Reticulosis of the Small Bowel as a Late Complication of Idiopathic Steatorrhœa

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(for R D Tonkin MD FRCP)

About 10% of all malignant lymphomata arise in the gastrointestinal tract (Skrimshire 1955). Of the cases of primary lymphoma of gut, approximately half involve the small intestine primarily (Skrimshire 1955, Dawson *et al.* 1960). In Parkhurst's series of 20 cases, however, only two arose in the small intestine (Parkhurst & MacMillan 1962).

The association between intestinal reticulosis and steatorrhœa has been known for at least twenty-five years (Fairley & Mackie 1937). Until recently, it has been assumed that the steatorrhœa has always been secondary to the reticulosis and Best & Cook (1961), from an analysis of the literature, concluded that between 5% and 10% of adult cases of apparently idiopathic steatorrhœa were secondary to lymphomata of small bowel or lymph nodes. Gough *et al.* (1962) were the first to suggest that reticulosis of the small bowel may occur, in some cases, as a late complication of idiopathic steatorrhœa, cœliac disease or tropical sprue. The case described here would appear to supply further evidence in support of their view.

Case History

Female, aged 31. Housewife. At the age of 14 months a well-substantiated diagnosis of cœliac disease was made. There was a satisfactory response to an unspecified diet. Subsequently she remained well, apart from occasional bouts of diarrhœa, until January 1962 when she began to pass ten to fifteen pale and offensive stools per day; by June she had lost 1 stone (6·3 kg) in weight.

On admission: She was noted to be of small build, thin and pale. Her skin was lightly pigmented and an ill-defined mass was palpable in the left upper quadrant of the abdomen. Rectal and sigmoidoscopic examinations were normal.

Investigations: Hæmoglobin 82%; W.B.C. 6,200; E.S.R. 42 mm in first hour (Wintrobe). Red cells showed generalized macrocytosis. Serum calcium 8·5, phosphate 4·1 mg/100 ml; albumin 2·7, globulin 3·8 g%. Formiminoglutamic acid test strongly positive. Gastrografin meal and follow through was inconclusive, although the bowel pattern in the jejunum was irregular. Peroral jejunal biopsy showed gross villous atrophy.

Course in hospital: The patient ran a high remittent temperature and had the symptoms of subacute small bowel obstruction. An emergency laparotomy was dictated by a massive hæmatemesis and severe abdominal pain. At operation the jejunum was found to be obstructed by a small ulcerating tumour. Histologically, this was a reticulum cell sarcoma (Fig 1). The lymph nodes were not involved. Sections of the macroscopically normal bowel showed blunting and occasional absence of villi (Fig 2).

Discussion: The length of history and the small area of bowel involved appear to rule out the reticulum cell sarcoma as the cause of the steatorrhœa. The childhood history of cœliac disease and the characteristic histological appearance of the jejunal biopsy are further points in favour of idiopathic as opposed to secondary steatorrhœa. While it is impossible to exclude a fortuitous association, it is at least likely that the reticulosis has developed as a late complication of idiopathic steatorrhœa. This latter condition,

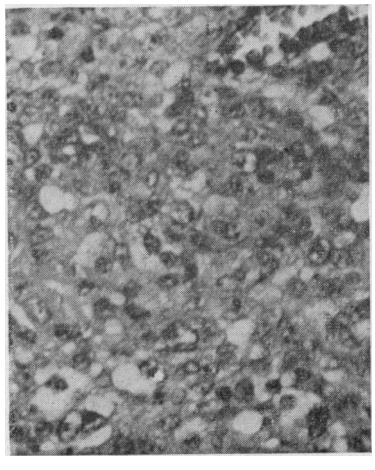

Fig 1 *Histological section of small bowel tumour – a reticulum cell sarcoma*

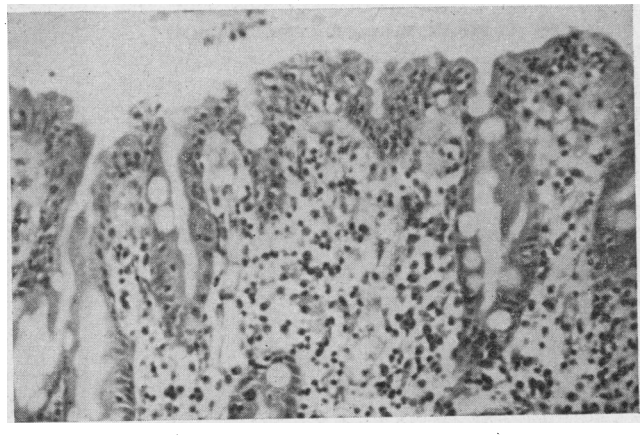

Fig 2 *Histological section of jejunal mucosa showing gross blunting of villi*

with its widespread mucosal abnormality, might reasonably be expected to be a premalignant condition.

