PHLEBECTASIA OF THE JEJUNUM IN A CHILD
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Abstract
A 12 year old boy presented with repeat episodes of lower gastrointestinal bleeding. We outline the pitfalls encountered en route to making the correct diagnosis and undergoing definitive surgery. Although not a classical presentation, his symptoms initially responded to therapy for ulcerative colitis (UC), confirmed clinically and on histology. However, he was readmitted with lower gastrointestinal bleeding. Repeat colonoscopy was grossly normal except a small polyp, and histology suggested UC. It was not until, on the third presentation with a precipitant hemoglobin drop, that capsule enteroscopy in conjunction with CT angiography was performed, and the site of blood loss identified. Surgical resection of the affected jejunum was curative and histology confirmed the diagnosis. Identifying the site of small bowel bleeding can be challenging due to its inaccessibility, length, vigorous contraction and overlying loops. Phlebectasia is a rare cause of gastrointestinal bleeding in children. Our case highlights the importance of keeping an open mind and a multimodality approach to investigating ongoing obscure gastrointestinal bleeding in children.

Keywords: congenital vascular malformation, intestinal, phlebectasia, children, gastrointestinal bleeding, capsule enteroscopy, CT angiography, ulcerative colitis

Introduction
Intestinal phlebectasias is a rare benign vascular anomaly consisting of varicosities without portal hypertension and is a rare cause of gastrointestinal bleeding in children. We present a case of 12-year-old boy presenting with recurrent lower gastrointestinal bleeding simulating ulcerative colitis. The first case of phlebectasia of the jejunum in a child was reported in 2008 who presented with life-threatening gastrointestinal bleeding due to this special vascular anomaly [1]. This is the second paediatric case reported in the English literature with a chronic presentation. Our case highlights the importance of a multimodality approach to investigating on-going obscure gastrointestinal bleeding.

Case report
In the first sub-acute presentation, a previously fit and well 12-year-old boy presented with a 2 week history of painless passing of altered blood per rectum, pallor and easy fatigability. He had no other gastrointestinal symptoms and his bowel habits were normal. He had no discomfort in the abdomen. Meckel’s diverticulum with ectopic gastric mucosa, communicating duplication cyst containing gastric mucosa, inflammatory bowel disease especially ulcerative colitis, vascular anomalies and allergic colitis were considered in the differential diagnosis at first admission.

Complete blood count showed haemoglobin of 70 g/L with microcytic hypochromic picture suggestive of iron deficiency anemia due to blood loss. Other investigations including C-reactive protein, white cell count, stool microscopy, culture and sensitivity, immunoglobulins, pANCA, anti-amoeba antibodies, coeliac antibodies, were all within normal limits. Ultrasound scan of the abdomen was normal. Upper gastrointestinal endoscopy was normal. Lower gastrointestinal endoscopy showed erythema extending from rectosigmoid junction to hepatic flexure and biopsies revealed focal active colitis suggestive of ulcerative colitis. He was treated with prednisolone, ranitidine and ferrous fumarate. Outpatient clinic fol-

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low up at 1 month showed low ferritin and haemoglobin levels persistently low despite iron therapy. He was treated as ulcerative colitis and commenced on onolsalazine. On the second emergency admission, he presented 2 years later, while on maintenance treatment with massive lower gastrointestinal bleeding and hypotensive shock with haemoglobin of 5.7 g/L. He underwent colonoscopy which revealed polyp in cecum with multiple ectatic vessels and an ulcerative colitis on colonic biopsies. Treatment was changed to mesalazine. Follow up colonoscopy at 3 months later was better and biopsies were doubtful about the ulcerative colitis. Colonoscopy 9 months later showed all biopsies to be normal histologically. Medications were stopped as child was well.

On the third emergency admission, he presented 5 days later following cessation of medications, with lower gastrointestinal bleeding - altered blood and fresh rectal bleeding - and required transfusion. Contrast CT, endoscopies and Meckel’s scan did not reveal source of gastrointestinal bleeding. Capsule enteroscopy showed dilated veins within the lumen of the small bowel (Fig. 1). CT angiography identified site as jejunum (Fig. 2). He underwent superior mesenteric angiography which showed no arterio-venous malformation. Upper and lower gastrointestinal endoscopies were normal. At an exploratory laparotomy, proximal section of jejunum was thickened with dilated, prominent and tortuous veins (Fig. 3). Proximal jejunal segmental resection containing multiple phlebectasia and primary anastomosis was carried out uneventfully. A total of 53 cm of bowel was resected (Fig. 3). The cut section of the resected jejunal segment showed dilated vessels (Fig. 3 inset). Diagnosis was confirmed by histology which suggested phlebectasia (Fig. 4).

He was discharged home 3 days post-operatively. He is currently asymptomatic, thriving well and off all medications at 5 years after final surgery. Recently he has been discharged from follow up.

