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DERMATITIS HERPETIFORMIS IN ASSOCIATION WITH TREATED COELIAC DISEASE

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Abstract. The paper describes an adult patient with coeliac disease who developed classical dermatitis herpetiformis while receiving a gluten-free diet. The dermatitis is controlled by sulphapyridine but relapses when this is withdrawn, even though the patient continues with the diet, indicating that a gluten-free diet plays little or no part in controlling the skin condition.

Key words: Gluten-free diet; Dermatitis herpetiformis; Coeliac disease; Sulphapyridine

Since Marks et al. (11) first reported small bowel abnormalities in patients with dermatitis herpetiformis (D.H.), a number of papers have been published confirming the association of D.H. and coeliac disease (1, 3, 5–10, 12–14, 16, 18, 19). It is now accepted that about two-thirds of patients with D.H. have associated abnormalities of the jejunal villi (1, 2, 3, 5, 7–9, 12–14, 16, 18, 19).

Brow et al. (3) reported that when multiple jejunal biopsies are carried out on every patient with D.H., histological changes are found in 95% of patients. They suggest that the villous atrophy is patchy in distribution and unless multiple biopsies are carried out, about a third of these cases would be passed as normal.

The relation between intestinal villous atrophy and the skin eruption in D.H. is still obscure. Treatment with a gluten-free diet definitely improves intestinal symptoms and the histological appearance of the jejunum (8, 9, 11, 13, 14, 18, 19) but its influence on the skin eruption is debatable. Some investigators have found a gluten-free diet to be beneficial in controlling the skin eruption in D.H. (8, 9, 18) but others find this therapy unhelpful in controlling the skin eruption and in reducing the minimum effective dose of sulphones required in the treatment of D.H. (2, 13, 18).

This paper reports a patient with coeliac disease who developed D.H. while on a strict gluten-free diet which had been taken conscientiously for the preceding eight months.

Present history

A 31-year-old woman reported to the skin clinic with a four month history of an itchy urticarial and vesicular eruption involving the buttocks, shoulders and elbows.

On examination, papules and grouped vesicles were present on the affected areas.

A skin biopsy showed a subepidermal bulla with a number of papillary microabscesses, the features being consistent with D.H. Direct immunofluorescence studies on the skin showed the typical granular and fibrillar type of basement membrane fluorescence to antihuman IgA conjugate (Molar F, $P=3.9$) as described in D.H. (4, 15, 17). Tests by the indirect method with patient's serum were negative, thus confirming the diagnosis of D.H.

Previous history

Eight months prior to referral to the skin clinic the patient complained of progressive tiredness and bilateral ankle swelling. There was no history of blood loss nor of diarrhoea or other bowel symptoms.

Results of previous investigations

Haemoglobin	31%
R.B.C.	1.05/mm^3
P.C.V.	118%
M.C.H.C.	36.2%
M.C.H.	44.2 ng
W.B.C.	2800/mm^3

Blood film. Aniso-poikilocytosis and macrocytosis with reduction of white cell and platelet count.

Sternal marrow. Maximum cellularity with stainable iron present. There was gross megaloblastic erythroid hyperplasia. The erythroid element showed marked saturation. Giant metamyelocytes were present but otherwise the granular series was maturing normally. Megakaryocytes were present.

Faecal fat estimation over 3 days: 11.8% of dry weight
Serum vitamin B_{12}: 74 pg/ml (low)
Schilling test: normal
Corrected serum folate: 890 mg % (normal)
Liver function tests, serum iron/T.I.B.C., serum proteins, serum transaminases, xylose test and immunoglobulins: normal
Lactose tolerance test—normal

Barium meal. Showed distortion of mucosal folds in the first part of the duodenum and a suggestion of an ulcer crater. Follow through revealed a pattern of typical malabsorption with widening of loops of small bowel and slight thickening of the transverse mucosal folds.

Jejunal biopsy from the first part of the jejunum showed an entirely flat mucosa and marked cellular infiltrate with polymorphs and plasma cells. On the basis of these findings, the patient was diagnosed as a case of adult-type coeliac disease with megaloblastic anaemia.

Treatment

For eight months prior to referral to the skin clinic the patient was treated with a strict gluten-free diet, parenteral cyanocobalamin and oral folic acid.

Her skin eruption is controlled with 0.5 g of sulphapyridine daily but relapses on reducing to 0.25 g daily.

A recent jejunal biopsy shows only a minor degree of partial villous atrophy with a mild diffuse chronic inflammatory infiltrate involving the lamina propria.

Her general physical health has improved greatly with an increase in energy and weight gain.

Discussion

The patient originally presented with anaemia and had no symptoms to suggest a diagnosis of coeliac disease. She had been taking oral contraceptives for one year prior to investigation of malabsorption and it is thought that this accounts for the normal serum folate level in spite of a low serum B_{12}.

Berg et al. (2) reported a case of D.H. whose skin symptoms were not affected by the diet and required sulphone therapy. Shuster et al. (16) found no evidence that a gluten-free diet had any effect on the dermatosis. McNeish et al. (10) reported a case of D.H. developing in a coeliac child while under treatment with a gluten-free diet.

Our patient has now been on a strict gluten-free diet for nearly three years. Her jejunal biopsy is now showing signs of reverting to normal. It is of course impossible to be certain that the patient is keeping to the diet assiduously but the gastroenterologist is convinced that this is the case and the patient is adamant that she has kept to her diet strictly.

We feel this case is further evidence that a gluten-free diet is not always an effective treatment for the skin eruption of D.H.

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