CASE REPORT

Jejunal angiodysplasia causing recurrent gastrointestinal bleeding presenting as severe anaemia and melena

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SUMMARY

Angiodysplasia of the gastrointestinal (GI) tract consists of ectasia of the submucosal vessels of the bowel. The evaluation of such patients needs proctoscopy, colonoscopy, small bowel enema, enteroscopy, capsule enteroscopy and angiography. Capsule enteroscopy has come up as an alternative to GI enteroscopy and colonoscopy in patients with occult GI bleeding; up to 52% cases of small bowel angiodysplasia in patients with occult GI bleed with negative upper GI and colonoscopy have been reported. The use of capsule enteroscopy potentially limits the hazard of radiation exposure from angiography and is less invasive than double balloon endoscopy. The treatment options for angiodysplasias include intra-arterial vasopressin injection, selective gel foam embolisation, endoscopic electrocoagulation and injection of sclerosants, with each of these being technically demanding, and requiring centres with good access to enteroscopy technology and trained gastroenterologists. Operative intervention has been indicated for refractory bleeding or lesions in sites not accessible to endoscopic interventions.

BACKGROUND

Intestinal vascular malformations are a rare entity, and should be considered the differential diagnosis of gastrointestinal (GI) bleeding and chronic anaemia. They can be present in 8–23% of cases evaluated for mid-lower GI bleeding.^{1 2} Angiodysplasias of the large bowel are frequently associated with renal failure and aortic stenosis.^{3 4} Durayet *et al* have also reported a possible correlation of GI angiodysplasias with Von Willebrand's disease.⁵

Angiodysplasia of the (GI) tract consists of ectasia of the submucosal vessels of the bowel. The exact mechanism of development has not been proven, however, chronic venous obstruction has

been implicated as the leading cause.^{5 6} Angiodysplasias present as lower GI bleeding in about 23% cases,² and the incidence of active GI bleed in these cases is usually <10%.⁴ The patients usually present with chronic/occult anaemia and recurrent episodes of melena, and, rarely, haematemesis. The evaluation of such patients needs proctoscopy, colonoscopy, small bowel enema, capsule enteroscopy, enteroscopy and angiography.⁷ Capsule enteroscopy has come up as an alternative to GI enteroscopy and colonoscopy in patients with occult GI bleeding; up to 52% cases of small bowel angiodysplasia in patients with occult GI bleed with negative upper GI and colonoscopy have been reported.⁸ The use of capsule enteroscopy potentially limits the hazard of radiation exposure from angiography and is less invasive than double balloon endoscopy.

CASE REPORT

A 21-year-old man presented with recurrent attacks of melena and weakness, with reduced appetite, for 6 months. Physical examination revealed no significant findings, and there was no evidence of active GI bleed at the time.

The patient had been treated for active GI bleeding with melena in internal medicine 5 months prior; he was investigated and received three units of blood transfusion. He underwent upper GI endoscopy, enteroscopy, colonoscopy, ultrasonography and barium series at the time; all the investigations were normal. Occult blood test of stool was positive. He underwent CT angiography, which revealed a small (7×6×15 mm) strongly enhancing lesion in the distal jejunum (*figure 1*), with intense contrast enhancement in arterial phase and multiple small feeders from the jejunal branches of the

Figure 1 CT angiography revealing small enhancing the distal jejunal lesion.

Figure 2 CT angiography with intense contrast enhancement in the distal jejunum.

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Figure 3 CT angiography showing central dilation and multiple feeders from the jejunal arteries.

superior mesenteric artery (figure 2). The central nidus of the lesion was dilated, with tortuous vessels (figure 3); arteriovenous shunting was noted with faint opacification of the superior mesenteric vein tributaries. The lesion showed drainage into the portal vein via SMV. There was active contrast extravasation in the jejunal lumen (figure 4).

At presentation, the patient's haemoglobin (Hb) was 9 g/dL, but he had repeated melena due to which his Hb% fell to 5.2 g/dL. He was considered for surgical intervention; three units of packed red cells were transfused and the patient was operated electively after 2 days. Intraoperative enteroscopy was also kept on standby during the procedure. A midline laparotomy was performed, revealing a 2×2 cm bluish-red lesion at the antimesenteric border of the jejunum, 5.5 ft proximal to the ileocaecal junction (figure 5). Externally, it appeared as a raised area over the serosal surface. The involved segment was resected and end-to-end jejuno-jejunal anastomosis was carried out. The cut section of the intestine revealed a vascular lesion, a few millimetres long, on the mucosal surface (figure 6).

The patient made an uneventful recovery and was discharged 5 days after the procedure. Biopsy of the lesion revealed angiodysplasia with dilated capillaries, venules and arterioles (figure 7). The patient had no symptoms at 6-month follow-up.

