

CASE REPORT

Superior mesenteric vein stenosis complicating Crohn's disease

R S Hodgson, J E Jackson, S D Taylor-Robinson, J R F Walters

Abstract

Background—Superior mesenteric vein stenosis as a consequence of mesenteric fibrosis, causing the development of small bowel varices, is an unrecognised association of Crohn's disease.

Case reports—Two cases of gastrointestinal bleeding occurring in patients with Crohn's disease, and a third case, presenting with pain and diarrhoea, are described. In all three patients, visceral angiography showed superior mesenteric vein stenosis with dilatation of draining collateral veins in the small bowel. Overt gastrointestinal bleeding or iron deficiency anaemia resulting from mucosal ulceration is common in Crohn's disease, but acute or chronic bleeding from small bowel varices as a result of superior mesenteric vein stenosis due to fibrosis has not previously been reported.

(Gut 1999;45:459–462)

Keywords: Crohn's disease; superior mesenteric vein stenosis; mesenteric fibrosis; small bowel varices

Bleeding from the gastrointestinal tract is a common presenting symptom or complication of Crohn's disease.^{1–3} Acute or chronic blood loss results from mucosal ulceration. Vasculitis involving small vessels is a frequent finding in affected segments of gut,⁴ but involvement of larger blood vessels such as the superior mesenteric vein is reported rarely.⁵ The aetiology in the described cases is venous thrombosis caused by a procoagulant tendency. Peripheral venous thrombosis and pulmonary embolism occur most commonly, but superior mesenteric vein thrombosis or portal vein thrombosis causing recurrent gastrointestinal bleeding from varices has also been described. Occlusion of the superior mesenteric vein caused by a fibrotic reaction has not been described.

We describe three patients with Crohn's disease who were found to have superior mesenteric vein stenosis as a consequence of mesenteric fibrosis and small bowel varices at visceral angiography. One patient presented with abdominal pain, another with iron deficiency anaemia, but in the third case, frank variceal bleeding from the jejunum was the predominant symptom.

Case 1

A 34 year old man with a 21 year history of Crohn's disease was referred to this hospital for visceral angiography and consideration of embolisation of a reported arteriovenous malformation in the small intestine.

The patient had had predominantly inflammatory small intestinal disease causing relapses of right iliac fossa pain, nausea, and vomiting which was successfully treated with courses of oral prednisolone and sulphasalazine or mesalazine for the first 17 years of his disease. He then presented with massive rectal bleeding requiring blood transfusion. Exploratory laparotomy revealed extensive inflammatory Crohn's disease involving the entire small intestine. The small intestine was oedematous, ulcerated, and thick walled, and was noted to be encroaching on the mesentery. A "haemangioma" at the duodenojejunal junction was resected, and an inflamed caecum adherent to the posterior abdominal wall was found. The colon was noted to be full of blood.

A limited right hemicolectomy to the hepatic flexure was performed and 20 cm of affected terminal ileum was resected to remove the suspected source of blood loss. The mesentery was noted to be oedematous and it bled easily. Macroscopic examination of the resected specimens revealed oedematous and fibrosed small and large bowel with loss of the normal mucosal pattern and numerous ulcers which caused a cobblestone appearance. Microscopically there was a moderate chronic inflammatory infiltrate and numerous fissures and ulcers with granuloma formation, which were consistent with the diagnosis of Crohn's disease. During the subsequent three years the disease was difficult to control, requiring treatment with oral prednisolone 10–30 mg/day and azathioprine 150 mg/day. Over the next six months the patient presented with severe rectal haemorrhage on six further occasions necessitating blood transfusion of 20 units in total. Barium meal and follow through examination showed inflammation involving the jejunum in a pattern typical of Crohn's disease. Visceral angiography performed at the referring hospital was thought to show a vascular malformation in the proximal jejunum and a second, smaller lesion in the terminal ileum. He was then referred to this hospital for repeat visceral

Gastroenterology Unit,
Department of
Medicine, Imperial
College School of
Medicine, London, UK
R S Hodgson
S D Taylor-Robinson
J R F Walters

Department of
Diagnostic Radiology,
Imperial College
School of Medicine,
London, UK
J E Jackson

Correspondence to:
Dr J E Jackson, Department
of Diagnostic Radiology,
Imperial College School of
Medicine, Hammersmith
Hospital, Du Cane Road,
London W12 0NN, UK.

