated, erythematous papules and plaques with a yellow-brown scale on the

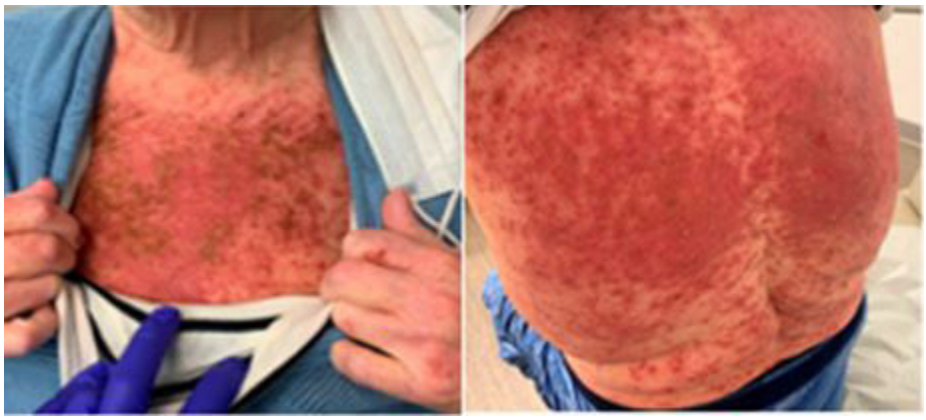

Figure: Images taken at initial visit. Papular errythematous rash with overlaying yellow crusting on the anterior chest (Left) and posterior torso (Right).

chest, back, bilateral forearm, buttocks, and proximal legs. (Figure)There was no mucosal involvement. Skin culture of the yellow-brown scale grew methicillin-susceptible *Staphylococcus aureus*. Her clinical presentation was initially diagnosed as nonspecific allergic-contact dermatitis with secondary impetigo. The patient was started on cephalexin for 7 days and received an intramuscular injection of triamcinolone 60 mg as well as a topical ointment of triamcinolone 0.1% . She presented to the dermatology clinic 3 weeks later with persistent erythematous papules and pruritis. A scabies prep test was negative. The patient was treated with oral prednisone (5-day course), hydroxyzine, doxycycline, and topical clobetasol 0.05 % ointment and her symptoms showed improvement over 1 month but then recurred. Topical clobetasol and oral hydroxyzine provided symptomatic relief.

The patient saw the dermatology clinic again 2 months later and had bloodwork and a shave biopsy performed. The blood serologies were positive for basement membrane zone antibody with a high titer (1:160) and positive direct immunofluorescence assay. Serum anti-BP180 autoantibodies were negative but serum anti-BP230 autoantibodies were positive. Shave biopsy of a perilesional site (buttocks) was performed, taking care to shave to deep dermis, and this showed subepidermal vasculopathy with eosinophils. These findings were consistent with a diagnosis of BP, even though the patient never presented clinically with bullae.

Differential Diagnosis

Older adults with new-onset dermatitis have high rates of diagnosis change after skin biopsy or patch testing procedures.[4] This patient's clinical presentation was concerning for drug hypersensitivity syndrome or toxic epidermal necrolysis, but her lack of new medications ruled out drug reactions. Allergic contact dermatitis and cutaneous T-cell lymphoma were also considered as part of the differential for refractory dermatitis. The patient's age, lack of childhood dermatitis, and clinical features of eczematous eruptions that excluded her face was consistent with CEEA, which is more common in patients treated with thiazides or calcium channel blockers.[5] Due to the persistent nature of the dermatitis, serologies and biopsies were ordered to rule out autoimmune bullous disorders and were consistent with BP despite her clinical presentation not being classic BP.

Clinical Reasoning

Even though 1 in 8 patients with BP do not develop bullae, 100% of these patients complain of pruritis.[6] Mucosal involvement may only be present in 10–30% of cases.[7] In rare cases, the only clinical presentation may be pruritis and a higher index of suspicion is required to consider work-up of a pruritic dermatitis in older adults.[7] The fluctuating course of this patient's pruritis and dermatitis may have been attributable to rebound flares related to systemic steroids which generally should only have short courses and serve as a bridge to steroid-sparing therapies.[8] Direct immunofluorescence was positive in this case and this has a 91% sensitivity, which is the highest among the serological tests.[3] Additionally, drug-induced BP is something to consider, as hydrochlorothiazide has been reported as a

potential inducing agent.[9] In this case, a subsequent punch biopsy was not performed as the tangential skin biopsy was deemed to have adequately sampled the deep dermis and this procedure would not be expected to change the diagnosis or clinical management.

Treatment Pathway

The skin biopsy demonstrating epidermolysis helped confirm the diagnosis, as immune deposits in BP are found at either the epidermal site or mixed at both the epidermal and dermal sites of the skin.[6] There is high-quality evidence supporting the use of doxycycline to control BP in mild to moderate cases of the disease.[3] In more severe cases, immunomodulatory treatments such as dupilumab can be considered.[10]

Outcomes

The patient's dermatitis waxed and waned over 2 months but was eventually controlled with oral doxycycline 100 mg twice daily. Over the next 9 months her dermatitis was maintained in remission and only required occasional short-term use of topical clobetasol 0.05% for pruritis.

Key Learning Takeaways

This case report contributes to practice change by underscoring the value of a skin biopsy in cases of treatment-resistant dermatitis with clinical uncertainty. The patient's presentation also demonstrated the need to consider the potentially serious diagnosis of BP in an older adult with either treatment-refractory dermatitis or unexplained pruritis.

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