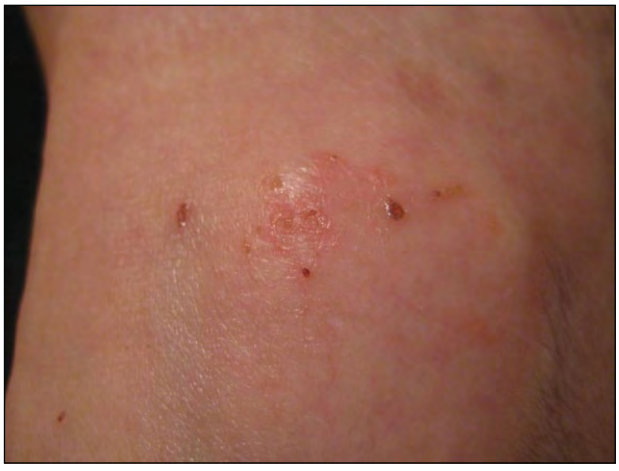
Letter to the Editor

Non–Dermatitis Herpetiformis Gluten-Sensitive Dermatitis: A Personal Account of an Unrecognized Entity

To the Editor:

As a dermatologist of European descent practicing in the United States, I wanted to share my account of an unrecognized identity to alert practicing physicians to the existence of a gluten-sensitive dermatitis that clinically resembles dermatitis herpetiformis (DH) but lacks its expected laboratory features.

At 55 years of age, I developed dermatitis on my dorsal hands that progressed despite use of topical steroids and tacrolimus ointment and avoidance of various topical exposures. In 2 years it gradually spread to the elbows, knees (Figure), buttocks, nucha, upper back, dorsal feet, ankles, legs, and ears and caused severe burning pruritus that greatly interfered with sleep and quality of life. There were no intestinal symptoms, but an unexplained steady weight loss of 15 lb occurred over a year.



Typical eruption on the knee with erythema and excoriated 1- to 2-mm vesicles.

The morphology and distribution were typical for DH, the skin disease associated with gluten-sensitive enteropathy.[1] Although I had a firm clinical diagnosis without anything else in the differential, I performed a confirmatory biopsy before committing myself to a lifetime gluten-free diet, which is not an easy one to keep. To my surprise, results of repeated skin biopsies only showed nonspecific dermatitis and negative direct immunofluorescence. Serum IgA or IgG anti–tissue transglutaminase or antiendomysial antibodies also were negative. I have HLA-DQ2 and HLA-DQ8 genotypes, which are expected in DH and celiac disease but also present in 40% of the normal population.[2] Protein contact dermatitis, delayed hypersensitivity, and IgE-mediated allergy to gluten were ruled out. Small intestinal biopsy would have been helpful, but it was not approved in my managed-care setting without the laboratory hallmarks for DH. Because DH was ruled out, my dermatology colleagues recommended empirical treatment with methotrexate, psoralen plus UVA, and systemic steroids for my dermatitis. Oral steroids were somewhat helpful, but antihistamines and dapsone were ineffective.

Because the clinical picture was characteristic of DH, I decided to try a strict gluten-free diet despite the non-DH histology and immunofluorescence results. The intensity of the pruritus drastically decreased in a few days. The eruption improved 50% within 2 months and 95% to 99% in 2 years, restricted only to occasional lesions on the dorsal hands. Oral iodide and ibuprofen caused the dermatitis to flare. After 3 years of being essentially symptom free, I resumed a regular diet. Within 3 months the eruption appeared once again in the same distribution along with severe pruritus. Upon returning to the gluten-free diet, the rash again cleared in a few months and remains 99% clear 6.5 years after the initial diagnosis of non-DH gluten-sensitive dermatitis.

Early studies in the 1970s reported that 97% of patients with a DH-like clinical picture had cutaneous IgA deposits,[3] which became the diagnostic hallmark of DH[4]; therefore, patients without this sine qua non finding were excluded from later studies. The information on the fate of these early IgA-negative patients is scant; most of them eventually were considered to have eczema,[5] which is a nonspecific diagnosis. They understandably were not put on a strict gluten-free diet, as such a diet was not yet developed and would have been impossible to effectively maintain more than a quarter century ago.

It usually takes many years before an average gluten-sensitive patient is diagnosed with DH. If I had not been a dermatologist convinced enough by a

clinically gluten-sensitive dermatitis pattern to start a serious diet, would I eventually have developed IgA deposits and antibodies to earn a diagnosis of DH some years later? There is no way to know.

I report this case to alert physicians to think of gluten-sensitive dermatitis when a clinical picture suggests DH but lacks laboratory confirmation. A severely pruritic rash affecting the elbows, knees, buttocks, nucha, and upper back might warrant an honest trial of a gluten-free diet for a few months. It can save much misery from the disease and potential harm from unnecessary treatments, and it might possibly avert an enteropathy-related malignancy.[6]

I also report this rare case of gluten-sensitive dermatitis to stimulate thinking about the possible mechanism through which gluten may lead to a skin eruption. As Bateson[7] advised in 1908, "[t]reasure your exceptions" for they guide where research should go. If IgA has a pathognomonic role in DH, which is not yet proven, it might not be the only pathway leading to the typical skin eruption. It also is possible that its presence is only a later side event of gluten sensitivity and not a necessary cause of the dermatitis.

Sincerely,
Katalin Soltesz Korossy, MD
Kensington, Maryland

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