



Review Article

Dermatitis Herpetiformis- its Presentation and Management

M Moazzem Hossain¹

Abstract

Dermatitis herpetiformis (DH) is a chronic, recurrent, intensely pruritic eruption occurring symmetrically on the extremities and the trunk and comprising tiny vesicles, papules and urticarial plaques that are arranged in groups. It is associated with gluten-sensitive enteropathy and IgA deposits in skin¹. Dermatitis herpetiformis responds well to dapsone 100mg/day and then reducing to 50mg/day. The dose of dapsone may further be reduced with gluten free diet.

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Introduction

Dermatitis herpetiformis is a vesico bullous disorder which is based on auto-immunity. In this disease, there is deposition of IgA antibodies at the tips of the dermal papillae. Clinically, the disease is characterised by recurrent episodes of intense itching and eruptions of grouped papules, papulo-vesicles or vesicles on the skin over the trunk and extremities². Lesions are usually distributed-symmetrically on extensor surfaces. The blisters vary in size from very small up to 1 cm across. The condition is extremely itchy, and the desire to scratch can be overwhelming¹⁵. This sometimes leading to the blisters being scratched off before they are examined by a doctor. Intense itching or burning sensations are sometimes felt before the blisters appear in a particular area^{7,18}. The severity can vary from week to week but it rarely clears up without specific treatment. Untreated, the severity of DH can vary significantly over time, probably in response to the amount of gluten ingested¹⁶. This disease in addition, is associated with an enteropathy which may be completely asymptomatic in some patients^{2,3}.

Despite its name, DH is not related to or caused by herpes virus: the name means that it is a skin inflammation having an appearance similar to herpes. DH was first described by Dr. Louis Duhring in 1884⁸. A connection between DH and gluten intolerance (coeliac disease) was recognised in 1967^{5,6}, although the exact causal mechanism is not known. The age of onset is usually about 15-40, but DH can also affect children and the elderly. Men and women are equally affected. Estimates of DH prevalence vary from 1 in 10000 to 1 in 400^{10,11,12,13}.

Diagnosis

Diagnosis is confirmed by a simple blood test for IgA antibodies¹⁷, and by a skin biopsy in which the pattern of IgA deposits in the dermal papillae, revealed by direct immunofluorescence, distinguishes it from linear IgA bullous dermatosis⁸ and other forms of dermatitis. These tests should be done before the patient starts on a gluten-free diet, otherwise they might produce false negatives. If the patient has already started a gluten-free diet, it might be necessary for them to come off it for some weeks before the tests can be done reliably^{1,2,3}.

¹ Associate Professor, Department of Dermatology, Rajshahi Medical College, Rajshahi.

Treatment

Dermatitis herpetiformis responds well to medication and changes in diet. Dapsone is an effective treatment for most patients. DH responds to dapsone so quickly that itching is significantly reduced within 2-3 days¹⁵ that this response may almost be considered diagnostic. However, dapsone treatment has no effect on any intestinal damage¹³. A strict gluten-free diet must therefore also be followed¹⁷, and this will usually be a lifelong requirement. This will reduce any associated intestinal damage^{15,17}, and the risk of other complications. After some time on a gluten-free diet, the dosage of dapsone can usually be reduced or even stopped¹⁵, although this can take up to anything from 1 to 3 years. Dapsone is an antibacterial, and its role in the treatment of DH, which is not caused by bacteria, is poorly understood. It can cause adverse effects on the blood, and regular blood monitoring is required⁷.

Dapsone is the drug of choice, but for patients unable to tolerate dapsone for any reason, the following can be tried, although they are less effective: colchicine, lymecycline, nicotinamide, tetracycline⁴, sulfamethoxypyridine, sulfapyridine^{7,17}.

Topical steroid, systemic corticosteroid can also be used as for other autoimmune diseases. Antihistamine is used for itching.

Complications

DH is an autoimmune disease, and patients with DH are more likely than others to have thyroid problems^{11,17} and intestinal lymphoma.^{11,12,14}

Conclusion

Dermatitis herpetiformis is most common among the bullous diseases. This disease is mostly diagnosed clinically by the experienced dermatologist. Selection of the effective drugs, dose adjustment and monitoring of the haematological and other side effects are important. Dapsone may cause acute haemolytic anaemia which may be severe in glucose-6-phosphate dehydrogenase deficiency. Glucose-6-phosphate dehydrogenase level should be done before therapy if possible. Dapsone should be

prescribed at a low starting dose (25mg/day) and watch the patient closely for dark urine. Dark urine is due to excess haemolysis.

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All correspondence to:
M Moazzem Hossain
Associate Professor
Department of Dermatology
Rajshahi Medical College, Rajshahi