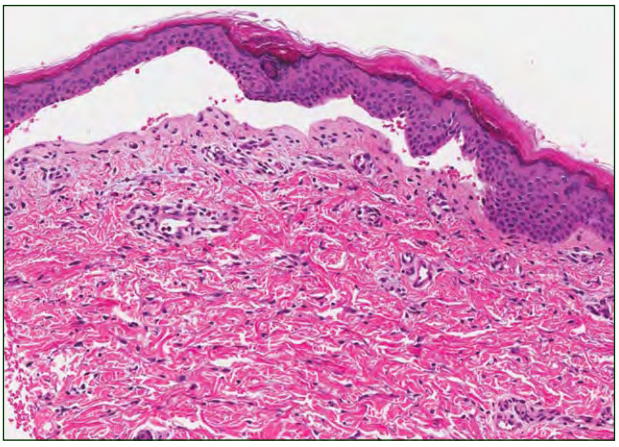
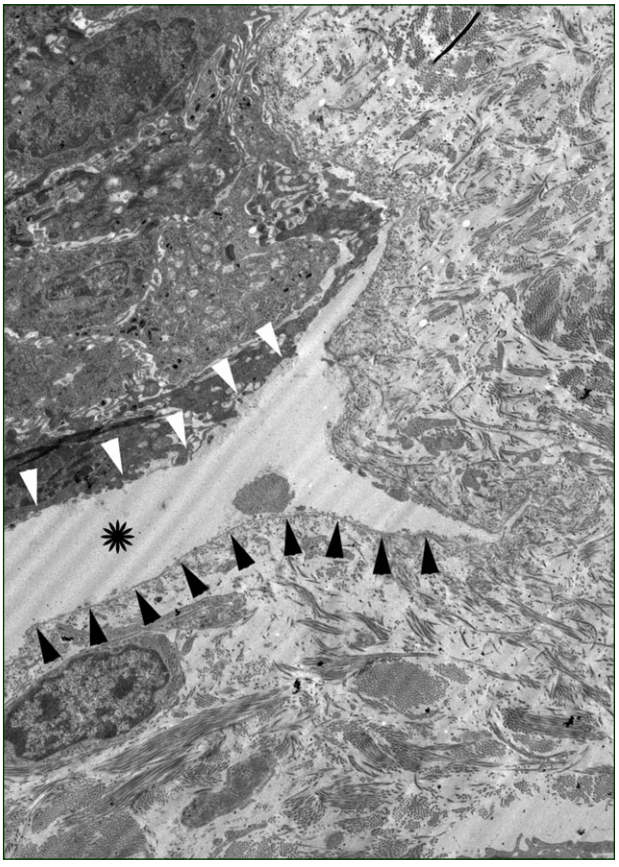


H&E, original magnification ×10.



Electron microscopy, original magnification ×4400.

The best diagnosis is:

a. bullous pemphigoid
b. junctional epidermolysis bullosa (JEB)
c. pemphigus vulgaris
d. staphylococcal scalded skin syndrome
e. toxic epidermal necrolysis

PLEASE TURN TO PAGE 81 FOR DERMATOPATHOLOGY DIAGNOSIS DISCUSSION

Mouhammad Aouthmany, MD, MHSA; Emily Keller, MD; Melissa Piliang, MD

Dr. Aouthmany is from the Department of Dermatology, Indiana University, Indianapolis. Drs. Keller and Piliang are from the Department of Dermatology, Dermatology and Plastic Surgery Institute, Cleveland Clinic, Ohio. Dr. Piliang also is from the Department of Anatomic Pathology, Cleveland Clinic.
The authors report no conflict of interest.
Correspondence: Mouhammad Aouthmany, MD, MHSA, 700 N Alabama St, Indianapolis, IN 46204 (mo@umich.edu).

Junctional Epidermolysis Bullosa

Junctional epidermolysis bullosa (JEB) is an autosomal-recessive mechanobullous skin disorder that clinically presents as tense blisters on the extremities and trunk with variable involvement of the mucosa and nails. Junctional epidermolysis bullosa can be divided into 3 major subtypes: Herlitz JEB, non-Herlitz JEB, and JEB with pyloric atresia. The subtypes are characterized by the formation of bullae within the lamina lucida. The Herlitz type is characterized by reduced or absent laminin-332, formerly laminin-5, while the non-Herlitz type is caused by a missense mutation, which leads to a reduction of functional laminin-332. Some cases of non-Herlitz JEB are due to complete absence of type XVII collagen. The third subtype, JEB with pyloric atresia, is caused by the genetic mutation that code $\alpha_6\beta_4$ integrin subunits, the main receptor for the ligand laminin-332 beneath the hemidesmosomes. Histology of JEB is remarkable for subepidermal separation with bullae, which usually are without substantial inflammatory infiltration (Figure 1). Electron microscopy reveals disruption in the basement membrane zone with separation through the lamina lucida (Figure 2).[1-3]

