CASE REPORT

Small intestine polypoid arteriovenous malformation: a stepwise approach to diagnosis in a paediatric case

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SUMMARY

We report a case of acute gastrointestinal haemorrhage due to a small intestine polypoid arteriovenous malformation (AVM) in a patient with a remote history of obscure gastrointestinal bleeding (OGIB) 8 years earlier. The diagnosis of a small intestine AVM was made using video capsule endoscopy (VCE) and confirmed using single-balloon push enteroscopy. The lesion was marked with submucosal tattoo to aid in subsequent surgical resection of the lesion with primary duodenoduodenostomy. Since our patient's initial bleeding episode, a variety of advanced tools have become widely available to aid in the localisation of OGIB. This case illustrates the use of a stepwise approach using new medical technology to identify and manage OGIB in children. VCE and push enteroscopy proved to be important diagnostic modalities in this paediatric case.

BACKGROUND

Obscure gastrointestinal bleeding (OGIB) originating from the small intestine presents a challenging diagnostic dilemma for clinicians. Small intestinal haemorrhage represents only 16% of paediatric gastrointestinal bleeding, primarily in the form of Meckel's diverticulum.¹ Polypoid arteriovenous malformations (AVMs) are rarely diagnosed in childhood. This case highlights the need for a stepwise approach to the diagnosis of OGIB in children.

CASE PRESENTATION

A 17-year-old boy presented to the emergency department with abrupt onset of haematochezia. A total of five grossly bloody stools were reported the day of presentation with associated lightheadedness. No abdominal pain, nausea, vomiting or diarrhoea were present. His vital signs revealed tachycardia but a normal blood pressure. He had no abdominal tenderness and normal abdominal auscultation. Rectal examination revealed scant bright red blood in the rectal vault.

Of note was a previous history of OGIB 8 years earlier with a positive technetium-99m pertechnetate scan (Meckel's scan) but no evidence of Meckel's diverticulum or any other potential cause of bleeding on laparoscopic exploration. A conventional esophagogastroduodenoscopy (EGD) and ileocolonoscopy did not reveal a source for the gastrointestinal bleeding. He also had a medical history of a right arm cavernous haemangioma that required surgical resection. His family history revealed a brother with a Meckel's diverticulum, a mother with psoriatic arthritis and a paternal uncle with Crohn's disease.

INVESTIGATIONS

Laboratory studies revealed a haemoglobin concentration of 9.1 g/dL (normal, 11.0–14.5 g/dL), haematocrit of 26.6 with a mean corpuscular haemoglobin of 83.4. His white blood cell count of 6810 cells per cubic millimetre (normal, 3840–9840 cells per cubic millimetre) and a platelet count of $175 \times 10^3 \text{ mm}^3$ (normal, $140\text{--}440 \times 10^3 \text{ mm}^3$). His coagulation studies included a prothrombin time of 14.6 s (normal 12.1–14.6 s), partial thromboplastin time of 24.3 s (normal 23.5–36.4 s) and INR of 1.0.

DIFFERENTIAL DIAGNOSIS

- ▶ Meckel's diverticulum.
- ▶ Small intestinal AVM/telangiectasia.
- ▶ Small intestinal varices.
- ▶ Aorto-enteric fistula.

TREATMENT

The patient underwent a stepwise evaluation to determine the source of his OGIB. A tagged red blood cell nuclear medicine scan was performed initially that was negative. A contrast-enhanced CT angiography scan (CTA scan) did not reveal the source of bleeding. Conventional EGD and ileocolonoscopy again did not identify any abnormalities. A video capsule endoscopy (VCE) device was deployed into the duodenal bulb during the EGD procedure. The VCE revealed a lesion in the small intestine with stigmata of a recent bleed (figure 1). An antegrade single balloon endoscopy was then performed and the lesion was discovered in the proximal jejunum. The site was marked with submucosal tattoo to aid subsequent surgical approach to the lesion (figure 2). Ultimately, laparoscopy showed the tattooing immediately distal to the

Figure 1 VCE image revealing a small intestine polypoid malformation. VCE, video capsule endoscopy.

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Figure 2 Push enteroscopy images revealing this lesion as well as the transmural 'tattooing'.

Figure 3 Gross pathology specimen image revealing the venous malformation.

ligament of Treitz. The surgeons decided to switch to an open procedure due to the location and a laparotomy was performed. The ligament of Treitz was mobilised and the lesion identified via palpation in the area of tattooing. The affected small intestine was removed and a primary duodenoduodenostomy performed (figure 3).

OUTCOME AND FOLLOW-UP

Postoperative pathology report of the resected tissue was consistent with a submucosal venous malformation. Our patient developed a postoperative ileus that required nasogastric tube decompression and temporary parenteral nutrition. On postoperative day 6, his ileus resolved and he was able to start advancement of oral feedings. He was discharged home on postoperative day 8. The patient was followed clinically over time without further endoscopic evaluation. No further episodes of gastrointestinal bleeding were reported in 1 year of follow-up.

