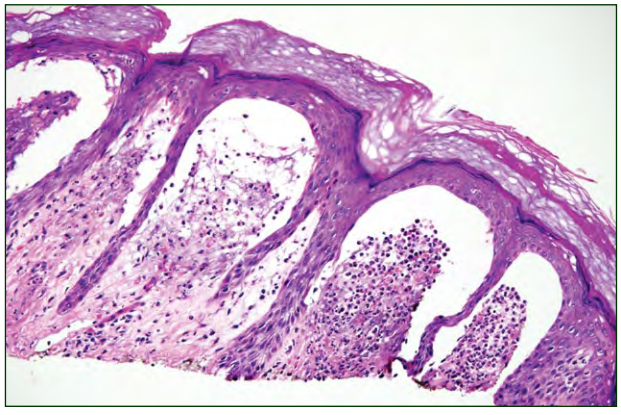

H&E, original magnification ×200.

The best diagnosis is:

a. bullous lupus erythematosus
b. bullous pemphigoid
c. dermatitis herpetiformis
d. epidermolysis bullosa acquisita
e. linear IgA dermatosis

PLEASE TURN TO PAGE 230 FOR DERMATOPATHOLOGY DIAGNOSIS DISCUSSION

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The authors report no conflict of interest.
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Dermatitis Herpetiformis

Dermatitis herpetiformis (DH) clinically presents with excoriated papules, often prominently involving the elbows, knees, and buttocks. Intact vesicles rarely are seen due to intense pruritus and consequent scratching. A superficial dermal neutrophilic infiltrate prominently involving the dermal papillae typically is noted on histopathology (Figure 1). Subepidermal bullae may be observed.[1] Direct immunofluorescence examination of perilesional skin shows granular deposition of IgA within the dermal papillae (Figure 2). Granular deposition of IgA along the dermoepidermal junction also may be observed in some cases (Figure 3). Most or all patients with DH have concurrent enteropathy secondary to celiac disease, which histologically presents as villous atrophy (Figure 4). Bullous pemphigoid,

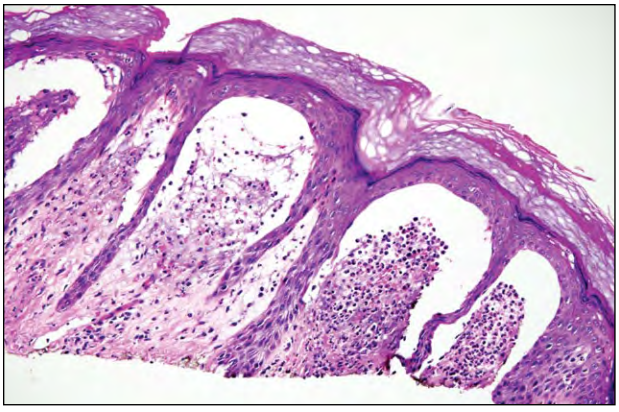

Figure 1. Perilesional skin of dermatitis herpetiformis (H&E, original magnification ×200).

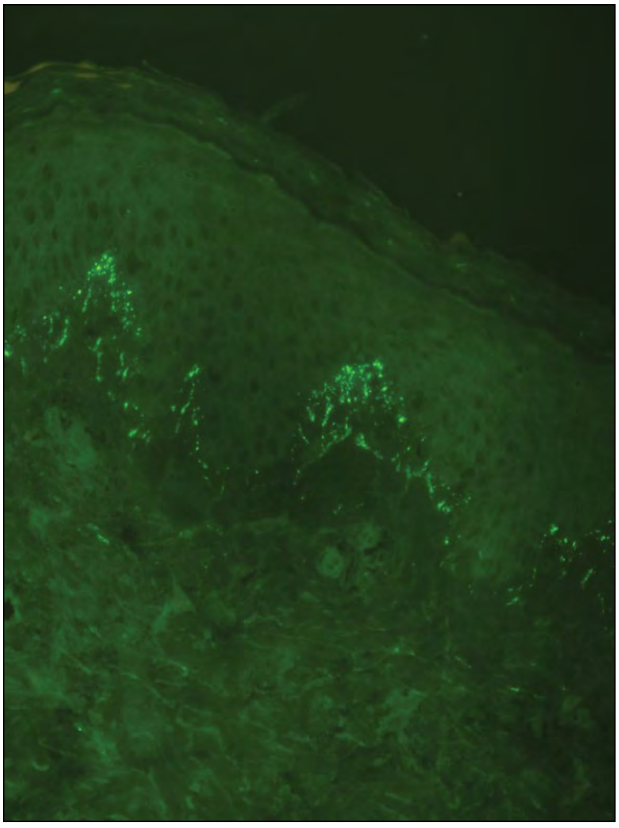

Figure 2. Direct immunofluorescence showing granular deposition of IgA within the dermal papillae that is characteristic of dermatitis herpetiformis (original magnification ×40).