Discussion
There are several causes of altered lower gastrointestinal blood in stool in a child [2]. Intestinal phlebectasias are venous varicosities in
the absence of portal hypertension. The vein is grossly dilated and tortuous, but with a normal vascular wall and little surrounding connective tissue stroma. The most common site is the jejunum [3,4]. This has been a somewhat blind area in the gastrointestinal system; beyond the reach of conventional upper and lower gastrointestinal endoscopes. However, advent of video capsule enteroscopy (VCE) [5] and double balloon enteroscopy (DBE) [6] has opened up the door to this relatively inaccessible segment of small intestine. The identification of small bowel lesions, presenting as chronic gastrointestinal bleeding, is a challenge. Upper and lower gastrointestinal endoscopies tend to be negative. Capsule enteroscopy or double-balloon enteroscopy are not frequently used in the pediatric population.

In our case the initial colonoscopy findings facilitated the error of ‘satisfaction of search’. History of altered lower gastrointestinal blood and the lack of raised inflammatory markers might have led us to question the initial diagnosis of ulcerative colitis. However, the diagnosis seemed to be confirmed with an absence of positive findings in other investigations, a response to treatment, and further confirmatory histology reports on repeat endoscopy and biopsy. It was not until his third presentation, with precipitant hemoglobin drop on this occasion, that a small bowel source was considered and the bleeding source finally identified with a combination of capsule enteroscopy and CT angiography.

While this case highlights the importance of a multimodal approach to investigating ongoing obscure gastrointestinal bleeding, it also emphasizes the importance of keeping an open mind.

Despite an initial diagnosis being made on clinical findings there is still the potential for an alternate pathology being present, particularly when the story does not completely fit with the histopathology. Identifying the site of small bowel bleeding can be challenging due to its inaccessibility, length, vigorous contraction and overlying loops. 99mTc labelled red cell scans are sensitive but it is difficult to localise and impossible to characterize the lesion. Digital subtraction angiography is useful in identifying active bleeding but not so helpful when bleeding has slowed down [7]. It is possible to evaluate the whole small bowel using double balloon enteroscopy. Capsule enteroscopy is the current investigative modality of choice as an initial screening investigation in patients with occult gastrointestinal bleeding with negative upper GI and colonoscopy. However while it is a less invasive procedure, it is non-therapeutic, may not be able to image the whole small bowel if slow transit, and does yield false positive results.

A recent case series by Ohmiya N et al. [8] has demonstrated that VCE was useful for screening and DBE enabled diagnosis and endoscopic treatment for bleeding lesions deep within the small bowel. Surgery has been regarded as the definitive therapy for intestinal phlebectasias in actively bleeding lesions. However in cases of multiple or extensive lesions resection might not be practical. In this small series [8], enteroscopic injection sclerotherapy using polidocanol proved beneficial for treating phlebectasias. However these lesions are known to relapse. At present, surgical resection remains the gold standard.

Congenital vascular malformations are classified into haemangioma and vascular malformation and both differ from each other so much that they have nothing in common except for their external appearance may be similar [9]. Haemangioma is further classified into common infantile hemangioma [rapidly involuting...
congenital hemangioma (RICH) or noninvoluting congenital hemangioma (NICH), kaposiform hemangioendothelioma (KHE) tufted angioma and intramuscular hemangioma. While the vascular malformations are classified into high-flow lesions—arteriovenous malformations (AVM) and arteriovenous fistulas (AVF) and low-flow lesions—capillary malformations (CM) – port wine stain, venous malformations (VM), cavernous lesion, lymphatic malformations (LM) and lymphatic-venous malformations (LVM). Both can have rare syndromic presentations and an additional class of lesions. Our lesion was venous cavernous type of vascular malformation.

Conclusion

Jejunal vascular phlebectasia, although congenital, may be missed in the prenatal and postnatal period and may present in the adolescent period or in the adult life. Diagnosis is elusive and misleading unless there is high index of suspicion in the light of unusual clinical features, so health professionals should be aware of these uncommon congenital anomalies. Identifying the site of small bowel bleeding can be challenging due to its inaccessibility, length, vigorous contraction and overlying loops. Phlebectasias are a rare cause of gastrointestinal bleeding in children. We should keep our options open when investigating obscure gastrointestinal bleeding. Multimodal approach to investigating on-going obscure gastrointestinal bleeding is vital to establish the diagnosis and management, capsule enteroscopy is the key investigation. Current gold standard treatment of symptomatic phlebectasia of the gastrointestinal tract remains surgical resection; however, recent evidence has shown that enteroscopic interventional sclerotherapy may be an alternative therapy available.

Figure 4: Histology: dilated, thin walled blood vessels (V) in submucosa (SM) mainly with occasional subserosal dilated channels. Some ulceration of the villi tips. Appearances suggestive of cavernous type of venous vascular malformation (venous phlebectasia)
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