DISCUSSION

Approximately 5% of OGIB cases remain undiagnosed despite thorough investigations of the GI tract.¹ In particular, haemorrhage from the small bowel can be a challenge as the bleeding can be intermittent with variable rates of blood loss.² Markedly few cases of paediatric polypoid AVM have been empirically documented and remains a rare cause of haematochezia.³⁻⁶ Most AVMs are ileocolic in nature and are more commonly seen in the adult population.⁴⁻⁸ AVMs of the small bowel are most commonly found in the jejunum or ileum and are more likely congenital lesions in contrast to the acquired adult AVMs of the large bowel.^{4,5}

Patients with intestinal AVMs typically present with painless bright red rectal bleeding and iron deficiency anaemia that results in further evaluation.⁷ This presentation is similar to that of a Meckel's diverticulum, the most common cause of paediatric small intestine haemorrhage.¹ Additionally, variceal bleeding and aorto-enteric fistulas can present with massive rectal bleeding. However, variceal bleeding is typically preceded by a known history of chronic liver disease and aorto-enteric fistulisation is an exceedingly rare cause of rectal bleeding. Those patients that present with massive rectal bleeding with transfusion dependent symptomatic haematochezia warrant rapid diagnostic evaluation given the risk of life threatening haemorrhage.

Diagnosis of AVMs of the gastrointestinal tract can pose a challenge. Prior to the advent of VCE, angiography was the gold-standard for diagnosis of AVMs. Vascular congestion is considered evidence of a likely AVM when using conventional angiography or CTA.^{6,8} However, the benefits of angiography are more notable with larger AVMs but less so for small bowel telangiectasias.⁹ Diagnostic evaluation has progressed to the use of push enteroscopy in OGIB. Angiography and push enteroscopy can delineate the lesion in up to 40% of cases.⁸

We report a polypoid lesion that was diagnosed with VCE, which has a higher sensitivity than arteriography for diagnosis of AVM.¹⁰ Studies have demonstrated its ability to identify causes of blood loss in the small bowel twice as often as push enteroscopy.¹⁰ In addition, it has been shown to be equivalent to the use of push enteroscopic evaluation of the complete gastrointestinal tract and should be considered as a first-line modality for localisation of a lesion.¹¹ These benefits are further enhanced with the use of VCE early on in OGIB as well as with more severe bleeding.^{12,13} It is also a preferred modality of initial investigation by patients.¹¹ The potential complication of retention or aspiration is uncommon.^{14,15} The use of VCE, however, can be limited by the physical constraints of capsule size according to the lumen diameter of younger paediatric patients.

Given our case and the available literature on VCE, we feel that there is sufficient support to advocate for the use of VCE early in the diagnosis of OGIB in the paediatric population. We believe that the early use of VCE with subsequent endoscopic modalities is superior to the use of CTA for the diagnosis of GI bleeding and warrants further investigation in a larger study.

Patient's perspective

The first time our son had internal bleeding he was almost 9 years old. He had hit his side on a handle of a foosball table while horse playing with his older brother. It did hurt him, but we played it off as just another bump or bruise kids get until about an hour later when he went to the bathroom. I did not know what to think when I saw the blood he had passed. We rushed him to the Emergency Room which was 20 miles away. During his exam there he passed a small amount of blood. Because of this along with some liver issues in the past, they sent him by ambulance to the Children's Hospital in Birmingham, Alabama. The staff was constantly asking us questions about his condition. We felt for a while that they were suspecting us of having done something to him. After 9 days in the hospital and numerous tests, doctors did not know what had caused his blood loss. We went home not knowing what had happened or what to expect in the future.

Now at age 17 with no pain or any other warning, he passes blood again. We were driving home from vacation so I continued to drive to Huntsville Hospital emergency room. After a quick preliminary exam, they sent him down the street to Huntsville Women and Children Hospital. He passed more blood there that they were able to witness. This convinced them to send him by ambulance to Children's Hospital in Birmingham since they were familiar with his medical history.

Back in Birmingham all the testing started again. Nothing seemed to show the true problem. Then we were told about the 'PillCam'. The 'PillCam' showed the problem in his small intestines, it was an 'AVM'. Later on they were able to perform surgery and remove the area of his intestines that were affected. This time as we left the hospital we felt like his problem had been solved. We hoped that we would not have to return. He healed from the surgery and appears to be fine now.

Learning points

- ▶ Paediatric small intestinal arteriovenous malformation is a rare cause of haematochezia.
- ▶ The diagnosis is challenging, given the intermittent haemorrhage of these lesions, and often requires multiple modalities to assist in localising these lesions.
- ▶ A standardised algorithmic approach is important in arriving at the final diagnosis and we recommend early use of video capsule endoscopy as well as push enteroscopy.
- ▶ Treatment options include primary surgical resection and embolisation with interventional radiology.

Contributors ALM is the primary author, drafted the initial manuscript and approved the final manuscript as submitted. He also cared for this patient during their hospitalisation. TJ provided project leadership and content, critically reviewed and approved the final manuscript as submitted. She also cared for this patient during their hospitalisation. JN provided project leadership and content, critically reviewed and approved the final manuscript as submitted. He also cared for this patient during their hospitalisation.

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