A further case of chronic ulcerative enteritis

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SUMMARY A further case of chronic ulcerative enteritis is presented. In this case there was clear evidence that the ulcerative process was superimposed upon atrophic jejunitis with malabsorption. The development of ulceration produced a picture of small bowel obstruction and its differential diagnosis from neoplasm was not possible without laparotomy.

Chronic ulcerative enteritis is a rare disease. Although it was first described in 1949, a review in 1971 by Moritz, Moran, and Patterson included only 25 cases. The majority of these were from North America, but some have been reported from Australia and Europe (Goulston, Skyring, and McGovern, 1965; Davidson, 1969).

The disease is characterized by multiple chronic benign ulcers of the small bowel, most frequently the jejunum, with resultant colic, fever, diarrhoea, and later progression to intestinal strictures. It is closely associated with intestinal malabsorption and atrophic jejunitis, and there is debate whether the ulceration precedes or follows this lesion. Most authors favour the latter possibility.

This paper reports a further case of chronic idiopathic ulcerative enteritis because of the rarity of this disease and the problems of differential diagnosis and management which it presented. In this instance there is clear evidence that malabsorption was present before the development of intestinal ulceration.

Case Report

A 58-year-old Englishwoman presented to a surgical unit at another hospital in July 1968. For 20 years she had had epigastric pain and flatulence, followed by progressive lassitude and diarrhoea for seven years. Her presentation followed anorexia and weight loss (54 to 43 kg) for three months, and nausea, vomiting, and colicky lower abdominal pain for two months. There was no significant previous history except for infantile rectal prolapse at 12 months of age.

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time. No macroscopic abnormality was apparent. However, after this the diarrhoea ceased and her weight increased to 43 kg. She remained in fair health on a gluten-free diet for 18 months.

Further symptoms developed in May 1971, with the onset of anorexia, postprandial lower abdominal colic, and loss of 9 kg weight but without a recurrence of diarrhoea. Relevant investigations included ESR, 33 mm drop in one hour; d-xylose absorption (25 g), 1·3 g in five-hour urine; faecal fat excretion (three days), 7·9 g per day. Barium studies of the small bowel using intravenous metoclopramide showed dilatation of the proximal jejunum with an obstructing stricture approximately 90 cm from the duodenojejunal flexure (fig 1).

A further laparotomy was therefore undertaken in July 1971. There were multiple, irregular, tumour-like thickenings of the small bowel, extending from 25 cm distal to the duodenojejunal flexure to within 100 cm of the ileo-caecal valve. The largest and most proximal of these was the site of an incomplete small bowel obstruction. A macroscopic diagnosis of malignant obstruction was made, and the obstruction bypassed by entero-anastomosis, biopsies being taken from the suspected tumours and jejunal and ileal mucosa. Examination of these did not show neoplastic tissue, so a resection of 62 cm of small bowel, including the obstructing lesions, with end-to-end anastomosis was performed a few days later. The postoperative course was uneventful.

Examination of the resected specimen showed two large deep circumferential ulcers 4 cm and 3 cm in diameter, as well as 13 other shallow ulcers distributed in irregular fashion along the mucosa (fig 2). The latter ranged in size from 3 cm to 3 mm in diameter. The mesentery was thickened and oedematous. The regional mesenteric lymph nodes were enlarged to 3 cm diameter and soft in consistency.

Histological examination showed a necrotizing ulceration extending in depth for a variable distance and in some instances involving the muscle coat (fig 3).

The fibrinopurulent exudate on the surface contained a mixture of Gram-positive cocci and bacilli but acid-fast organisms and fungi were not found. Fibrinoid change was prominent in the walls of small blood vessels in the necrotic parts of the bowel and there was a diffuse mixed inflammatory infiltrate composed of neutrophil and eosinophil leucocytes, plasma cells, and lymphocytes. The submucosa adjacent to the ulcers was thickened by fibroplastic connective tissue; scar tissue replacement of portion of the muscle coat was evidence of a chronic disease process. The mesenteric lymph nodes showed non-specific inflammation and reactive changes. Adjacent to one of the large ulcers there was a focus of necrotic mesenteric fat containing obliterated arteries.

Following discharge from hospital the patient was initially well and gained some weight. However, pain and vomiting later recurred despite a gluten-
free diet and antibiotic therapy. She became depressed and was admitted to a psychiatric hospital. She died in October 1971 during insulin-shock therapy. Postmortem examination was not obtained.

Discussion

This case meets fully the clinical, radiological and histological criteria for the diagnosis of chronic ulcerative enteritis. There is clear evidence that atrophic jejunitis with malabsorption was present for some years before the onset of intestinal ulceration. This is shown by the history, initial biochemical, radiological, and biopsy findings, and the macroscopic appearances at the first laparotomy in July 1968. This appears to be the usual sequence, although some cases have come to medical attention only with the onset of symptoms due to the presence of jejunal ulcers.

There is no satisfactory explanation of the development of intestinal ulceration in some patients with atrophic jejunitis.

It does not seem to be secondary to vasculitis. The arteritis observed adjacent to the ulceration is probably a secondary effect, and there are no reports of vascular disease in other parts of the body.

Ulceration secondary to a chronic bacterial infection is a further possibility, and bacteria were observed in the ulcerated areas of mucosa in the present case. But this hypothesis does not explain facts such as the chronicity of the disease and clinical and histological differences from known bacterial ulcerative diseases of the small intestine due to organisms such as Salmonellae, Clostridia, and Mycobacterium tuberculosis.

The possibility of an ingested toxin is suggested by distribution of the ulcers. These are in most cases larger and more numerous in the proximal intestine and often absent from the ileum, so that many cases have been reported as ulcerative jejunitis rather than ulcerative enteritis. However, no such toxin has been identified. Some preparations of potassium chloride are known to cause intestinal ulceration (Baker, Schrader, and Hitchcock, 1964), but these have not been mentioned in most reports of patients with ulcerative enteritis. The present patient did receive supplements of potassium chloride, but at laparotomy after cessation of this therapy there were no signs of ulceration. Nevertheless, the possibility of an allergic reaction to some ingested agent seems the least unlikely explanation of the distribution and pathology of the lesions and their occurrence in patients with gluten enteropathy.

The clinical problem presented by patients with chronic ulcerative enteritis is that of clinical deterioration with subacute intestinal obstruction in a patient with atrophic jejunitis. The differential diagnosis is from intestinal neoplasm, since this is also a recognized late complication of atrophic jejunitis (Joske, 1960; Read, 1970). There seems no
way to resolve this without laparotomy. Differentiation may be difficult even on macroscopic inspection of the gut, as in the present case, so that the surgeon must obtain adequate material for histological examination.

The treatment of this disease is unknown, but in most recorded cases the prognosis is poor. No certain benefits appear to derive from steroid or antibiotic therapy, but the rarity of the condition makes definitive conclusions impossible. Surgery is indicated both for diagnosis and treatment of obstructing lesions.

References

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