Accepted for publication
5 March 1999

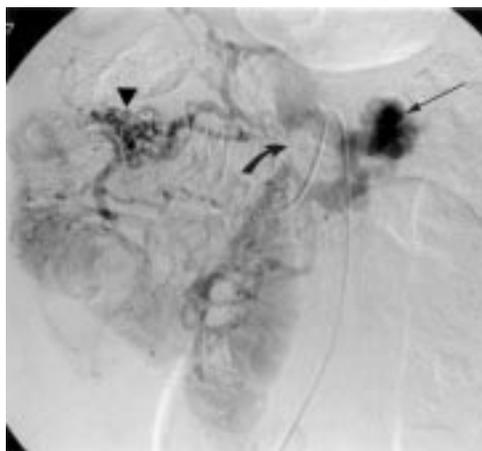


Figure 1 Case 1: venous phase from cannulation of the superior mesenteric artery shows a tight stenosis of the main superior mesenteric vein (long curved arrow) with numerous surrounding small bowel varices most prominent within the jejunum (straight arrow). The stenosis is regular and concentric with tapered shoulders strongly suggesting a fibrotic aetiology as opposed to recanalisation following venous occlusion by thrombus. Varices are also seen crossing the ileocolic anastomosis (arrowhead).

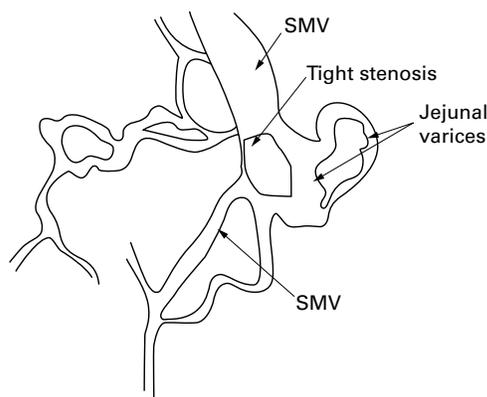


Figure 2 Diagram illustrating the stenosis present in the angiographic picture shown in fig 1.

angiography and consideration of embolisation of the vascular lesions.

The repeat visceral angiogram revealed an extremely tight stenosis of the superior mesenteric vein and notable dilatation of the collateral jejunal veins (figs 1 and 2). He was commenced on propranolol to reduce the splanchnic vascular pressure, and has not had a further episode of rectal bleeding for 10 months.

Case 2

A 23 year old woman with a two year history of occult gastrointestinal blood loss of uncertain aetiology was referred to this hospital for visceral angiography.

The patient had experienced cramping upper abdominal pain and lethargy lasting several weeks on two occasions, but was otherwise asymptomatic. She had an iron deficiency anaemia and was dependent on oral iron supplementation. Her menstrual cycle was regular with normal blood loss. Physical examination was unremarkable, but multiple faecal occult blood tests were positive. Upper gastrointestinal endoscopy, colonoscopy, barium

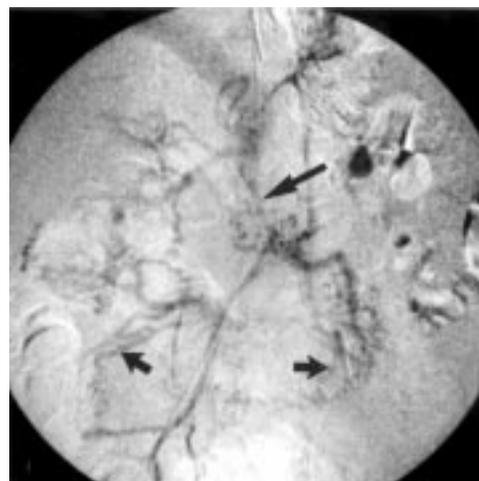


Figure 3 Case 2: venous phase from the main superior mesenteric arteriogram shows a tight stenosis of the superior mesenteric vein (long arrow) and resultant small bowel varices (short arrows).

meal and follow through examination, and technetium-99m pertechnetate scintigraphy for Meckel's diverticulum performed at the referring hospital were reported as normal.

