A rare cause for severe recurrent lower gastrointestinal bleeding in a 12 year old patient

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ABSTRACT

The cause for severe, recurrent lower gastrointestinal (LGI) bleeding in children can usually be diagnosed readily by means of the commonly used investigative/diagnostic techniques such as colonoscopy, laparoscopy ± laparotomy. Occasionally less commonly used investigations may be necessary to look for more elusive causes of LGI bleeding such as capsule endoscopy, angiography, technetium-99m (99m Tc) labeled red blood cell (RBC) scintigraphy, cross-sectional imaging such as CT/MRI (including angiography) and laparotomy combined with on-table small bowel enteroscopy. We report a case of severe, recurrent LGI bleeding that had occurred over several years, where the cause remained elusive despite numerous investigations and interventions. The etiology of this was eventually found to be a gastric duplication cyst infiltrating into adjacent transverse colon and causing bleeding from peptic ulceration in the colon. The process by which this diagnosis was made and the lessons learned are discussed.

Subsequently, from 9 years of age per rectal bleeding was more significant leading to hemoglobin drop of 30–50 g/L on multiple occasions necessitating blood transfusions. Bleeding episodes were usually preceded by a few days’ history of poor appetite, weight loss and significant peri-umbilical pain which persisted until bleeding ceased. A CT angiogram was normal but a formal catheter angiogram suggested a right colonic source with a vascular blush. At age 9.5 years a laparotomy with entire small bowel enteroscopy was performed with no diagnostic yield. Her small bowel appeared normal at laparotomy, except that there was blood in the colon from the cecum distally. In view of the recurrent nature of the bleeding and the transfusion dependency (7 transfusions between 9 and 10.5 years of age), a right hemi-colectomy from the terminal ileum to the cecum was performed at 10.5 years. She remained well for a few months but then she had a further bleed. She was given 20 mg of omeprazole daily throughout this period and subsequently.

Between 10 and 11 years of age she had two normal abdominal ultrasounds and a red cell scan. The red cell scan showed a blush in the pelvis which may have been from the transverse colon but repeat colonoscopy was normal, albeit on two occasions good mucosal visualization was prevented by the presence of large amounts of blood in the colon.

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1. Case report

An 11 year old female patient was transferred to our unit for a second opinion in view of severe, recurrent and obscure LGI bleeding.

The first presentation of LGI bleed was at 6 weeks of age with intermittent history of fresh unaltered blood in her stool. Assumption of cow’s milk enteropathy was made and hence she was started on hydrolyzed formula. At five months of age she was passing fresh blood with clots and hence she was referred to the gastroenterology team. Upper GI endoscopy and ileo-colonoscopy were normal. She had a further large fresh PR bleeding with clots during infancy and mini-laparotomy occurred which excluded Meckel’s diverticulum. She had a history of poor appetite, weight loss and significant peri-umbilical pain which persisted until bleeding ceased. A CT angiogram was normal but a formal catheter angiogram suggested a right colonic source with a vascular blush. At age 9.5 years a laparotomy with entire small bowel enteroscopy was performed with no diagnostic yield. Her small bowel appeared normal at laparotomy, except that there was blood in the colon from the cecum distally. In view of the recurrent nature of the bleeding and the transfusion dependency (7 transfusions between 9 and 10.5 years of age), a right hemi-colectomy from the terminal ileum to the hepatic flexure was performed at 10.5 years. She remained well for a few months but then she had a further bleed. She was given 20 mg of omeprazole daily throughout this period and subsequently.

Between 10 and 11 years of age she had two normal abdominal ultrasounds and a red cell scan. The red cell scan showed a blush in the pelvis which may have been from the transverse colon but repeat colonoscopy was normal, albeit on two occasions good mucosal visualization was prevented by the presence of large amounts of blood in the colon.

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bleeding. After effective bowel clear-out colonoscopy revealed a colonic stricture with an adjacent polypoid lesion and ulceration at around 30 cm from the anal verge. Injected contrast (on-table) showed an apple-core appearance of the stricture.

This was followed by a dual-injection single-phase (Afghan trauma protocol) contrast-enhanced CT abdomen which revealed a 3–4 cm diameter rounded lesion with a fluid-filled center and a stratified wall (Fig. 1), which was consistent with a duplication cyst arising from the stomach or colon.

A few days later a further severe LGI bleed necessitated an emergency laparatomy. This was preceded by a colonoscopy and the stricture was tattooed with methylene blue to facilitate serosal identification at laparotomy. Upper GI endoscopy showed a 2 cm dome-shaped indentation into the stomach with no mucosal invasion or erosion (Fig. 2). At laparotomy a gastric duplication cyst arising from the greater curve of the stomach was identified. Dissection revealed that this cyst was not communicating with the stomach but was infiltrating and fistulising into the adjacent colon at the splenic flexure, resulting in an associated stricture of the colon (Fig. 3). Stapled resection/anastomosis of the splenic flexure stricture was performed. The gastric duplication cyst was resected by means of a stapled partial gastrectomy. Post-operative period was uneventful and patient was discharged one week later. No further bleeding has been reported 6 months later. Histopathological examination of the specimen showed regenerative changes, sub-mucosal fibrosis and ulcerated mucosa of the resected part of transverse colon with a duplication cyst showing perforated gastric duplication cyst with fundic type mucosa (Figs. 4 and 5).