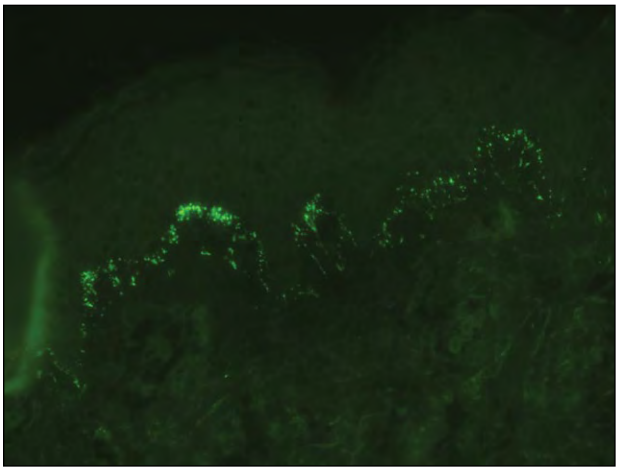

Figure 3. Direct immunofluorescence with granular deposition of IgA along the dermoepidermal junction and within dermal papillae that is characteristic of dermatitis herpetiformis (original magnification ×20).

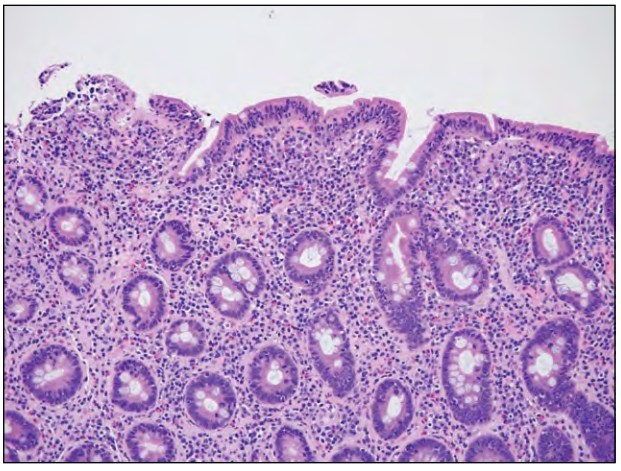

Figure 4. Jejunal villous atrophy of dermatitis herpetiformis (H&E, original magnification ×200).

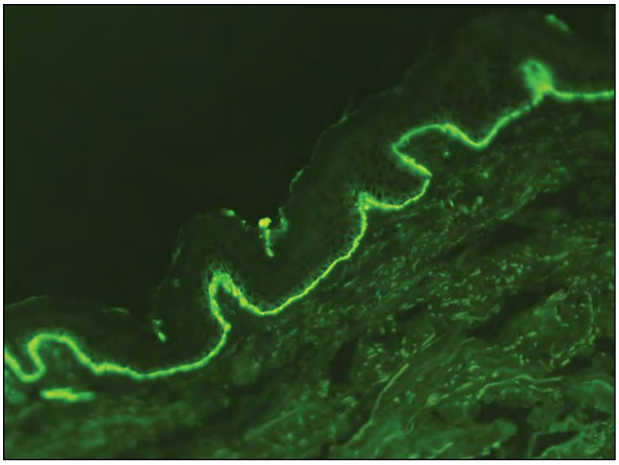

Figure 5. Direct immunofluorescence with a linear pattern of C3 deposition along the dermoepidermal junction that is characteristic of bullous pemphigoid (original magnification ×20).

linear IgA dermatosis, bullous lupus erythematosus, and epidermolysis bullosa acquisita (EBA) can cause subepidermal bullae and may present with collections of neutrophils within the dermal papillae. However, these disorders differ from DH because they lack granular IgA within the dermal papillae and there is a linear pattern of immunofluorescence along the dermoepidermal junction (Figure 5). In bullous pemphigoid, direct immunofluorescence typically demonstrates linear deposition of IgG and C3 deposition at the basement membrane zone in an *n*-serrated pattern, while linear IgA dermatosis shows IgA deposition in a similar pattern. Bullous lupus erythematosus and EBA often show linear deposition of IgG and C3 deposition in a *u*-serrated pattern.[2] Direct or indirect immunofluorescence on salt-split skin also can help distinguish bullous pemphigoid from EBA and bullous lupus erythematosus.

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1. Elston DM, Ferringer T, eds. *Requisites in Dermatology: Dermatopathology*. China: Saunders Elsevier; 2009.
2. Vodegel RM, Jonkman MF, Pas HH, et al. U-serrated immunodeposition pattern differentiates type VII collagen targeting bullous diseases from other subepidermal bullous autoimmune diseases. *Br J Dermatol*. 2004;151:112-118.