Dermatitis Herpetiformis: A Case Report

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Abstract

Dermatitis herpetiformis (DH) is an intensely pruritic, chronic recurrent papulovesicular disease. Definitive diagnosis of DH is made on the characteristic histological findings and direct immunofluorescence finding of granular or fibrillar IgA deposits along the basement membrane zone of biopsied perilesional skin. Although it is a relatively common disease in Caucasian populations, it is rare in Asian populations. Here, we report a 28-year-old male with DH who had severely pruritic, papular and papulovesicular lesions and was being treated as chronic dermatitis for a long time. Routine histopathology and the direct immunofluorescence supported the diagnosis of DH. He was started on dapsone therapy and his skin lesions cleared rapidly.

Introduction

Dermatitis herpetiformis (DH) is characterized by pruritic papules and vesicles occurring in herpetiform grouping (clustering like herpes simplex). There is symmetrical involvement of the extensor surfaces and involvement of the elbows, knees, shoulders, nape of the neck, sacral area, scalp, and mucous membranes. Although this is a bullous disorder, large blisters such as seen in pemphigus vulgaris or bullous pemphigoid are distinctly uncommon. It is usually associated with gluten sensitive enteropathy. Although it is a relatively common disease in Caucasian populations, it is rare in Asian populations and the association of gluten sensitive enteropathy is also rare[1]. Young male adults are most commonly affected but it may occur at any age. Definitive diagnosis of DH depends on the characteristic histological findings and direct immunofluorescence finding of granular or fibrillar Ig A deposits along the basement membrane zone of biopsied perilesional skin[2]. Here, we present a 28-year-old male patient who had been suffering from DH for more than 4 years.

Case report

A previously healthy 28-year-old man presented to our dermatology OPD with 4 and a half years history of persistent, papulovesicular lesions. The lesions had first appeared as small reddish papules and plaques accompanied by intense pruritus.

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Latter, there developed vesicles and superficial erosions (Figure 1).

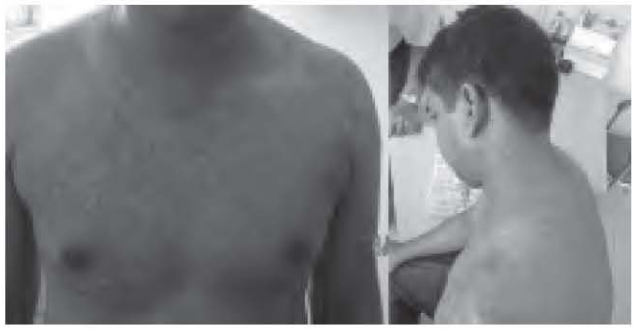

Figure1. Grouped vesicular lesions on erythematous base on the neck, shoulder and front of the chest.

There was severe burning sensation. He was treated with topical corticosteroid and sometimes with topical as well as systemic corticosteroids. Although there was a minimal decrease in intensity of symptoms, the lesions were always there. There was much worsening of the symptoms during the summer. Gastrointestinal and other constitutional symptoms were not present. On examination, there were grouped vesicular lesions on erythematous base predominently on the shoulders, nape of the neck, front and back of the chest, buttocks, extensor surface of the arms, forearms and hands, elbows and also the lower limbs including front of the knees, with relative sparing of the face, palms, soles and the mucous membrane. On laboratory work-up complete blood count (CBC), liver function tests, urine analysis, creatinine, thyroid-stimulating hormone (TSH), and serum IgE were normal. Considering the long history of the eruption together with the poor response to topical and systemic corticosteroids, differential diagnoses included autoimmune bullous diseases. So, a biopsy specimen was taken from the margin of a fresh lesion situated on the neck and clinically uninvolved skin. The specimen was processed for routine histology and direct immunofluorescence (DIF) testing. Hematoxylin-eosin-stained sections revealed neutrophilic microabscesses in the dermal papilla with subepidermal vesicle formation, suggestive of DH. Superficial and deep perivascular and interstitial infiltrate consisting of lymphocytes and neutrophils was also noted. Perilesional skin was submitted for direct immunofluorescence studies. DIF revealed finely granular IgA deposits along the basement membrane with marked accumulation at the tips of dermal papilae (Figure 2).