2. Discussion

Significant LGI bleeding is usually investigated by means of an algorithm of sequential investigations and a cause can usually be found [1]. The appearance of a gastric duplication cyst in older children can present a diagnostic dilemma. Due to their uncommon occurrence there is no established diagnostic algorithm to aid in the
diagnosis and treatment of such a potentially life threatening GI bleeding.

Duplication cysts are defined by a smooth muscle coat, an intimate attachment to the native GI tract and a GI mucosal lining [2]. Gastric duplication cysts are rare and account for only 4–8% of all duplication anomalies affecting the intestinal tract [3]. A majority of duplication cysts occurred in females regardless of age with a female to male ratio range from 2:1 to 4:1 reported to date [4]. Occasionally identified by antenatal scan [5], the usual presenting symptoms include PR bleeding, bowel obstruction, abdominal pain or an abdominal mass usually in infancy but less commonly in older children. Complications include torsion of pedunculated cysts, malignant changes, hemorrhage, pancreatitis and fistula formation. Gastrointestinal bleeding is a rare presenting feature and may be caused by peptic ulcer formation due to the cyst containing gastric mucosa. The bleeding can be painless, brisk and life threatening, with the first bleed occurring usually early in life [6].

Amouei et al. reported a paediatric patient with hematemesis and melena and endoscopic appearance of a large sub-mucosal mass in the fundus of stomach with a large 3 cm² ulcer noted with a significant artery at its base — differential diagnoses considered included leiomyoma or gastrointestinal stromal tumor (GIST) [7]. Interestingly, a review of the article reveals only 3 other similar cases where a gastric duplication cyst caused fresh LGI bleeding by infiltrating into the adjacent colon [8–10]. The first described a 5 month old with severe lower gastrointestinal bleeding due to a gastric cyst eroding into the transverse colon [8], whereas another case reported an intra-pancreatic gastric duplication cyst causing massive lower gastrointestinal bleeding in an 8 month old resulting from a peptic ulceration of the adherent jejunum [9]. Surridge et al. [10] describe rectal bleeding in a 13 month old from a perforated gastric duplication cyst that had eroded into the right lobe of the liver as well as the hepatic flexure of the colon. Although it is difficult to diagnose GDC preoperatively, recent imaging modalities have provided some informative findings. CT scan and endoscopic ultrasound (EUS) are the best ways to identify GDC [11]. It is of note
that the duplication cyst was not identified during the mini-laparotomy the year before the diagnosis. Possible explanations for this might be the cyst was in development, was varying in size at different times or that the left-side of the colon was not examined. Why the gastric duplication preferentially infiltrates into the adjacent colon rather than into the gastric lumen remains unclear. In this case it was clear at laparotomy that this was a gastric duplication as it shared muscle and serosal layers with the stomach and the involvement with the colon was only secondary to fistulation. It is known that colonic duplication cysts can contain gastric mucosa but this is not the case here.

Once a gastric duplication cyst is detected, surgical excision of the entire gastric cyst remains the treatment of choice in all complicated intra-abdominal enteric duplications [3].

Other bleeding duplication cysts are well documented in the literature. Duodenal duplication cysts can lead to GI bleeding as a result of peptic ulceration of the ectopic gastric mucosa within the cyst and eroding the duodenal wall [12]. Ileal duplication cysts can present with LGIB of varying severity — stool occult blood positivity with anaemia to fresh bleeding per rectum. Diagnosis can be made by Tc(99m) pertechnetium scan, DBE and laparotomy [13]. Rectal duplication cyst can present at times with per rectal bleeding described in adults [14].

We feel that in this case the diagnosis — which had remained elusive over a period of 11 years — may have been aided by a combination of disease progression leading to the development of significant changes in the colon enabling visualization at colonoscopy and on cross-sectional imaging of the abdomen (CT scan). The previous CT did not detect the lesion probably because it was performed as an angiogram (with no portal venous phase) and because the scan area was limited and did not include the whole upper abdomen.

The second consideration is that previous colonoscopies suffered from poor vision due to acute bleeding and blood obscuring the abnormal mucosal appearances. Referral to a different team of clinicians who are approaching the problem as new may have been a factor.

A review of the management preceding referral to the quaternary GI bleeding unit reveals the following possible lessons:

- The importance of ‘starting again’ with referrals from other tertiary centers;
- Colonoscopy with good mucosal visualization and bowel preparation that is adequate;
- Lesion recognition on concurrent upper GI endoscopy; and that
- CT angiography alone is not good enough for anatomical lesion identification.

A dual injection CT scan with simultaneous arterial and venous phase contrast can be used to identify lesions as well as delineate the mesenteric vessels.

In conclusion, this case demonstrates the challenges in diagnosing rare conditions despite the availability of a variety of investigation modalities at the disposal of the paediatric gastroenterologist and surgeon. A multi-disciplinary approach, team work and the sharing of difficult problems between teams and institutions is best practice and is most likely to yield a positive and safe result.

References