Successful management of severe gastrointestinal bleeding from jejunal angiodysplasia in a patient with Bernard-Soulier syndrome

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DESCRIPTION

A 57-year-old woman presented to the emergency department with melena, easy fatigability and dizziness for 2 days. She did not have haematemesis, abdominal pain, ascites or jaundice. Her routine master health check-up done 4 years previously revealed a diagnosis of Bernard-Soulier syndrome (BSS). She did not have any bleeding manifestation so far. Her personal history and family history were not contributory. On examination, she was drowsy, pale, tachycardic and hypotensive. Her abdomen examination was unremarkable, and digital rectal examination revealed melenic stool staining. Her blood investigations showed severe anaemia (haemoglobin: 54 g/L) and thrombocytopenia (platelet count: $65 \times 10^9/L$), with giant platelets on a peripheral smear. She received two units of packed red blood cells. Her initial upper endoscopy, colonoscopy and CT abdominal angiogram did not reveal any abnormality. Video capsule endoscopy showed multiple brisk bleeding spots in the proximal jejunum (figure 1A–D). She received octreotide infusion initially. However, there was no response for over 72 hours, and her haemoglobin value did not improve. Hence, we decided to proceed with diagnostic laparoscopy and intraoperative

Figure 2 (A) Photograph of the resected segment of the proximal jejunum. (B) Photomicrograph showing the biopsy of the resected segment with normal mucosa, thickened walls of the submucosal blood vessels, veins exhibiting arterialisations of walls and thin-walled capillaries (H&E: 10x).

enteroscopy. It revealed multiple ectatic cherry red spots in proximal jejunum, with active diffuse pinpoint ooze. She underwent resection of 50 cm of proximal jejunum in the same sitting (figure 2A). Histopathological examination of the resected bowel confirmed diagnosis of angiodysplasia (AD) (figure 2B). She had an uneventful postoperative period and got discharged in a stable condition. She did not have further bleeding episodes and is doing well for the past 2 years.

AD is the most frequently reported cause for bleeding from the small bowel.¹ It presents as chronic anaemia due to occult gastrointestinal (GI) blood loss or an overt GI bleed like melena or haematochezia. The presence of a concomitant systemic condition like chronic kidney disease, aortic stenosis or haemostatic disorder increases the risk of bleeding from ADs.² BSS presenting with GI bleeding from ADs is rarely encountered.^{3,4} The association between BSS and AD is not well established, and whether such ADs are more prevalent in BSS or incidentally diagnosed because of bleeding is not known. Video capsule endoscopy is the first-line modality for small bowel evaluation and has a higher diagnostic yield if performed early. Other modalities include CT angiography, push enteroscopy and device-assisted enteroscopy. Intraoperative enteroscopy should be reserved as a final option when other diagnostic modalities fail or are not available. The various treatment modalities for intestinal AD include pharmacological treatment, endoscopy, radiological intervention and surgery. Somatostatin analogues and thalidomide have shown promising results in some patients.⁵ Endoscopic treatment includes argon plasma coagulation, sclerotherapy

Figure 1 Video capsule endoscopy images showing (A) normal duodenal mucosa, (B) proximal jejunum showing active ooze of blood, (C) distal jejunum showing altered blood and (D) terminal ileum showing black-coloured stool staining of the mucosa.

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Images in...

with polidocanol and clipping. In patients with haemodynamic instability, selective angiography and embolisation or coiling may achieve haemostasis.⁶ Due to advancements in endoscopic and radiological techniques, such lesions rarely mandate a surgical treatment nowadays. However, surgery plays an important role when the source of the GI bleeding is not detectable and when there is haemodynamic instability. The prognosis after surgery is generally good, and they have to undergo haemogram during periodic follow-up.

Patient's perspective

I thank the team of doctors who tried all their best to save me. But I am worried that the bleeding may recur in the future.

Learning points

- ▶ Gastrointestinal angiodysplasias may cause life-threatening bleeding, when associated with a concomitant haemostatic disorder.
- ▶ Intraoperative enteroscopy may be helpful when all other options fail.
- ▶ Recurrence of bleeding must be anticipated for the rest of their life.

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