Bullous pemphigoid (BP) also has subepidermal separation with bullae. However, it is differentiated from JEB with the predominance of eosinophils in the dermis and blister cavity (Figure 3).[2,4,5] Additionally, BP is an immunobullous skin disorder that can be differentiated from JEB by immunofluorescence. Although negative in JEB, linear IgG to BP antigen 180 (BP180, BP antigen 2, or type XVII collagen)

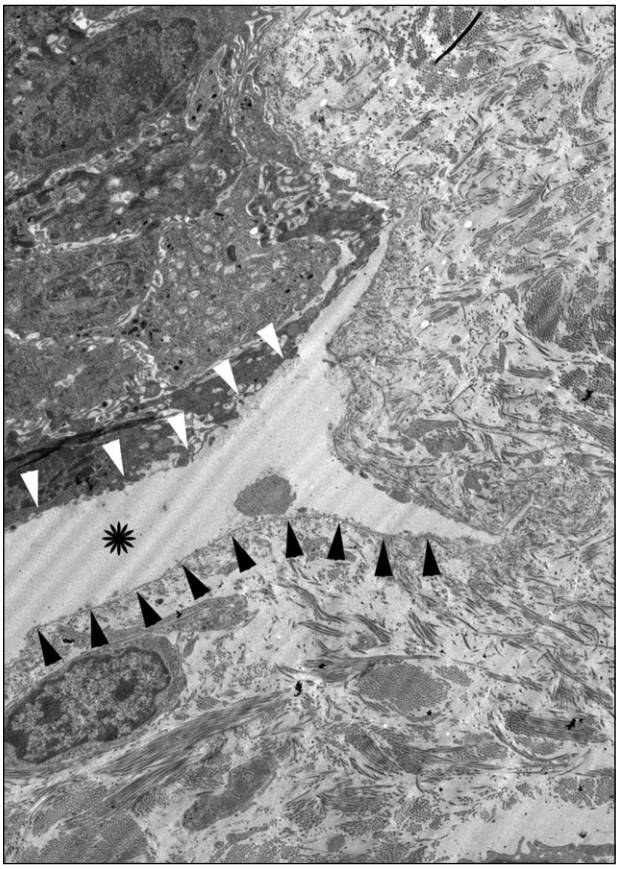


Figure 2. Electron microscopy of junctional epidermolysis bullosa shows a blister (asterisk) within the lamina lucida between the lamina densa (black arrowheads) and the basal keratinocyte (white arrowheads)(original magnification ×4400).

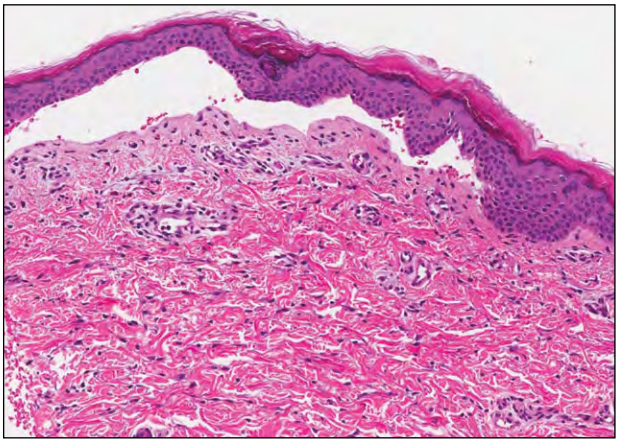


Figure 1. Subepidermal separation with bullae of junctional epidermolysis bullosa (H&E, original magnification ×10).

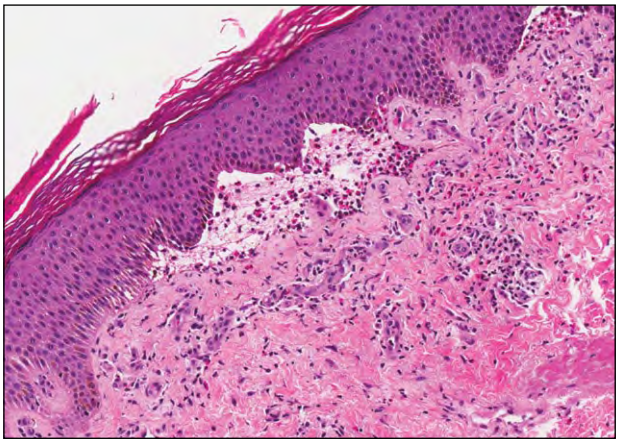


Figure 3. Subepidermal separation with eosinophils in the dermis and blister cavity of bullous pemphigoid (H&E, original magnification ×10).

