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CASE REPORT

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Celiac disease presenting as dermatitis herpetiformis: A case report

Ali Kirik[1], Sinan Ozcelik[2], Eren Altun[3], Figen Aslan[3], Gulhan Zorgor Ucdu[1], Teoman Dogru[4]

[1]Balikesir University, Faculty of Medicine, Department of Internal Medicine, Balikesir, Turkey
[2]Balikesir University, Faculty of Medicine, Department of Dermatology, Balikesir, Turkey
[3]Balikesir University, Faculty of Medicine, Department of Pathology, Balikesir, Turkey
[4]Balikesir University, Faculty of Medicine, Department of Gastroenterology, Balikesir, Turkey

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Abstract

Dermatitis herpetiformis, a specific cutaneous manifestation of Celiac disease, is characterized by herpetiform clusters of pruritic urticated papules and vesicles on the skin and granular IgA deposits in the dermal papillae. In the present report, we describe a case of 39-year old male who is previously diagnosed as DH, and after this diagnosis he was serologically and histologically confirmed to have CD.

Keywords: Dermatitis herpetiformis, Celiac disease, autoimmune disease

Introduction

Dermatitis herpetiformis (DH), an inflammatory disease of the skin, is characterized by herpetiform clusters of pruritic urticated papules and vesicles on the skin and granular IgA deposits in the dermal papillae. It is associated with several autoimmune disorders, including type I diabetes mellitus, autoimmune thyroid diseases, and connective tissue diseases, such as Sjögren syndrome.

DH is also considered as a specific cutaneous manifestation of Celiac disease (CD). Both DH and CD occur in gluten-sensitive individuals, share the same Human Leukocyte Antigen (HLA) haplotypes (DQ2 and DQ8), and improve following the administration of a gluten-free diet. Moreover, almost all DH patients show typical CD alterations at the small bowel biopsy, ranging from villous atrophy to augmented presence of intraepithelial lymphocytes, as well as the generation of circulating autoantibodies against tissue transglutaminase (tTG).

In the present report, we describe a case of 39-year old male who is previously diagnosed as DH, and after this diagnosis he was serologically and histologically confirmed to have CD.

***Coresponding Author:** Ali Kirik, Balikesir University, Faculty of Medicine, Department of Internal Medicine, Balikesir, Turkey
E-mail: alikirik87@hotmail.com

Case Report

A 39-year-old male patient was admitted to the dermatology department with pruritic lesions on the limbs, lower back skin with 2 months. The patient had no gastrointestinal symptoms. His medical history revealed no particularities such us history of chronic alcohol, herbal and drug use. His family history was unremarkable. On physical examination, the patient was in good general condition. Small vesicles and papules were seen symmetrically distributed on the extensor surface of the limbs, buttocks and lower back (Figure-1). No other abmormalities were observed in physical examination. In serial blood testing, elevated liver enzymes (AST: 51 IU/L, ALT: 67 IU/L, GGT: 151 IU/L) were found. The other laboratory analysis results were as follows; hemoglobin: 13.7 g/dL, creatinine: 0.81 mg/dL, INR: 1, erythrocyte sedimentation rate: 34 mm/h, C reactive protein (CRP): 20.3 mg/L. Viral serologies for hepatitis (A, B, C, and E) and markers for autoimmune liver disease (antinuclear antibody, antimitochondrial antibody, liver kidney microsomal type 1 antibody) were negative. The abdominal ultrasonography of the patient was normal. Skin punch biopsy was performed by a dermatologist with the result compatible with DH.

The skin biopsy revealed collections of neutrophils in the papillary dermis as well as clefting at the dermoepidermal junction. Direct immunofluorescence (DIF) showed granular IgA deposition in the papillary dermis (Figure-2A, 2B).