Visceral angiography performed at this hospital showed a stenosis of the superior mesenteric vein with resultant dilatation of collateral veins in the jejunum and ileum (fig 3). The ileal branches of the superior mesenteric vein were noted to be stretched around several loops of dilated ileum. A small bowel enema was therefore performed. This allowed the mucosa to be distended to maximum advantage as a result of the enteroclysis procedure in order to highlight small bowel pathology. This revealed multiple tight strictures in the ileum accompanied by proximal dilatation of the ileal loops, with no evidence of mucosal ulceration or fistula formation. Laparotomy confirmed the presence of multiple ileal strictures with a normal colon. Two segments of small intestine were resected and stricturoplasty was performed on a further narrowed segment. Histology of the resected specimens revealed an active, patchy, chronic inflammatory bowel disease featuring mucosal ulceration and fissuring, full thickness inflammation of the bowel wall including submucosal, intramural, and serosal lymphoid aggregates, and the presence of granulomas which were consistent with a diagnosis of Crohn's disease. The patient was subsequently treated with oral prednisolone and mesalazine. She recovered well from the operation and is being followed up at the referring hospital.

Case 3

A 63 year old man presented to this hospital with abdominal pain and diarrhoea. His past medical history included two myocardial infarctions, bilateral carotid endarterectomies, and a sigmoid colectomy performed for severe diverticular disease. There were no features indicating a diagnosis of Crohn's disease in the history or examination. Blood tests performed included a haemoglobin of 125 g/l, with normal mean corpuscular volume, a normal white cell

count and platelet count, an erythrocyte sedimentation rate of 36 mm in the first hour, C reactive protein 22 mg/l (normal range 0–10 mg/l), and albumin 34 g/l. A plain abdominal radiograph did not show any abnormal features. A visceral angiogram was performed to exclude mesenteric ischaemia. This revealed a stenosis of the superior mesenteric vein with dilatation of collateral veins in the jejunum and ileum. A barium meal and follow through examination showed multiple long strictures affecting the terminal ileum. Terminal ileal inflammation was confirmed by radiolabelled ^{99m}Tc white cell scintigraphy. A diagnosis of Crohn's disease was made and he was commenced on treatment with oral prednisolone and sulphasalazine. His symptoms resolved and a repeat barium meal and follow through examination showed no progression of the strictures. His disease is now well controlled with oral prednisolone and azathioprine.

Discussion

Diarrhoea, abdominal pain, and gastrointestinal bleeding are among the presenting symptoms of Crohn's disease originally described by Crohn *et al* in 1932.¹ Approximately 45% of patients with Crohn's disease will have overt rectal bleeding during the course of their illness,^{2,3} and although profuse rectal bleeding is not mentioned in the original description of the disease, it occurs in about 1 in 20 cases.^{6,7} Life threatening haemorrhage has a lower prevalence of 1.3–1.4%.^{8,9} This was first described in Crohn's disease in a case report by Fallis in 1941.¹⁰ Rubin *et al* subsequently described seven cases of Crohn's disease where acute, severe bleeding due to extensive mucosal ulceration was the presenting feature.⁷ Superior mesenteric vein thrombosis or portal vein thrombosis as a result of a procoagulant tendency causing recurrent gastrointestinal bleeding from varices has been described.⁵ However, occlusion of the superior mesenteric vein caused by a fibrotic reaction has not previously been described. Case 1, which we present, is the first report of overt bleeding caused by small intestinal varices due to mesenteric vein stenosis as a consequence of fibrosis in Crohn's disease.

In Crohn's disease, angiography of the small intestine is abnormal in over 90% of cases. Characteristic features include proliferation, dilatation, and tortuosity of the blood vessels and early venous return. Extension of these abnormalities into the adjacent mesentery due to associated mesenteric fibrosis is said to be diagnostic of Crohn's disease.¹¹ The vascular changes may occasionally be so notable that the appearances are mistaken for those of an arteriovenous malformation. This has previously been reported in two patients who presented with massive rectal bleeding.^{12,13} In both cases, visceral angiography was thought to show an arteriovenous malformation, but the correct diagnosis of Crohn's disease was subsequently made when examination of the resected segments of bowel revealed transmural inflammation and the presence of granulomas.

