Erythrodermic Epidermolysis Bullosa Acquisita

Epidermolysis bullosa acquisita (EBA) is a rare acquired sub-epidermal blistering autoimmune disease of the skin and mucosae associated with autoantibodies directed against type VII collagen, the major component of the anchoring fibrils of the dermal-epidermal junction. Various clinical presentations of EBA have been described, including a noninflammatory mechnobullous form, an inflammatory bullous pemphigoid (BP)-like form, and a mucous-membrane pemphigoid-like form. These forms may show clinical overlap, and their courses are often unpredictable.1,3

Report of a Case | A 60-year-old man was admitted for evaluation of a 3-week history of widespread pruritic cutaneous lesions. The patient had taken no drugs, and his medical history was unre-
markable. On examination, almost his entire trunk, buttocks, and thighs were erythematous and infiltrated with papular and urticarial lesions (Figure 1). Islands of normal-appearing skin were observed. On his soles and palms, isolated lesions with a target-like appearance were noted. The head as well as the oral and genital mucous membranes were spared. During hospital admission, the patient developed isolated vesicles and serous blisters on erythematous skin on his wrist and ankle.

Light microscopy studies of a skin biopsy specimen obtained from patient’s back showed a diffuse spongiosis with a mixed perivascular inflammatory infiltrate consisting of eosinophils and neutrophils in the upper dermis (Figure 2). Direct immunofluorescence microscopy studies disclosed linear deposits of IgG and C3 along the epidermal basement membrane zone. By indirect immunofluorescence microscopy using sodium chloride-separated normal human skin, circulating IgG autoantibodies binding the dermal side of the split were detected.

Immunoblotting analysis using human dermal extracts showed a reactivity with a 290-kDa protein showing the same electrophoretic migration to the protein band recognized by the control monoclonal antibody directed against type VII collagen.

Based on the clinical features and immunopathological findings, the diagnosis of EBA was made. The patient was given oral prednisolone, 0.75 mg/kg of body weight, which resulted in rapid clearance of the lesions within 2 weeks. Corticosteroid doses were subsequently slowly tapered. At a dose of 10 mg/d, the patient experienced a relapse and began treatment with methotrexate, 15 mg subcutaneously once weekly. The prednisolone dose was then tapered to 2.5 mg/d, and the patient remained asymptomatic at 6-month follow-up.

Discussion | The clinical features of EBA are protean. The classic presentation is that of a noninflammatory mechanobullous disease characterized by the development of acral blisters that heal with atrophic scarring, milia, and hyperpigmentation or hypopigmentation. They are localized to trauma-prone surfaces such as elbows, knees, hands, and feet. Acral involvement may be mutilating. Scalp involvement occurs in up to 20% of patients. The inflammatory BP-like presentation is associated with widespread vesicles and bullae involving intertriginous and flexural areas that heal without atrophic scarring. Epidermolysis bullosa acquisita may also present as mucous membrane pemphigoid or as Brunsting-Perry pemphigoid phenotype. The potential causes of erythroderma include psoriasis, atopic dermatitis, drug reactions, and cutaneous T-cell lymphoma. With the exception of pemphigus foliaceus, the other autoimmune bullous diseases of the skin have been only anecdotally implicated as cause of erythroderma. Specifically, single cases of erythrodermic BP have been described. Epidermolysis bullosa acquisita is potentially associated with a number of systemic diseases, including inflammatory bowel diseases, but our patient showed no evidence of any of these.
Our case was striking because the patient initially showed features suggestive of either a severe drug reaction or a paraviral eruption, but immunopathological studies were diagnostic for EBA. Our observation provides a further example about the polymorphous and misleading presentations of EBA. Hence, EBA should be considered as a rare cause of erythroderma.

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Conflict of Interest Disclosures: None reported.

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CORRECTION

Missing Middle Initial in Author’s Name: In the Observation letter entitled “Acute Onset of Acrokeratosis Paraneoplastica (Bazex Syndrome)” published in the March 11, 2015, online first issue of JAMA Dermatology (doi:10.1001/jamadermatol.2014.5622), an author’s middle initial was not included in the byline. The author’s full name is Amara S. Hussain. This article was corrected online.

Incorrect Figure: In the article titled “Intralesional Rituximab in the Treatment of Refractory Oral Pemphigus Vulgaris,” published online December 23, 2014, in JAMA Dermatology (doi:10.1001/jamadermatol.2014.3674), incorrect images appeared in a figure. As a result of human error, Figure 2A was inadvertently published upside down. Both images in Figure 2 are now replaced. In addition, the figure caption should have read as follows: “Clinical photographs of the right buccal mucosa in patient 3 show progress of oral PV with rituximab treatment. A, Pretreatment photograph showing persistent oral erosions. B, Posttreatment photograph at week 16 showing complete healing.” This article was corrected online.

Error in Byline: In the article “Narrowband UV-B Phototherapy for Steroid-Refractory Sclerotic Chronic Cutaneous Graft-vs-Host Disease,” published online March 25, 2015 (doi:10.1001/jamadermatol.2015.0175), the name of the second author listed in the byline has been corrected from Rachel Holtzman, BS, to Rachel McAndrew, BS. The author contributions have been corrected to reflect this change and now read as follows: “Author Contributions: Mr Sorenson and Dr Levin had full access to all of the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Sorenson, Logan, Koo, Levin. Acquisition, analysis, or interpretation of data: All authors. Drafting of the manuscript: Sorenson, McAndrew, Patel, Logan, Koo. Critical revision of the manuscript for important intellectual content: All authors. Study supervision: Sorenson, Logan, Koo, Levin.”

Error in Text: In the Research Letter by Chen et al titled “γ-Secretase Mutation in an African American Family With Hidradenitis Suppurativa,” published online February 18, 2015, in JAMA Dermatology (doi:10.1001/jamadermatol.2014.5306), an incorrect number of families was given in the second sentence of the first paragraph. The sentence should read as follows: “Heterozygous mutations in the γ-secretase genes have been identified in the pathogenesis of autosomal dominant forms of HS in 2 British, 11 Chinese, 1 Japanese, and 3 French families.” This article was corrected online.