Case report

Monozygous twins concordant for duodenojejunal villous atrophy and dermatitis herpetiformis

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SUMMARY A pair of monozygous twins concordant for both duodenojejunal villous atrophy and dermatitis herpetiformis are described, the first example to be reported in the literature. The observation provides strong support for the concept of an underlying genetic component being involved in the pathogenesis of these two conditions.

Duodenojejunal villous atrophy, morphologically identical to that seen in coeliac disease,¹ is encountered in association with two-thirds of cases of dermatitis herpetiformis.¹ Although the duodenojejunal villous atrophy of dermatitis herpetiformis does respond to a gluten free diet,² however, clinically detectable malabsorption is rare.¹ Although the concordance rate among identical twins for coeliac disease is virtually 100%,³ only a single pair of male monozygotic twins both suffering from clinically diagnosed dermatitis herpetiformis (with no evidence of duodenojejunal villous atrophy) have been reported, and neither brother was subjected to skin or jejunal biopsy.⁴ We therefore wish to report a pair of female monozygous twins with histologically proven dermatitis herpetiformis and duodenojejunal villous atrophy.

Case reports

CASE ONE

A 53 year old Scottish woman presented in 1964 with a 10 year history of an intermittent blistering itchy rash. Dapsone was started with good symptomatic effect. A flare up in 1980 prompted a skin biopsy and granular IgA deposition in the dermal papillae confirmed a diagnosis of dermatitis herpetiformis. In January 1984, she developed diarrhoea and weight loss. Serum and red cell folate levels were subnormal and a small bowel biopsy revealed subtotal villous atrophy, crypt hyperplasia, and an

increased intraepithelial lymphocyte count. Her gastrointestinal symptoms settled on a gluten free diet but she refused a repeat small bowel biopsy.

CASE TWO

The twin sister of case one, who was strikingly similar in physical appearance to her, presented in 1970 with a 14 year history of a recurring pruritic vesicular rash which responded well to dapsone. In May 1985, a skin biopsy revealed granular IgA deposition, concentrated at the dermal papillary tips, and a small bowel biopsy showed severe partial villous atrophy, crypt hyperplasia and an increased intraepithelial lymphocyte count. Serum folate levels were normal. The patient refused a gluten free diet but has remained free of cutaneous symptoms on dapsone. At no point in her illness has she been troubled by gastrointestinal symptoms.

Monozygosity of the sisters was supported by identical blood groups for ABO, MNS, Pi, Rh, Kell, Le^a, Chilano, Fy^a, Fy^b, Jk^a, Jk^b, Co^a and Co^b.

Discussion

This is the first reported pair of monozygous twins in which both individuals have been shown to have both dermatitis herpetiformis and duodenojejunal villous atrophy. The multifactorial aetiology of duodenojejunal villous atrophy without dermatitis herpetiformis – that is, coeliac disease, is generally accepted. As dermatitis herpetiformis is rare, however (with a prevalence of 1:80 000⁴), there have been relatively few well documented family studies,^{4,5} but these studies have appeared to show an increased risk among family members for

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dermatitis herpetiformis. The twins reported here, with their proven monozygosity, provide evidence for a genetic component in the pathogenesis of dermatitis herpetiformis, although their different disease patterns despite their identical genes indicate the importance of environmental factors in the disease.

The relative contributions of genetic factors – for example, HLA status, and environmental agents – for example, dietary gluten, in coeliac disease have been confirmed and investigated using detailed family studies.^{3 5} The situation with respect to the pathogenesis of dermatitis herpetiformis is more complex – the syndrome differs from coeliac disease in that it possesses dermatological features (which are responsive to a gluten free diet⁶) while the bowel lesions cause clinically significant malabsorption much less frequently than is the case with coeliac disease¹: interestingly the duodenojejunal villous atrophy of dermatitis herpetiformis does not respond to the drugs used to treat the skin lesions.¹ HLA associations (similar to those encountered in coeliac disease) and a variety of immunological abnormalities have been described in dermatitis herpetiformis,^{7–10} but despite considerable speculation as to the nature of the underlying immunopathogenesis of dermatitis herpetiformis, several central aspects of the puzzle remain to be resolved.^{7 10} A detailed discussion of the various hypotheses is beyond the scope of a case report, and the reader is referred to more detailed reviews.^{7 10}

Dermatitis herpetiformis is responsible for considerable morbidity and there is a possible relationship to lymphoma and carcinoma,¹¹ thus greater understanding of the pathogenesis of this condition would be highly desirable.

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