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Vascular malformations of the small intestine manifesting as chronic anemia: Two pediatric cases managed by single-site umbilical laparoscopic surgery

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ABSTRACT

INTRODUCTION: Vascular malformations affecting abdominal viscera, especially the gastrointestinal tract, are less common than that in other body segments. Nonetheless, it seems to be one of the important causes of gastrointestinal bleeding in not only adults but also children as well. It occurs during the development stage of vascular system, and may increase in severity as the child grows.
PRESENTATION OF CASE: We present here two cases of lesions developed at the small intestine in an 8-year-old girl and 3-year-old girl, which were identified during the management for chronic anemia. Although there were some limitations associated with diagnosis, a histology confirmed the presence of arteriovenous malformations in both cases, they were successfully treated with surgical resection, especially minimal invasive procedure.
DISCUSSION: Vascular malformations of abdominal viscera, especially the small intestine, are rare clinical manifestations in pediatric patients but are among the important causes of acute massive or chronic obscure LGI bleeding. Unless there is significant GI bleeding, patients are usually treated for anemia with obscure LGI bleeding. In the present study, selective angiography was useful in one case and CT enterogram with angiography was useful in the other case.
CONCLUSION: Considering the rarity and possibility of gastrointestinal bleeding due to vascular malformations, it is necessary to be regarded as one of differential diagnosis when managing a lower gastrointestinal bleeding in pediatric patients. Besides, a minimal invasive procedure could be suggested as a good surgical option when necessary.

1. Introduction

Vascular malformations affecting the gastrointestinal (GI) tract are less common than that of the head and neck, trunk, extremities, and other soft tissues. Despite its rarity, it may cause a GI bleeding ranging from chronic obscure bleeding to acute massive bleeding. Vascular malformations can occur during the development of vascular system and increase in severity as the child grows. Even though vascular malformations can manifest anytime during infancy, childhood, or adolescence, they often remain unrecognized until the development of symptoms.

In adults, vascular malformations of the colon are regarded as one of the important causes of lower gastrointestinal (LGI) bleeding, while LGI bleeding in pediatric patients is more commonly associated with anal fissure, Meckel's diverticulum, intussusception, polyps, and or necrotizing enterocolitis. Contrary to these conditions, GI bleeding from vascular malformations occasionally may affect the delay in diagnosis because of its rarity and limitations in the diagnostic approach in pediatric patients.

Herein, we present two pediatric cases of LGI bleeding due to vascular malformations in the small intestine that were identified during the management of iron-deficiency anemia.

2. Presentation of case

2.1. Case 1

An 8-year-old girl was admitted to the emergency room with a massive hematochezia. The patient had been followed-up for treatment of iron-deficiency anemia, 3 months prior, but presented with no gastrointestinal symptoms at the time. Initial laboratory findings showed a hemoglobin level of 4.4 g/dL and we proceeded with immediate resuscitation. Assessments for GI bleeding (Meckel's scan, fiberoptic gastroduodenoscopy, and colonoscopy) did not reveal any specific focus of the abnormal bleeding. After

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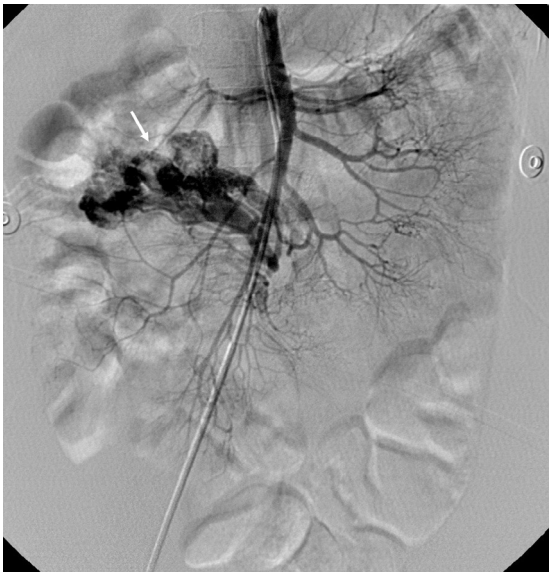

Fig. 1. Selective angiography of Case 1 shows an early venous drainage with vascular tangle in distal jejunal branch (white arrow).

stabilization, we were able to identify a focus of bleeding at the distal jejunum by selective angiography following a red blood cell (RBC) scan, which revealed tortuous and early venous return of distal jejunal branch (Fig. 1). An exploratory laparotomy through single-site umbilical laparoscopic approach was then performed, which revealed a portion of the jejunal segment involving the serosal surface to be hyperemic (Fig. 2). Approximately 15 cm of the jejunal segment including the hyperemic portion was resected. Pathology indicated an arteriovenous malformation with features of tortuous, engorged vascular structures on the serosal surface and congested mucosa along the antimesenteric border of intestine (Fig. 3).