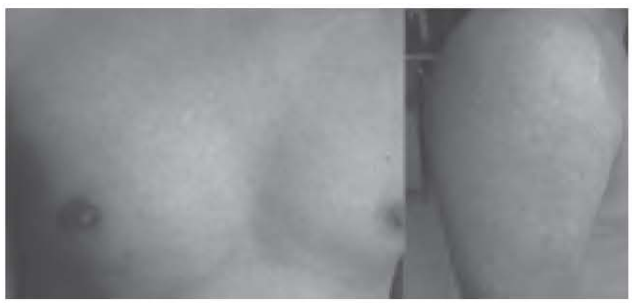

Figure 2. Clearing of the lesions from the shoulder and front of the chest four months after treatment with dapsone. Only depigmented areas are seen.

As the patient did not have any gastrointestinal symptoms he did not give consent for gastrointestinal endoscopy which we wanted to perform to see if there was any asymptomatic celiac disease or not. Antiendomysial antibodies of the IgA isotype were found in the serum. The diagnosis of dermatitis herpetiformis was confirmed and the patient was put on a dapsone and gluten-free diet (gluten-containing cereals, including wheat, barley). Soon after the starting of the treatment the patient started improving symptomatically and there was no new active lesion after one week and the older lesion was healing. Later the patient was completely free of symptom and when we took the final photographs after 4 month there was no lesions except post inflammatory depigmentations. (Figure 3)

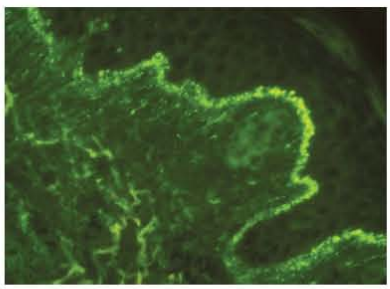

Figure 3. Direct immunofluorescence of skin specimen taken from a patient with DH showing granular-linear deposition of IgA along the dermoepidermal junction[7]

Discussion

Dermatitis herpetiformis usually presents in the third decade, although individuals of any age can be affected. It is predominately a disorders of Caucasians; the incidence was found to be 11.5 per 100 000 in Scotland and ranging form 19.6–39.2 per 100 000 in Sweden[3]. In our country the diagnosis of dermatitis herpetiformis is not that frequent. Many of the patients get treatment as atopic dermatitis for a considerable period of time without much benefit. The key reason may be the fact that it is not that common in Asian populations[1,3]. Moreover the association of gluten-sensitive enteropathy is rare in comparison to Caucasian populations[1]. In fact, the severity of DH is inversely proportional to the severity of the intestinal symptoms (i.e., severe DH is associated with mild intestinal disease). So any long-standing itchy, inflammatory and vesicular/erosive eruption in extensor distribution that is non-responsive to topical and systemic corticosteroids should include dermatitis herpetiformis in its differential diagnostic considerations and be accompanied by a prompt skin biopsy with appropriate histopathologic and immunofluorescence analysis[4].

The severity of DH is inversely proportional to the severity of the intestinal symptoms[5]. Our patient had severe DH which may explain the reason for the absence of gastrointestinal symptom in this case.

Dapsone is the drug of choice for DH and is highly effective for controlling skin lesions and pruritus, often clearing the eruption within 48–72 hours (dosages of 1/mg/kg/day)[3,6]. This was true in our patient's case also. Dapsone is associated with dose-related hemolytic anemia and methemoglobinemia in all patients; glucose-6-phosphate dehydrogenase deficiency is a contraindication to its use. Agranulocytosis is a rare complication and can occur early in the treatment course[7]. Weekly CBC and reticulocyte counts should be done until hemoglobin levels are stable, then monthly to bimonthly testing is appropriate[3]. In our patient the regular monitoring of laboratory parameters were uneventful and no adverse drug reaction was observed.

References

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