Dermatitis Herpetiformis - A skin disorder

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What is Dermatitis Herpetiformis?

Dermatitis herpetiformis is celiac disease of the skin. Dermatitis herpetiformis accompanies celiac disease of the small intestine. Celiac disease is a permanent intolerance to gluten, a protein found in various wheats (e.g., durum, kamut, spelt), rye, barley and triticale. Gluten consumption causes damage to the absorptive surface of the small intestine and can result in malnutrition, anemia, nutritional deficiencies, and an increased risk of other diseases including osteoporosis and specific cancers of the gut.

The skin rash of dermatitis herpetiformis is intensely burning and itchy. The earliest findings are groups of small blisters (2-5 mm) which soon erupt into small erosions, either spontaneously or from being scratched.

The elbows, knees and buttocks are most often involved in symmetrical fashion. The back of the neck, upper back, scalp, hairline and face are less frequently involved (1). The blisters resemble those of herpes simplex (herpetiformis) but there is no relationship to herpes viruses.

Dermatitis herpetiformis and celiac disease are inherited and related autoimmune disorders that share the same genetic pathways and chromosome features. Both respond to gluten withdrawal from the diet.

Prevalence

Dermatitis herpetiformis occurs in 10% of patients with celiac disease that itself affects 1 in 100-200 Canadians. Onset of dermatitis herpetiformis is usually early to middle adult life but can occur in children and later in life.

Diagnosis

Dermatitis herpetiformis is only diagnosed and confirmed by a dermatologist obtaining a slight skin biopsy from uninvolved skin adjacent to blisters or erosions (1, 3). Other forms of dermatitis can mimic dermatitis herpetiformis necessitating skin biopsy for correct diagnosis (4).

Small bowel biopsies will confirm a diagnosis of coexisting celiac disease but are not essential if the skin biopsy confirms the diagnosis of dermatitis herpetiformis (5).

Referral to a gastroenterologist may be necessary for assessing the extent of the underlying intestinal injury and associated deficiencies of iron, calcium and vitamins.

The skin symptoms usually predominate over intestinal symptoms (1, 3). Blood tests for celiac disease may be negative, reflecting the absence or paucity of intestinal symptoms expected when there is milder, more patchy villous atrophy seen on small bowel biopsies (2).

Treatment

Dapsone is necessary for the immediate treatment of the dermatitis by providing rapid relief from the burning and itching. Ongoing treatment with dapsone may be necessary to maintain remission. Flare-ups brought on by inadvertent or intentional gluten ingestion will require additional courses of dapsone (6).

A life-long gluten-free diet is required to reduce the risk of associated conditions and to decrease or eliminate the need for dapsone (6). The gluten-free diet is complex so that patients should be referred to a registered dietitian with expertise in celiac disease for nutrition assessment, education and follow-up. All people with dermatitis herpetiformis are encouraged to join the local chapter of the Canadian Celiac Association for valuable practical information and ongoing support.

Bibliography

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5. NIH consensus document web link: http://consensus.nih.gov/cons/118/118celiacPDF.pdf

6. Alaedini A and Green PHR. Narrative review : celiac disease: understanding a complex immune disorder. Ann Intern Med 2005; 142: 289-98.

CCA Chapters

For more information on celiac disease, dermatitis herpetiformis, and the gluten-free diet, please contact the national office or the local chapters of the Canadian Celiac Association.

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