Pruritic eruption on the chest, arms, and buttocks

JENNIFER CLAY CATHER, MD, AND M. ALAN MENTER, MD

Figure 1. Erythematous papules, vesicles, and tense bullae on the chest of a middle-aged man.

Figure 2. Few intact vesicles and excoriated lesions with crusting are seen on the elbows.

An otherwise healthy middle-aged white man presented with a long-standing, extremely pruritic eruption on his chest, arms, and buttocks. Previous therapies, including multiple topical steroids, systemic antihistamines, and an occasional course of oral steroids, had provided limited relief. Examination revealed erythematous and urticarial papules and discrete small bullae and vesicles (*Figure 1*). Closer examination revealed numerous crusted lesions along with the vesicles (*Figure 2*). A skin biopsy taken from the edge of a small vesicle is shown in *Figure 3*.

What is the diagnosis?

Figure 3. Skin biopsy of a vesicle reveals a subepidermal split with a collection of neutrophils in the suprapapillary dermis (hematoxylin-eosin).

From the Division of Dermatology, Department of Internal Medicine, Baylor University Medical Center, Dallas, Texas.

Corresponding author: Jennifer Clay Cather, MD, 5310 Harvest Hill Road, Suite 260, Dallas, Texas 75230.

DIAGNOSIS: Dermatitis herpetiformis (Dühring's disease).

DISCUSSION

Symmetrical, extremely pruritic, and often urticarial papules and vesicles that have a predilection for the elbows, knees, and sacrum are characteristic features of dermatitis herpetiformis. Intact vesicles are rare, and frequently only crusts or scars are seen. Bullae, if present, are usually small and tense. Intense pruritus or burning usually precedes visible lesions by 8 to 24 hours. Oral lesions are less common and rarely symptomatic. While this disease may begin at any age, the lesions typically appear in the second or third decade of life, with men being affected more often than women. The disease is chronic in most patients; however, symptoms may lessen after the first 10 years. Recurrent crops of lesions are commonly followed by a brief remission. Exacerbations have been seen after potassium iodide intake. Several haplotypes have been associated with dermatitis herpetiformis, including HLA-A1, HLA-B8, HLA-DR3, and HLA-DQw2 (1).

Gluten-sensitive enteropathy is present to some degree in 60% to 70% of patients with dermatitis herpetiformis; however, only 5% to 10% of patients have symptoms, including diarrhea, bloating, and abdominal pain. Approximately 20% to 30% of patients have evidence of malabsorption, such as steatorrhea, abnormal D-xylose absorption, and abnormal iron, folate, glucose, water, and bicarbonate absorption (1).

Some authors report an increased incidence of gastrointestinal lymphomas and other malignancies in these patients (2). Recently, a gluten-free diet was found helpful in reducing the risk of gastrointestinal lymphoma (3). Additionally, autoimmune diseases, especially thyroid disorders, may be seen in patients with dermatitis herpetiformis (4).

The diagnosis of dermatitis herpetiformis relies on clinical examination, routine histology, and immunofluorescence studies (5). Skin biopsies obtained from lesions reveal collections of neutrophils at the tips of dermal papillae, along with a separation of the dermal papillary tips from the overlying epidermis (Figure 3). Direct immunofluorescence studies on perilesional or even normal skin reveal granular deposits of IgA with or without C3 deposits at the dermal-epidermal junction.

The pathogenesis of dermatitis herpetiformis is unknown but is believed to involve IgA deposition followed by complement activation via the alternative pathways. IgA may be directed against gluten protein or other cross-reacting antigens that originate in the gastrointestinal tract (1). Additionally, an increased incidence of HLA-B8, HLA-DR3, and HLA-DQw2 are seen in both celiac sprue and dermatitis herpetiformis, suggesting a common pathogenesis.

Extreme pruritus, often out of proportion to skin findings, may be seen in dermatitis herpetiformis, scabies, eczema, and urticaria. If intact vesicles or bullae are seen, then bullous pemphigoid, bullous lupus, chronic erythema multiforme, and linear IgA dermatoses are often considered in the differential diagnosis.

Rapid improvement in cutaneous lesions is seen with sulfones and sulfapyridine; however, no change is seen in the gastrointestinal lesions. Diaminodiphenylsulfone (dapsone) is the most effective sulfone at doses ranging from 50 to 300 mg daily. Acute hemolytic anemia may occur in patients with glucose-6-phosphate dehydrogenase deficiency; therefore, this enzyme level should be tested prior to therapy. Patients treated with dapsone should be closely monitored for bone marrow suppression. A strict gluten-free diet will produce remissions in intestinal and cutaneous lesions; however, it takes 6 to 12 months for skin improvement to occur (6), thus limiting patient compliance. Cutaneous lesions reappear within 1 to 3 weeks after patients discontinue a gluten-free diet.

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