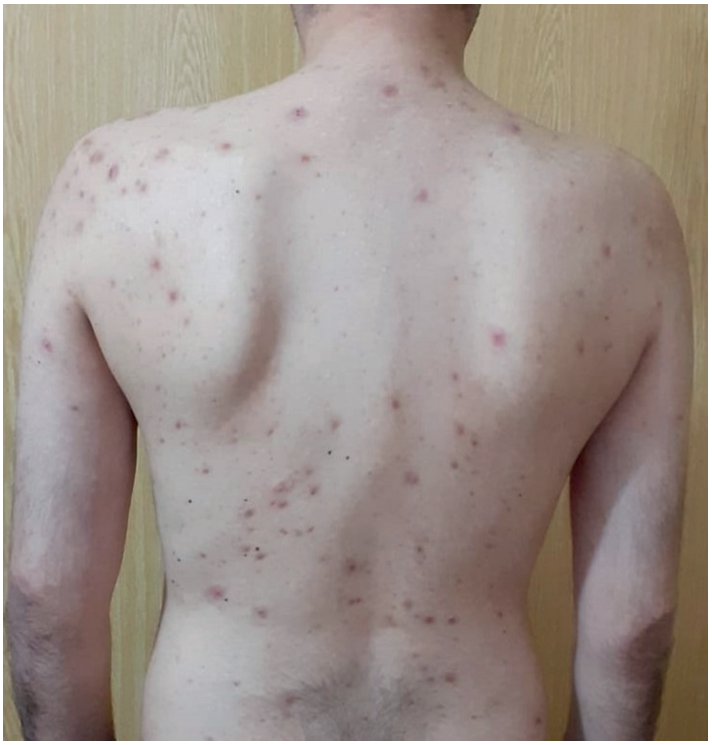

Figure 1. Small vesicles and papules, on theextensor surface of the limbs and lower back

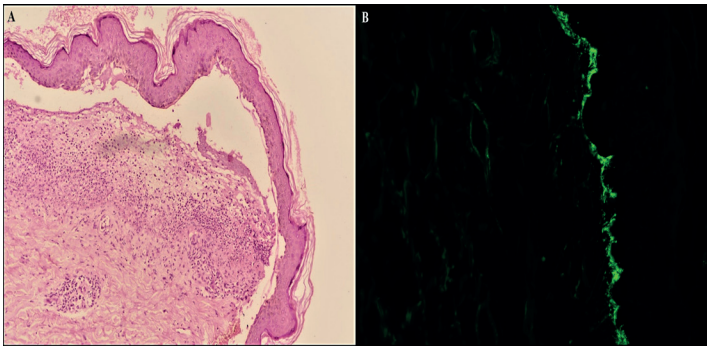

Figure 2A. Subepidermal dissociation and intense inflammatory infiltration from neutrophils (H&E, x40). **Figure 2B.** Granular IgA deposition in the papillary dermis (DIF, x400)

The patient was consulted with gastroenterology department because of the persistently elevated liver enzymes. Because of the well-known association between CD and DH, serologic tests and upper gastrointestinal endoscopy were planned. Antibodies to tissue transglutaminase IgA (36.9 IU/mL), endomysial IgA (95 IU/mL) and gliadin IgA (45.8 U/mL) were all positive. Subsequently, upper gastrointestinal endoscopy and duodenal biopsy was performed. During the endoscopy, the duodenal mucosa had a mosaic appearance in the bulb and complete villous atrophy was found in the second part of the duodenum (Figure-3). Histopathological examination of the duodenal biopsies revealed villous atrophy, as well as marked intraepithelial lymphocytosis, suggesting stage 3a mucosal damage (Marsh-Oberhuber classification) (Figure-4). A gluten-free diet was introduced with the confirmed diagnosis. The patient presented favorable clinical evolution with partial regression of lesions.

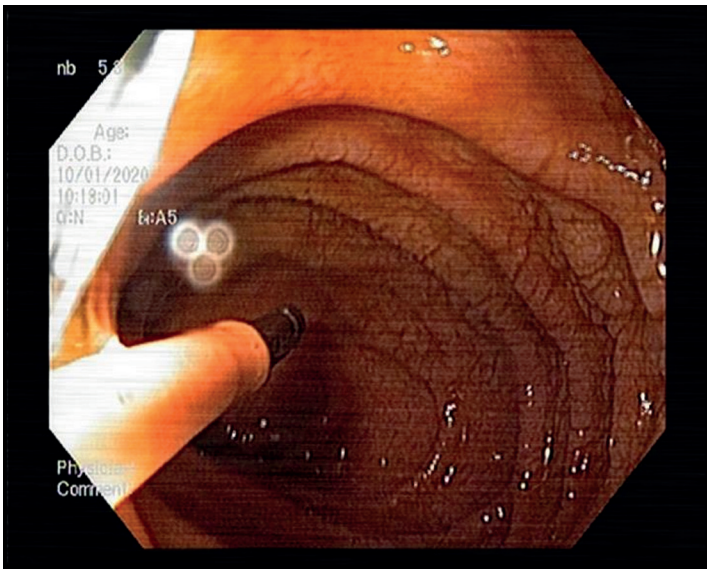

Figure 3. Complete villous atrophy in the second part of the duodenum and a mosaic appearance in the duodenal mucosa.

