JOINT DECISIONS

Dx: Epidermolysis Bullosa Acquisita

The woman's bullous lesions previously improved with dapsone 50 mg daily, but treatment was discontinued after she developed a hypersensitivity syndrome. She then was prescribed azathioprine 75 mg daily, hydroxychloroquine 200 mg twice daily, and systemic corticosteroids in varying strengths. At time of presentation, she was taking 15 mg prednisone.

"Differential diagnoses included bullous systemic lupus erythematosus (BSLE), bullous pemphigoid, and epidermolysis bullosa acquisita," Dr. Alia Sampson Brown said at the Caribbean Dermatology Symposium.

Physicians took punch biopsies to perform hematoxylin and eosin staining and direct immunofluorescence, as well as serum for an indirect immunofluorescence assay.

Histology showed a split at the dermal-epidermal junction where a blister cavity formed with paucicellular inflammation. Also, indirect immunofluorescence demonstrated dermal staining of IgG at the basement membrane zone, as well as presence of IgG bullous pemphigoid antibodies 180 and 230.

A diagnosis of epidermolysis bullosa acquisita was made. A major feature, subepidermal blistering, is caused by antibiodies to collagen type VII.

"Our patient had an underlying diagnosis of [systemic lupus erythematosus] and then developed epidermolysis bullosa acquisita," said Dr. Brown, a dermatology resident at the University of Louisville (Ky.). Because of its rarity, there are no randomized studies of this disease, only case reports in the literature (Cutis 2002;70:31-4; Clin. Exp. Dermatol. 1993;18:378-80).

"Histology cannot distinguish epidermolysis bullosa acquisita from BSLE. There is, however, histological overlap," Dr. Brown said in an interview. For example, BSLE and the inflammatory variant of epidermolysis bullosa acquisita each can feature neutrophils, but this finding is variable.

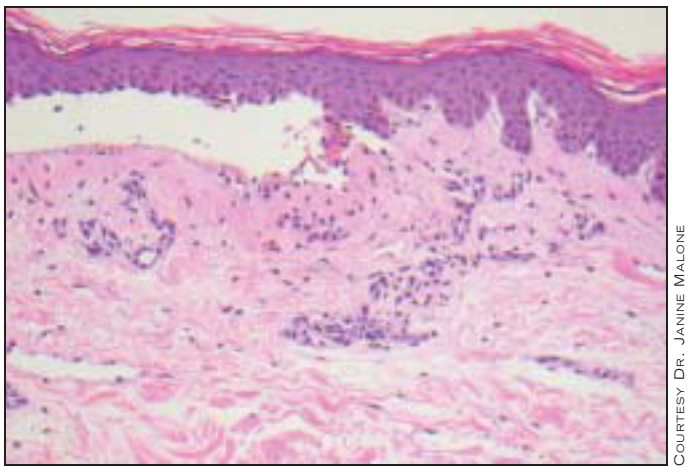
"What helps you most to distinguish between these entities is the presence or absence of certain clinical clues, as well as the course of the disease," Dr. Brown said. BSLE in general does not occur over trauma-prone areas, nor does it heal with scarring, milia, or the development of calcinosis. Epidermolysis bullosa acquisita is difficult to treat, whereas patients with BSLE generally respond to dapsone therapy.

Even given this challenge, a differential diagnosis is important between these two diseases. "They have similar histology but act very differently," Dr. Brown said.

While hospitalized, the patient continued to receive azathioprine 75 mg daily and prednisone 15 mg daily. She was discharged after 10 days. A short time later, the woman was admitted to an institution closer to her home with acute renal failure. She died from a pulmonary embolism.

Dr. Jeffrey P. Callen and Dr. Carol Kulp-Shorten, also of the University of Louisville, contributed to this case.

—Damian McNamara



Courtesy Dr. Janine Malone

Histology shows a split at the dermal-epidermal junction with paucicellular inflammation within the blister cavity.