

## Case 63

# An abdominal mass in a young man

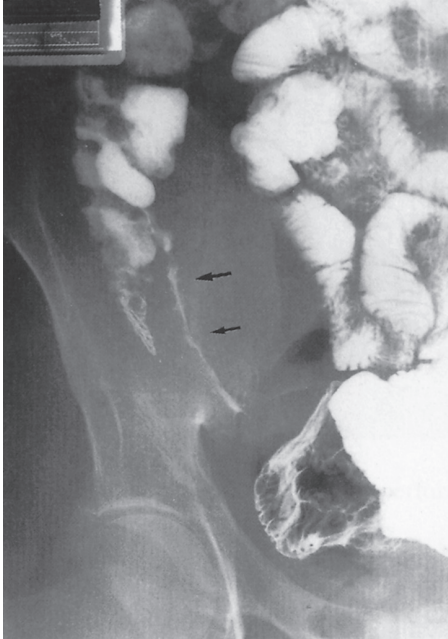


Figure 63.1

A trainee schoolmaster aged 25 years was referred to the gastroenterological medical outpatient clinic by his family practitioner with an urgent appointment. He gave a history of 6 months of attacks of colicky pain in the right iliac fossa, which were accompanied by diarrhoea. The bowels would act up to three or four times a day and would be loose, even watery, but he had never noticed any blood. The attacks were getting more frequent and more intense, and in the last couple of weeks he was in almost constant pain or discomfort and was quite unable to work. His appetite was now very poor and he thought he had lost about 3 kg in weight.

When examined in the clinic he looked ill and anaemic. He was afebrile and his blood pressure was 134/70. Examination of the abdomen revealed a tender mass in the

right iliac fossa; there was no abdominal distension or clinical evidence of ascites. Rectal examination was negative, and there was no evidence of perianal disease.

He was admitted urgently to the unit. A battery of investigations was carried out, including a barium meal and follow-through examination. Figure 63.1 shows the long strictured segment of terminal ileum (arrowed).

### What is the name given to this appearance, and what disease characteristically produces this change?

The 'string sign' of Kantor.\* This, together with the clinical picture, strongly suggests the diagnosis of Crohn's disease.†

### What other radiological features may be seen in Crohn's disease?

There may be fine ulceration, which may give the so-called 'rose thorn' appearance of the mucosa, and this can be seen on careful inspection of Fig. 63.1. Several segments of stricturing may be seen, separated by apparently normal bowel ('skip lesions'), and fistulous tracks may be demonstrated into other structures – an adjacent loop of small bowel or large bowel, for example.

### What radiological test has replaced a barium meal in the diagnosis of Crohn's disease?

Magnetic resonance enterography, which demonstrates mural thickening in areas of affected bowel; in cross section this circumferential thickening gives the appearance of a target. MR is also useful in delineating fistulae-in-ano in Crohn's disease.

\*John Leonard Kantor (1890–1947), gastroenterologist, Presbyterian Hospital, New York.

†Burrill Bernard Crohn (1884–1983), gastroenterologist, Mount Sinai Hospital, New York. The condition was described by Morgagni (1680–1771).



**Figure 63.2** Cobblestoning and linear mucosal ulceration in Crohn's disease.

### Can Crohn's disease affect other parts of the alimentary tract?

Although the terminal ileum is the commonest site for Crohn's disease – hence the common but scientifically inaccurate name of regional ileitis – any part of the alimentary tract from mouth to anal canal may be involved, including the perianal skin. It is associated with severe multiple anal fissures and fistulae (see Cases 75 and 77, pp. 150 and 154).

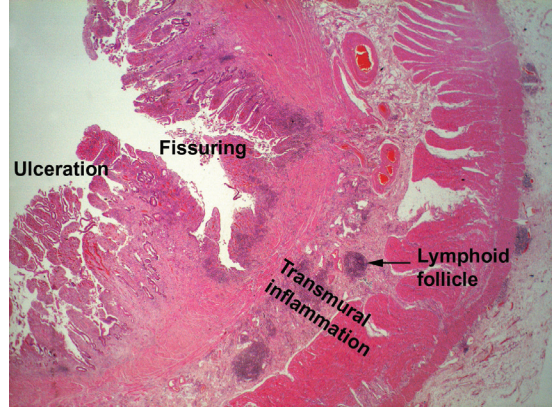
### What are the typical naked eye and histological appearances of the bowel in this disease?

The bowel is bright red and swollen. Mucosal ulceration, which tends to be in a linear direction, and intervening oedema results in a 'cobblestone' appearance of the mucous membrane (Fig. 63.2). The wall of the intestine is greatly thickened, as is the adjacent mesentery, and the regional lymph nodes are enlarged. There may be skip areas of normal intestine between involved segments and there may be fistulae into adjacent structures.

Microscopic examination shows chronic inflammatory infiltrate through the whole thickness of the bowel with non-caseating foci of epithelioid and giant multinucleate cells. Ulceration is present, with typical fissuring ulcers extending deep through the mucosa (Fig. 63.3). These may extend through the bowel wall to form abscesses or may fistulate into adjacent structures.

### What is known about the aetiology of this disease?

The simple answer is not all that much! There is a genetic component, in that 20% of patients with Crohn's disease have an affected relative. Recently an association has been described with a genetic mutation in the NOD gene family – these are genes involved in the innate immune



**Figure 63.3** Histology of Crohn's disease.

response to bacterial antigens within the gut. *Mycobacterium avium* subspecies *paratuberculosis* has also been suggested as a cause, and is known to affect immune signalling. In reality it may be a synthesis of many of these theories, with impaired innate immunity, both genetically and environmentally acquired, affecting the host response to otherwise innocuous gut flora.

### What would be your initial non-operative management?

The initial management of an acute Crohn's ileitis involves anti-inflammatory treatment with oral corticosteroids, such as prednisolone (0.5–1 mg/kg/day). As the disease responds, elemental diet is introduced to maintain the remission. This is a liquid diet comprising protein, in the form of amino acids or short chain peptides, as well as carbohydrate, fats, vitamins and minerals. If the ileitis is not severe, elemental diet alone, without oral steroids, may enable disease remission and long-term control of symptoms.

### What would be the indication for surgery in this man?

The most likely indication for surgery in this man would be intestinal obstruction secondary to the stricturing. The underlying aim when operating on Crohn's disease is to avoid resecting more bowel than necessary. Many patients may require multiple operations and bowel resections, resulting in short gut syndrome and a dependence on total parenteral nutrition.

There are two main procedures

- For short strictures, the stricture is opened longitudinally and closed transversely, so-called 'strictureplasty'.
- For longer strictures, the affected bowel is resected with an end-to-end anastomosis of the two ends.