DISCUSSION

Galdabini, in 1974, first used the term 'angiodysplasia'.⁹ Among cases of lower GI bleeding, up to 95% are from the large bowel,

whereas only 5% of cases are from the small bowel. In the large bowel, up to 40% of cases have been reported to be from angiodysplasias.³ Small intestinal angiodysplasias are a rare entity, although several authors have reported cases in children and young adults.^{10 11} Extra-colonic angiodysplasias are noted with colonic lesions in up to 17% of cases.¹² Angiodysplasias occur commonly in the population above 60 years of age. The presence of angiodysplasia in the small bowel has been reported by a few authors.^{13 14} These lesions are characterised by venous dilation in the early stages, followed by dilation of venules, capillary bed and arterioles supplying. In the later stages, there is loss of the precapillary-sphincter mechanism and disruption of the capillary bed, leading to bleeding.^{6 15} Advances in molecular biology have identified several growth factors responsible for the angiogenesis cascade. VEGF is still by far the most well known and most studied molecule in pathogenesis. The unifying hypothesis of upregulation of VEGF in aortic stenosis, von Willebrand's disease and chronic renal failure, is most studied in the angiogenesis-malformation-bleeding cascade. Inhibition of VEGF is the most promising therapy in cases of refractory angiodysplasia.¹⁶

The presence of angiodysplasia goes unnoticed until bleeding occurs. This remains a major cause for delayed recognition of

Figure 4 Extravasation of contrast in the jejunum in CT angiography.

Figure 5 Intraoperative exposure of distal jejunal angiodysplasia.

Figure 6 Specimen photograph.

the disease. Among the diagnostic armamentarium, the use of wireless capsule enteroscopy and angiography has a key role. Easy localisation and marking can be achieved by localisation with CT and injection of methylene blue dye for obscure lesions.¹⁷ In their study, Tonea *et al* contend that selective angiography is the best tool, giving positive results in 85% patients, whereas upper and lower GI endoscopy were diagnostic in only 64% cases.⁴

The most common complication of mechanical circulatory support following ventricular-assist device-use is GI bleeding in 20–40% of patients postimplant.¹⁸ Acquired von Willebrand's factor deficiency and mucosal arteriovenous malformations are two key causes of bleeding. 'The perfect storm' of antiplatelet therapy plus oral anticoagulation, added to depletion of large molecular weight von Willebrand factor, in mechanical support cases, leads to bleeding.

The treatment option for angiodysplasias is medical therapy, for the most part. The first line and most frequently used treatment is endoscopic therapy for short term and active bleeding lesions, and includes intra-arterial vasopressin injection, selective gel foam embolisation, endoscopic electrocoagulation and injection of sclerosants, with each of these being technically demanding and requiring centres with good access to enteroscopy technology and trained gastroenterologists. Short-term control is achieved with endoscopic intervention and long-term control, with somatostatin analogues, hormonal therapy or antiangiogenics. Recently, the use of octreotide, thalidomide, oestrogen and even bevacizumab as antiangiogenic agents in patients has been tried with good response in a small series, however, large prospective randomised trials are needed to prove its efficacy.^{18–21}

Somatostatin analogues (octreotide) effectively reduce

Figure 7 Microphotograph of dilated capillaries, venules and arteries.

bleeding by inhibition of angiogenesis, decrease in splanchnic flow, increase in vascular resistance and improved platelet aggregation.¹⁶ Marked reduction in portal and splanchnic blood flow due to inhibition of vasodilator peptides by somatostatin is the basic mechanism of action, along with control of bleeding. Operative intervention has been indicated for refractory bleeding or lesions in sites not accessible to endoscopic interventions.³ Surgical correction of aortic stenosis is always recommended if associated with angiodysplasia, as it leads to a cure of the disease. Small or large bowel angiodysplasia refractory to medical treatment and poorly controlled with endoscopic therapy needs surgical intervention in the form of intra-operative localisation of the diseased segment followed by resection and anastomosis. In the current case, the patient was operated on as the lesion was located in the distal jejunum, could not be accessed by the gastroenterologist. Given the lack of literature with respect to the location and age of the angiodysplastic lesions in the current case, it can be deemed as a rare presentation.

Learning points

- ▶ Intestinal vascular malformations are a rare entity and should be considered in the differential diagnosis of gastrointestinal bleeding and chronic anaemia.
- ▶ Angiodysplasias usually present with chronic/occult anaemia and recurrent episodes of melena, and, rarely, haematemesis.
- ▶ The treatment option for long-term control in angiodysplasias is medical therapy, for the most part; the use of octreotide, thalidomide, oestrogen and even bevacizumab, as antiangiogenic agents, has been tried with good response considering the multifocal nature of the disease.
- ▶ The treatment options for short-term control in angiodysplasias include intra-arterial vasopressin injection, selective gel foam embolisation, endoscopic electrocoagulation and injection of sclerosants.
- ▶ Operative intervention has been indicated for refractory bleeding or lesions in sites not accessible to endoscopic interventions.

Contributors SKT and MZH drafted the manuscript and collected data. PK and AKK critically reviewed and drafted the final manuscript.

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