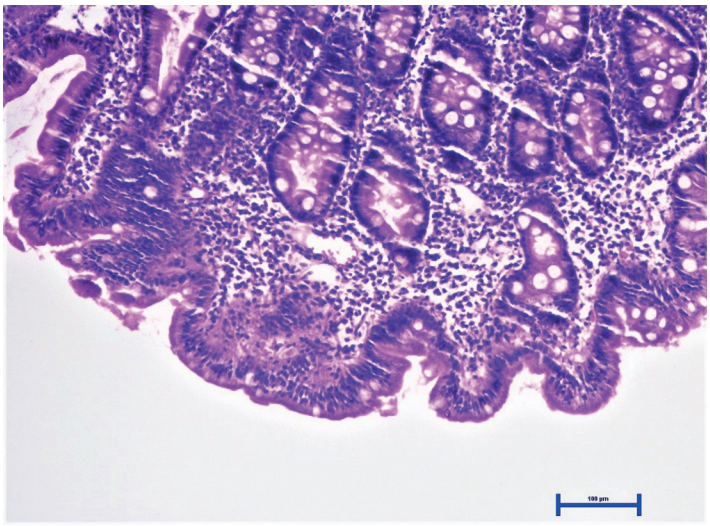

Figure 4. Intraepithelial lymphocytosis and villous atrophy in the duodenal biopsies (H&E, x100).

Discussion

In 1884, DH was firstly described as a clinical entity by Louis Duhring (1). It is a rare, chronic autoimmune skin disease caused by a reaction to gluten ingestion. In adults, the mean age at diagnosis is between 30 and 40 years for DM, but the disease can occur at any age. However, it is rarely occurs in children and elderly. The prevalence of DH has been reported as high as 10 cases per 100.000 population. Recent evidence is growing that men are more affected than women (2).

The clinical presentation of DH is often highly suggestive. The symmetrical distribution of small vesicles and papules which typically on the elbows, knees, and buttocks is the most prominent symptom of the disease. The upper back, abdomen, scalp and face can also be affected, but oral lesions are rare. These lesions are often eroded and crusted because of intense itch and scratching. Therefore, itchy skin disorders such as urticaria, atopic or nummular dermatitis, and scabies infestation should be considered

in the differential diagnosis (2). The ideal method for diagnosis of DH is a direct immunofluorescence biopsy of unaffected skin in close proximity to an active lesion (4). This reveals pathognomonic granular IgA deposits at the dermo-epidermal junction (5).

DH is associated with several autoimmune diseases including hypothyroidism, type 1 diabetes mellitus, pernicious anemia, and CD (6). On the other hand, the most common extraintestinal manifestation of CD (more than 90% of patients) is DH. Hence, these two condition share the same genetic background, with a high frequency of HLA-DQ2 or HLA-DQ8 haplotypes (7). In clinical practice, gastrointestinal manifestations rarely occur in patients with DH suffering from CD. However, intestinal (duodenal) biopsies show CD manifested by blunting of villi, crypt hypertrophy, and lymphocyte infiltration of crypts. However, it should be noted that, a quarter of the patients may have normal histological findings (8). In addition, CD is commonly associated with elevated liver enzymes that normalize on a gluten-free diet (9).

Currently, the only effective treatment for all patients with DH is a strict adherence to a gluten-free diet (GFD), regardless of whether they have pathological abnormalities in the small intestine. Skin manifestations in DH responds to a GFD, albeit slowly, and the symptoms recur on gluten challenge. Therefore, maintaining the life-long GFD is required for remission of this condition (5). As reported in CD, the increased risk of non-Hodgkin's lymphoma is the main factor for the long-term prognosis of DH. Hence, it has been reported that, a strict GFD for more than five years seems to protect against lymphoma (3).

Conclusion

DH is a cutaneous manifestation of CD, and its presence should prompt testing for CD even without gastrointestinal symptoms.

Conflict of interests
The authors declare that they have no competing interests.

Financial Disclosure
The authors don't need financial suppport.

Patient informed consent
Consent of patient was obtained.

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