Chronic ulcerative jejunitis without symptoms

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Summary

A patient is presented who, in addition to subtotal villous atrophy, had superficial ulcers of the jejunum, as well as a wide band of granulation tissue deep to the crypts suggesting recurrent past ulceration. In spite of these changes she had no intestinal symptoms at the time of her presentation with reflux oesophagitis, and her only nutritional abnormality was a mild folate deficiency. Withdrawal of dietary gluten produced some improvement of the jejunal mucosa but this was not sustained on continued low-grade ingestion. She has continued to be virtually symptom-free over a 3-year period of follow-up.

Introduction

Chronic ulcerative jejunitis (Moritz, Moran and Patterson, 1971) is an uncommon condition not clearly distinguishable from coeliac disease with mucosal ulceration (Jones and Gleeson, 1973). Both are characteristically accompanied by gross steatorrhoea, abdominal pain, weight loss and haemorrhage. Failure of response to a gluten-free diet (GFD) is usual, and surgical intervention may be required for complications, including strictures and perforation. In contrast, a patient is reported who had jejunal villous atrophy and recurrent mucosal erosions who has remained free of significant symptoms for 3 years.

Case report

A 61-year-old housewife presented in April 1975, with heartburn, associated with a hiatus hernia. At the time of her barium meal examination a tubular 'moulage' (Kantor, 1939) small bowel pattern was noted (Fig. 1). She had not travelled abroad and had no relevant family history, but in 1959 she had received liver injections for 'anaemia'. She ate a normal diet and had no diarrhoea, abdominal pains or systemic upset. Her weight was steady and physical examination was normal. The radiological findings prompted further investigations as follows: Hb concentration 12.2 g/dl; MCV 117 fl; serum vitamin B₁₂ 302 ng/l (normal); serum folate 11

μg/l (normal); RBC folate 128 μg/l (normal > 160 μg/l). The following were normal or negative: serum vitamin B₁₂; serum and RBC folate; WBC; ESR; plasma urea and electrolytes; serum calcium; alkaline phosphatase; standard liver function tests; immunoglobulins and autoantibodies. Serum albumin (32 g/l) was low. Xylose absorption was impaired (8.8% of a 25-g load excreted at 5 hr). Daily faecal fat excretion on 70 g of fat/day was high-normal at 19 mmol (5.4 g), and absorption of ⁵⁷Co-vitamin B₁₂ was normal.

Multiple jejunal biopsies taken with a hydraulic tube were grossly abnormal, showing subtotal villous atrophy, crypt hyperplasia, plentiful intra-epithelial lymphocytes, eosinophil and plasma cell infiltration of the lamina propria and mild enterocyte pleomorphism. Surface ulceration was seen in one biopsy (Fig. 2). A striking feature of all these and subsequent biopsies was a wide band of fibrovascular granulation tissue lying between the crypt bases and the muscularis mucosae (Fig. 2).

Initial oral folate therapy (5 mg daily) corrected her macrocytosis, but as a further jejunal biopsy 6 months later was indistinguishable from that of untreated coeliac disease, apart from the fibrovascular band, she was started on a GFD. Stunted villi were present in a biopsy taken 3 months later but, after a further 9 months of imperfect adherence to the diet, multiple biopsies again showed patchy subtotal villous atrophy, and a surface erosion in one biopsy. After 18 further months on a normal diet with folate supplements, the patient's weight is rising and she continues asymptomatic, although she passes occasional loose pale stools. Haematological and biochemical indices remain normal.

Discussion

The benign course of this patient's disorder may relate in part to the relative structural normality of her jejunal enterocytes, and in part to functional sparing of the distal ileum as judged by normal vitamin B₁₂ absorption. The anatomy and distribution of her intestinal lesion, with impaired xylose absorption and folate deficiency, would fit with a diagnosis of adult coeliac disease (Stewart *et al.*, 1967). However, this disease is generally defined in

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FIG. 1. Barium contrast radiograph of the stomach and upper small intestine showing the 'moulage' phenomenon - effacement of the jejunal valvulae conniventes.

terms of morphological response of an abnormal mucosa to a GFD (Booth, 1970). This patient's first follow-up biopsy suggested some improvement over the biopsy preceding gluten withdrawal, but multiple biopsies both previously and subsequently showed patchiness in the severity of villous atrophy, a recently confirmed finding in untreated coeliac disease (Scott and Losowsky, 1976). Since the authors have failed to demonstrate an unequivocal morphological response to gluten withdrawal, they do not feel justified in labelling this patient as having coeliac disease, nor in urging her to return to a GFD in the face of her lack of symptoms. Even if she is considered to have 'unresponsive coeliac disease', it is uncertain that long-term strict adherence to a GFD would reduce her risk, albeit small, of developing intestinal malignancy (Holmes *et al.*, 1976).

The earliest report of erosions in association with

the coeliac lesion does not, regrettably, provide details of the patient's clinical status (Paulley, 1959). Ulceration is now recognized as an occasional feature of coeliac disease, occurring even when the patient has been in remission on a gluten-free diet (Bayless *et al.*, 1967; Jeffries, Steinberg and Sleisenger, 1968; Davidson, 1969; Moritz *et al.*, 1971; Jones and Gleeson, 1973; Klaeveman *et al.*, 1975).

In most of the reported cases, the ulcers are deep and chronic, extending at least to the muscularis propria. They are associated with abdominal pain, melaena, weight loss, severe malabsorption, stricture formation and, frequently, with a rapid downhill course, necessitating surgery. The relationship to ulcerative jejunitis with patchy mucosal atrophy is uncertain (Jeffries *et al.*, 1968; Moritz *et al.*, 1971). In some cases (Clinicopathological Conference, 1972; Klaeveman *et al.*, 1975), ulceration may be an accompaniment of coeliac disease which has become

FIG. 2. Section of initial jejunal biopsy demonstrating villous atrophy, crypt hypertrophy and a superficial mucosal ulcer. A wide band of fibrovascular granulation tissue lies between the crypt bases and the muscularis mucosae (HE, $\times 30$).

unresponsive to a gluten-free diet, and has been considered as heralding the development of frank intestinal lymphoma (Whitehead, 1973). The mucosal fibrovascular band described here has not been reported previously either in coeliac disease, ulcerative jejunitis or in other small intestinal disorders. If it represents a reaction to recurrent past ulceration (H. Thompson, personal communication), then the 2 erosions biopsied here may be examples of a long-standing process. In this instance, however, jejunal ulceration has not led to the severe illness of which such lesions are assumed to be hallmarks.

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CORRIGENDUM

Postgraduate Medical Journal
March 1979 Vol. 55 No. 641

Article by S.P. Kane and R. A. Parkins: Chronic ulcerative jejunitis without symptoms.

Page 215, paragraph 1 of right hand column, lines 2 and 3:

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