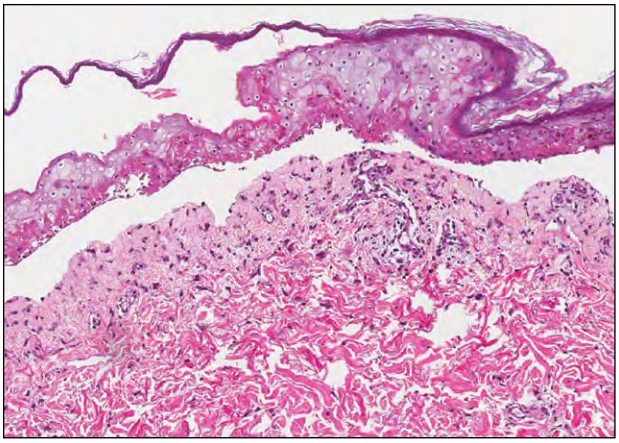


Figure 4. Necrotic epidermis with a subepidermal bulla in toxic epidermal necrolysis (H&E, original magnification ×10).

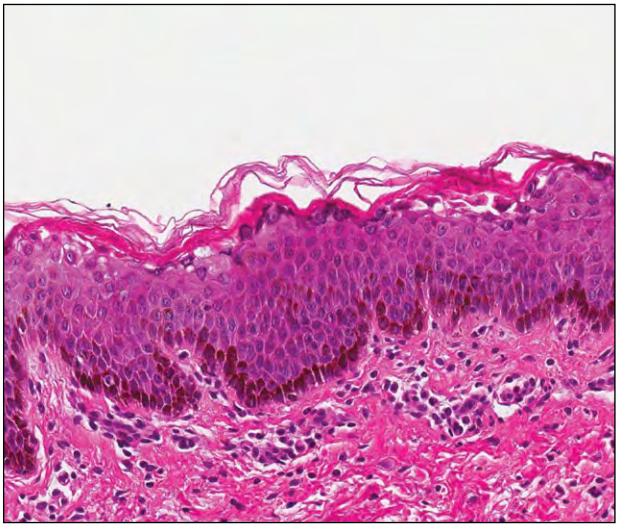


Figure 5. Subcorneal split of the epidermis of staphylococcal scalded skin syndrome (H&E, original magnification ×10).

and/or BP antigen 230 (BP230 or BP antigen 1) are seen in BP.[6] In toxic epidermal necrolysis there is a subepidermal bulla with necrosis of the epidermis and a sparse lymphocytic infiltrate (Figure 4).[3] Staphylococcal scalded skin syndrome is characterized by a subcorneal split of the epidermis with sparse acantholytic cells and neutrophils (Figure 5). Additionally, a mixed inflammatory infiltrate may be observed in the dermis.[3] Pemphigus vulgaris displays suprabasilar bullae with acantholysis that extends down the adnexa. The characteristic tombstone appearance of the basal cells is caused by the loss of intercellular bridges (Figure 6).[3,4] Similar to BP, pemphigus vulgaris also

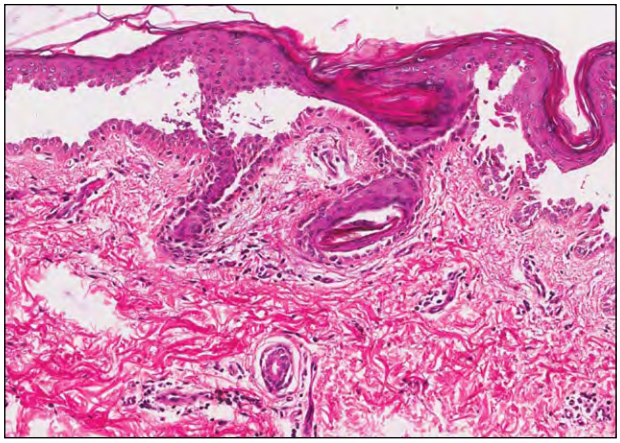


Figure 6. Suprabasilar bullae with acantholysis in pemphigus vulgaris and tombstone appearance of the basal cells with adnexal extension of the acantholysis (H&E, original magnification ×10).

is an immunobullous disorder that can be differentiated from JEB by the intercellular netlike IgG on immunofluorescence to desmogleins (desmoglein-3 and/or desmoglein-1).[7]

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