Features in this case which suggested the development of a reticulosis were the persistent pyrexia, high E.S.R. and evidence of subacute intestinal obstruction. Gough mentions two further points; namely, the failure to respond to a previously successful diet and dermatological manifestations of the reticulosis (Gough *et al.* 1962). The presence of any of these features in a patient with idiopathic steatorrhœa should raise the question of early laparotomy.

Where a reticulosis develops as a complication of idiopathic steatorrhœa, the jejunum is involved almost as often as the ileum, in contrast to isolated small bowel lymphomata which are commoner in the ileum. Although all types of reticulosis have been described complicating idiopathic steatorrhœa, all 5 of Gough's own cases were reticulum cell sarcomas, as in our case, and he comments on the striking histological similarity of the tumours (Gough *et al.* 1962).

The natural course of primary lymphomata of the gut is variable. Skrimshire (1955) gives a five-year survival of 10%, Allen *et al.* (1954) a five-year survival of 43% and Parkhurst & MacMillan (1962) found an average survival of three to four years. With treatment, and sometimes without, remarkably long remissions, and perhaps cures, may occur (*see* Skrimshire 1955 for references). Of the 22 cases of small bowel reticulosis associated with idiopathic steatorrhœa (Gough *et al.* 1962) the average duration of life after the diagnosis of steatorrhœa in 15 cases was twenty-five months.

A combination of surgery and radiotherapy or chemotherapy is generally thought to be the treatment of choice for small bowel reticulosis unassociated with idiopathic steatorrhœa. Dawson *et al.* (1960), however, concluded that post-operative radiotherapy did not materially alter the prognosis. In addition, in the cases secondary to idiopathic steatorrhœa, there are the theoretical risks of irradiation aggravating the steatorrhœa (Reeves *et al.* 1959) and, if Gough's hypothesis is correct (Gough *et al.* 1962), of encouraging further malignant change.

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Congenital Intraduodenal Diverticulum

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R C, male, aged 24 years

History: Born 15.11.37. Vomited after breast feeding the next day. Vomitus became yellow. He was seen on 5th day of life when barium feeding confirmed the diagnosis of duodenal obstruction, which was thought at the time to be due to complete duodenal atresia.

At operation the following day anterior gastro-enterostomy was performed (D L). The post-operative course was extremely stormy. A gastrostomy was performed and a tube inserted through the stoma into the jejunum. Leakage of bile-stained juice occurred around the tube with digestion of the abdominal wall. His condition slowly improved and the fistula closed without further operation.

March 1938: Shown at the Royal Society of Medicine by Dr K H Tallerman and Mr David Levi, who found, in the British and American literature at that time, records of only 6 other cases successfully operated on.

He was then well except for prominent abdominal rumblings without definite dyspepsia until February 1961, when he had the first of three episodes of melæna. Three barium meals were carried out at two different teaching hospitals but the characteristic appearances were not recognized. March 1962: After his third melæna, barium meal (L K) revealed a large diaphragm pouched down the dilated duodenum (Fig 1), forming an intraduodenal diverticulum.

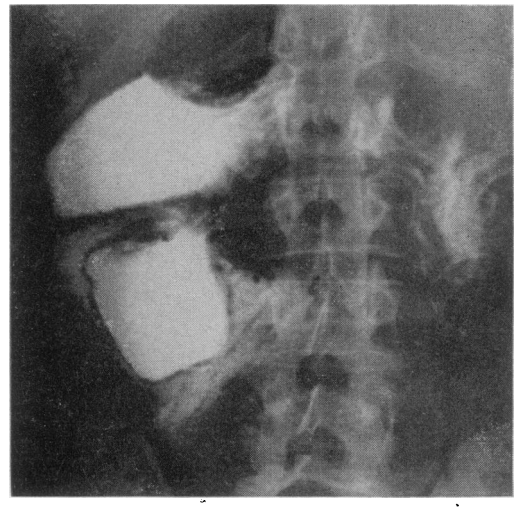

Fig 1 *Barium meal showing the intraduodenal diverticulum*