Venous and arterial thrombosis is a rare but recognised complication of inflammatory bowel disease described by Talbot *et al*,³ which may be associated with coagulation defects, thrombocytosis, sepsis, or surgery. Mesenteric inflammation and fibrosis are well recognised findings at laparotomy in patients with Crohn's disease¹⁴ and may be extensive. These pathological changes are reflected at visceral arteriography by angulation and corkscrewing of the arterial branches¹¹ which are not dissimilar to those angiographic features seen in patients with carcinoid tumours due to the mesenteric desmoplastic reaction which typically surrounds these tumours.¹⁵ All three of our patients showed stenosis of the main superior mesenteric vein with angiographic appearances of fibrosis of the superior mesenteric vein rather than recanalisation of a thrombosed vessel, with resultant dilatation of the surrounding small bowel venous branches, a previously unreported association of Crohn's disease. It seems likely that cicatrization of the mesenteric vasculature as a result of involvement of the mesentery is responsible for the superior mesenteric vein stenosis and the development of small bowel varices seen in the three cases which we report.

It is perhaps surprising, therefore, that venous stenosis as seen in our three patients (and which is commonly present around carcinoid tumours) has not been previously reported in association with Crohn's disease. It is possible that venous occlusion is, in fact, more common than the literature suggests. One reason for this discrepancy may be that the angiographic studies performed in most of the previous series reporting on the arteriographic findings seen in patients with Crohn's disease were obtained using conventional film on which venous opacification is often only moderate or poor. Recent, more frequent use of a digital subtraction angiographic technique for visceral angiography will, in most instances, produce better images of the venous phase of a superior mesenteric arteriogram due to its improved contrast resolution.¹⁶ Venous involvement will therefore become more apparent.

The diagnosis of venous stenosis as a consequence of fibrosis and subsequent segmental portal venous hypertension as a cause of gastrointestinal blood loss is a hitherto unrecognised association of Crohn's disease with potentially serious implications for patient management.

- 1 Crohn BB, Ginsburg L, Oppenheimer GD. Regional enteritis. A pathologic and clinical entity. *JAMA* 1932;99:1323–9.
- 2 Greenstein AJ, Kark AE, Dreiling DA. Crohn's disease of the colon. II. Controversial aspects of hemorrhage, anemia and rectal involvement in granulomatous disease involving the colon. *Am J Gastroenterol* 1975;63:40–8.
- 3 Mekhjian HS, Switz DM, Melnyk CS, *et al*. Clinical features and natural history of Crohn's disease. *Gastroenterology* 1979;77:898–960.
- 4 Pounder RE. The pathogenesis of Crohn's disease. *J Gastroenterol* 1994;29(suppl 7):11–15.
- 5 Talbot RW, Heppell J, Dozois RR, Beart Jr RW. Vascular complications of inflammatory bowel disease. *Mayo Clin Proc* 1986;61:140–5.
- 6 Daffner JE, Brown CH. Regional enteritis. I. Clinical aspects and diagnosis in 100 patients. *Ann Intern Med* 1958;49:580–94.

- 7 Rubin M, Lynwood Herrington J, et al. Regional enteritis with major gastrointestinal hemorrhage as the initial manifestation. *Arch Intern Med* 1980;140:217-19.
- 8 Homan WP, Tang CK, Thorbjarnarson B. Acute massive hemorrhage from intestinal Crohn's disease. *Arch Surg* 1976;111:901-5.
- 9 Robert JR, Sachar DB, Greenstein AJ. Severe gastrointestinal hemorrhage in Crohn's disease. *Ann Surg* 1991;213:207-11.
- 10 Fallis L. Massive intestinal hemorrhage in regional enteritis. Report of a case. *Am J Surg* 1941;53:512-13.
- 11 Brahme F, Hildell J. Angiography in Crohn's disease revisited. *AJR Am J Roentgenol* 1976;126:941-51.
- 12 DeNardi F, Burns K, Peterson RA, et al. Crohn's disease angiographically mimicking vascular malformation or neoplasm. *J Can Assoc Radiol* 1989;40:279-82.
- 13 Fowler DL, Fortin D, Wood WG, et al. Intestinal vascular malformations. *Surgery* 1979;86:377-85.
- 14 Williams JA. Crohn's disease of the small intestine. In: Zinner MJ, Schwartz SI, Ellis H, eds. *Maingot's abdominal operations*. 10th edn. New York: Appleton and Lange, 1997.
- 15 Makridis C, Oberg K, Junlin C, et al. Surgical treatment of midgut carcinoid tumours. *World J Surg* 1990;14:377-83.
- 16 Katzen BT. Peripheral, abdominal and interventional applications of DSA. *Radiol Clin North Am* 1985;23:227-41.