**CASE REPORT**

**Myelofibrosis and angiodysplasia of the colon: another manifestation of portal hypertension and massive splenomegaly?**

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A 74 year old man presented with a one year history of progressive exertional breathlessness, leg cramps, and numbness of his feet. He had had several months of severe night sweats, poor appetite, and a weight loss of 7 kg. Examination was notable for the passage of fresh blood, either coating or mixed with his stool, on several occasions over the months before this admission. His platelet count at the time of bleeding was 52 x 10^9/litre and his clotting screen was normal (prothrombin time, 14 seconds; activated partial thromboplastin time, 33 seconds; fibrinogen, 4.0 g/litre; and D dimer, 0.38 mg/litre). Plain abdominal x ray showed slightly dilated small bowel loops and thickening of the large bowel mucosa. He underwent emergency colonoscopy, which showed a very unusual appearance, with extensive telangiectatic submucosal vessels increasing with distance from the rectum to the caecum (fig 1). He was treated supportively with blood transfusion and the acute bleeding settled.

His recovery was complicated by post-pneumonic pleural effusion. Unfortunately, two months after discharge, some three and a half years after presentation, he had a further chest infection and died at home.

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Bleeding owing to portal hypertensive colopathy, a form of large bowel angiodysplasia, as a cause of increased blood transfusion requirement was described in a 74 year old man with idiopathic myelofibrosis. The proposed mechanism and the potential therapeutic options for this rare complication of myelofibrosis are discussed.

Angiodysplasia of the gastrointestinal tract causing blood loss and increased blood transfusion requirements in two patients with myelofibrosis was first described in two papers in 1991. In this report we describe what we believe to be only the third example of such an association, and one that we suggest is a consequence of portal hypertension, a recognised complication of myelofibrosis and associated gross splenomegaly.

“We suggest that this is an example of portal hypertensive colopathy, a manifestation of portal hypertension, which is a well recognised complication of patients with cirrhotic portal hypertension”

Although angiodysplasia of the lower gastrointestinal tract and the right side of the colon in particular is a well known cause of rectal bleeding in elderly patients, we illustrate here what our endoscopist reported as an unusual appearance for straightforward angiodysplasia. We suggest that this is an example of portal hypertensive colopathy, a manifestation of portal hypertension, which is a well recognised complication of patients with cirrhotic portal hypertension, also first reported in 1991.

We suspect that portal hypertensive colopathy may be an under recognised cause of gastrointestinal bleeding and increased blood transfusion requirement in patients with myelofibrosis with grossly enlarged spleens.

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DISCUSSION

Bleeding from the gastrointestinal tract in patients with myelofibrosis can be associated with oesophageal varices secondary to portal hypertension, which is a complication in some patients who have massive splenomegaly.1

Although no oesophageal varices were demonstrated in our patient, he did have haemorrhoids, which can be secondary to portal hypertension. In addition, extensive telangiectatic lesions were found throughout his colon. Telangiectatic lesions of the colon, termed portal hypertensive colopathy, are well recognised in patients with cirrhotic portal hypertension, and we suggest that portal hypertension is the mechanism for their occurrence in this case.

“This finding emphasises the importance of imaging the colon in addition to the upper gastrointestinal tract when searching for a cause of increased transfusion requirement in patients with myelofibrosis”

In 1991, Edoute et al reported a case of bleeding jejunoileal angiodysplasia in a patient with myelofibrosis resulting in recurrent malaena.2 Diagnosis was made by intraoperative endoscopy, following transfusion of a total of 65 units of blood to keep the haemoglobin value stable. Attempts were made to secure haemostasis by suturing of the angiodysplastic lesions, but the patient later died from massive gastrointestinal bleeding.

Freedman et al later reported the presence of bleeding telangiectatic lesions in the stomach, duodenum, and entire colon in a patient with myelofibrosis and massive hepatosplenomegaly.3 In this case, myelofibrosis had developed as a result of systemic mast cell disease. The telangiectatic lesions resolved completely within seven months of splenectomy, but the patient died later, cause unknown.

Our report of a third case confirms the association between angiodysplasia, which we attribute to portal hypertensive colopathy, and idiopathic myelofibrosis. This finding emphasises the importance of imaging the colon in addition to the upper gastrointestinal tract when searching for a cause of increased transfusion requirement in patients with myelofibrosis.

There is limited evidence for effective treatment. Lesions may be too extensive to consider endoscopic cautery, laser treatment, or angiographic embolisation. Splenectomy may limit the development of further lesions but may lead to progressive hepatomegaly. Transjugular intrahepatic portosystemic shunt, oral propanolol, octreotide, and other measures to reduce portal pressure have been shown to be effective in some cases of portal hypertensive colopathy.4,5 Whether thalidomide with its anti-angiogenic properties may have a role is necessarily a speculative thought.

Take home messages

- We report the case of a 74 year old man with idiopathic myelofibrosis in whom bleeding as a result of portal hypertensive colopathy, a form of large bowel angiodysplasia, was the cause of increased blood transfusion requirement
- We believe that this case is only the third example of such an association, one that we suggest is a consequence of portal hypertension, a recognised complication of myelofibrosis and associated gross splenomegaly
- Portal hypertensive colopathy may be an under recognised cause of gastrointestinal bleeding and increased blood transfusion requirement in patients with myelofibrosis with grossly enlarged spleens

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