Mesenteric arteritis

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Abstract

Four patients are presented with small bowel infarction secondary to a vascular arthritis. In three patients there was a history of rheumatoid arthritis. In each patient infarcted bowel was resected and a primary anastomosis performed. In one patient the anastomosis broke down and she subsequently died. One patient died from a disseminated rectal tumour three years later. The remaining patients remain well. If operated on early, intestinal infarction due to arteritis has a good prognosis.

Acute intestinal infarction is a surgical emergency. Except in patients with mechanical bowel obstruction, it is caused by arterial or venous occlusion, or a vasculitis, or may arise in a low flow state when mesenteric perfusion is inadequate.

The precise cause is difficult to define, even at laparotomy, which accounts for the variation in incidence reported. Nevertheless, the consensus indicates that superior mesenteric arterial occlusion is responsible for most cases of intestinal ischaemia, while mesenteric venous thrombosis and non-occlusive ischaemia together represent 30% of cases. The rarest cause of intestinal ischaemia is due to mesenteric vasculitis. Early recognition and treatment is important as intestinal infarction may progress unless the involved bowel is resected. We report the clinical features and outcome for four adults with small bowel infarction secondary to vasculitis.

Patients

A short history of non-specific abdominal pain and lack of clear physical signs was characteristic of each patient (Table). Three of the patients had a history of severe rheumatoid arthritis and were taking 5–10 mg prednisolone/day on presentation. The white cell count was raised in all four patients. The decision to operate was taken on clinical grounds alone; arteriography was not performed on any patient.

At operation there was clear evidence of ischaemic damage which was localised in all except the patient in case 1, who had many short gangrenous areas of small bowel extending from proximal jejunum to mid-ileum. In all cases the large bowel was normal. The superior mesenteric artery and its principal branches appeared normal. The necrotic segment of bowel was resected in each case and a primary anastomosis performed.

The histology in the four cases indicated that there was full thickness ischaemic necrosis of the bowel. There was an acute arteritis both in the affected bowel itself and in the adjacent mesenteric vessels which were thrombosed. There were prominent vascular channels lined by plump endothelial cells. The surrounding stoma of the vascular channels was infiltrated by a cuff of acute inflammatory infiltrate with oedema (see Figure). In addition, the patient in case 3 had fibrinous necrosis of the vessel walls and aneurysmal dilatation of the wall consistent with polyarteritis nodosa.

In the patient in case 1 a small bowel fistula developed which had not shown evidence of closing after six weeks’ intravenous feeding. In view of the patient’s age and severe rheumatoid disease, which prevented her changing the drainage bags, it was decided not to embark on longterm parenteral nutrition.

Discussion

Mesenteric vasculitis is a rare cause of intestinal infarction representing about 2% of cases. Indeed, this presentation is the largest reported series.

Intestinal vasculitis represents a group of clinical features of four patients with superior mesenteric vasculitis

<table>
<thead>
<tr>
<th></th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td>65</td>
<td>67</td>
<td>74</td>
<td>49</td>
</tr>
<tr>
<td>Sex</td>
<td>Female</td>
<td>Male</td>
<td>Male</td>
<td>Male</td>
</tr>
<tr>
<td>Relevant medical</td>
<td>Rheumatoid arthritis for 20 years</td>
<td>Rheumatoid arthritis for five years</td>
<td>Rheumatoid arthritis for 10 years</td>
<td>Rheumatoid arthritis for 10 years</td>
</tr>
<tr>
<td>Symptoms</td>
<td>One week severe diffuse abdominal pain</td>
<td>One week abdominal pain and vomiting</td>
<td>Two days’ abdominal pain</td>
<td>Two days’ abdominal pain</td>
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<tr>
<td>Signs &amp; Serology</td>
<td>Generalised tenderness</td>
<td>Generalised tenderness</td>
<td>Necrotic skin lesions</td>
<td>Necrotic skin lesions</td>
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<tr>
<td>Operation</td>
<td>Multiple segments of gangrenous small bowel</td>
<td>Infarcted 90 cm ileum resected.</td>
<td>Infarcted 150 cm small bowel</td>
<td>Infarcted 150 cm small bowel</td>
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<td>Outcome</td>
<td>Anastomotic dehiscence. Started on parenteral nutrition but subsequently abandoned. Died eight weeks</td>
<td>Well at two years</td>
<td>Well at three years then presented with disseminated rectal tumour</td>
<td>Well at one year</td>
</tr>
</tbody>
</table>

RIF=right iliac fossa.
Mesenteric arteritis

Histological appearance of mesenteric vasculitis, showing loss of columnar epithelial lining of the bowel, thrombosis of vessels, and inflammation around involved vessels.

disorders sharing the histological features of inflammation and necrosis of blood vessels. Classification is based on a combination of clinical, radiological, and pathological features. The intestinal involvement may occur in isolation (as in the patient in case 4) or multiple systems may be involved.

A variety of different vasculitic complications have been implicated as the cause of intestinal ischaemia. The reported diagnostic categories have been rheumatoid arteritis. This is generally regarded as a rare cause of intestinal infarction, though it was probably responsible for three cases in this series. Scleroderma has been described and the patient in case 4 had certain symptoms that may indicate this as an underlying abnormality. Polyanteritis nodosa may rarely cause intestinal ischaemia and infarction, and the patient in case 3 had histological evidence of fibrinoid necrosis of the vessel wall which would be consistent with polyarteritis nodosa. In addition, he had necrotic areas of skin.

In those cases with an isolated mesenteric vasculitis it is often impossible to identify a specific clinical syndrome and such patients have been described as having an atypical collagen vasculitis. The patient in case 4 would probably be regarded in this category. Indeed, difficulties arise in the classification of arteritis.

There is often considerable overlap between the different syndromes which will not be resolved until the pathogenesis of vasculitic disease is better understood.

Should a patient present with vasculitis it may be possible to prevent the development of mesenteric vasculitis by aggressive treatment with cyclophosphamide.

Preoperative diagnosis of intestinal ischaemia is notoriously difficult. It has been found that a high white cell count is frequently found in this condition. Interestingly, the expected leucocytosis was present in these patients, three of whom were taking prednisolone.

In all cases the infarcted bowel was resected and a primary anastomosis performed. In the patient in case 1, however, this subsequently broke down and ultimately was responsible for her death. It may be important that in the three patients who did well there was an isolated ischaemic segment. The patient in case 1 had many separated areas of necrosis and about three quarters of her small bowel had to be resected which had both necrotic and viable segments. In the patient in case 1 the vasculitis may have involved the suture line, and in retrospect we would not recommend primary anastomosis for anyone with diffuse small bowel involvement.

Mesenteric vasculitis causing bowel ischaemia is a rare condition; nevertheless, if operated on promptly it has a prognosis considerably better than that in patients with infarction due to superior mesenteric artery occlusion. Indeed, three of the patients made a good postoperative recovery. We therefore urge those responsible for the management of patients with abdominal symptoms and a connective tissue disorder to have a high index of suspicion as prompt operative intervention can be life saving.

We thank Dr D R Swinson for his advice.