2.2. Case 2

A 3-year-old girl visited the outpatient clinic, presenting with melena twice a month. For several months, she had been managed for an iron-deficiency anemia with initial hemoglobin level of 7.0 g/dL following a severe upper respiratory infection, which was remedied with oral iron supplements. At the time, the patient presented with intermittent vomiting, without any other GI symptoms. Assessments for melena (Meckel's scan, fiberoptic

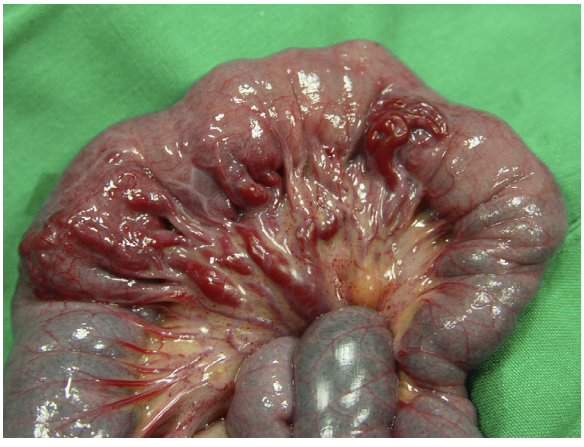

Fig. 2. Gross appearance shows tortuous vascular branch around mesenteric surface of the jejunum.

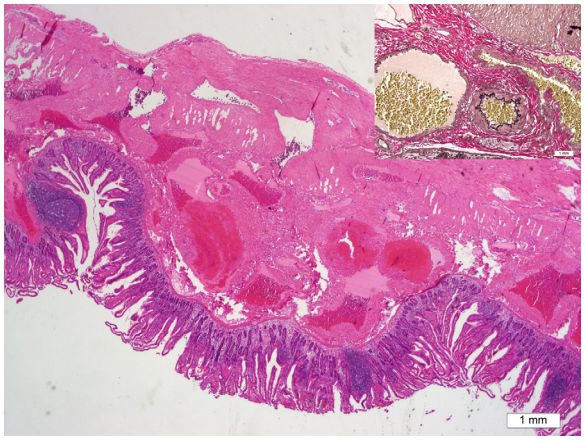

Fig. 3. Multiple dilated vascular structures of variable sizes and the thickness of the wall affecting the bowel submucosa, muscularis propria, and subserosa (H&E stain, ×20). In inlet, the abnormally dilated vein (left side of image) and artery (right side of image) (Elastic fiber stain, ×100).

gastroduodenoscopy, colonoscopy, and RBC scan) revealed no focus of the GI bleeding. Planned a computed tomographic (CT) angiography after having failed a capsule endoscopy, however, it showed segmental, circumferential wall thickening of the small bowel with multiseptated cystic lesions in the thickened wall, and multifocal punctate enhancement on portal venous phase, involving more than 10 cm in length (Fig. 4). Single-site umbilical laparoscopic surgery was then carried out and we identified two separate lesions, one large and one small, on the jejunum (Fig. 5). Approximately 20 cm of the jejunal segment, including the portion with large lesion, was resected, and wedge-resection of small discrete lesion was performed. Pathology revealed an arteriovenous malformation with dilated blood vessels in the submucosa, muscularis propria, and subserosa (Fig. 3).

3. Discussion

There are only a few existing reports about vascular malformations of the small intestine in children [1–3]. Vascular malformations can affect all parts of the body, and those in the skin and soft tissue tend to more common in children. Conversely, visceral involvement of the abdomen is relatively rare in comparison. There are four types of vascular malformations involving the abdominal viscera; lymphatic malformation (LM), capillary malformation (CM), venous malformation (VM), and arteriovenous malformation (AVM) [4,5]. Intra-abdominal LMs usually arise from the mesentery, omentum, or retroperitoneum, but symptoms may not manifest until adulthood. CM and AVM in the abdomen are rarely encountered in clinical practice. However, VM in the abdomen often presents with GI bleeding [6,7], and appears to result from mucosal vascular abnormality associated with mucosal ulcers. The cases reported here, also revealed LGI bleeding due to vascular malformations of the small intestine. Lesions in GI tract may cause a chronic obscure bleeding followed by anemia as shown in our cases, whereas it could cause spontaneous, significant LGI bleeding at first. We could assume that these vascular malformations were causes of child's anemia in our cases.

Vascular malformations at body surface can be diagnosed based on patient's history and physical examination; nevertheless, appropriate imaging studies are still necessary in most cases. Although proper diagnosis is crucial for appropriate treatment, the diagnosis of intestinal vascular malformations is occasionally difficult due to the limitations associated with diagnostic methods [8,9]. In cases of LGI bleeding, a GI endoscopy or RBC scan is usually performed to localize the bleeding focus, but in some cases, this may not

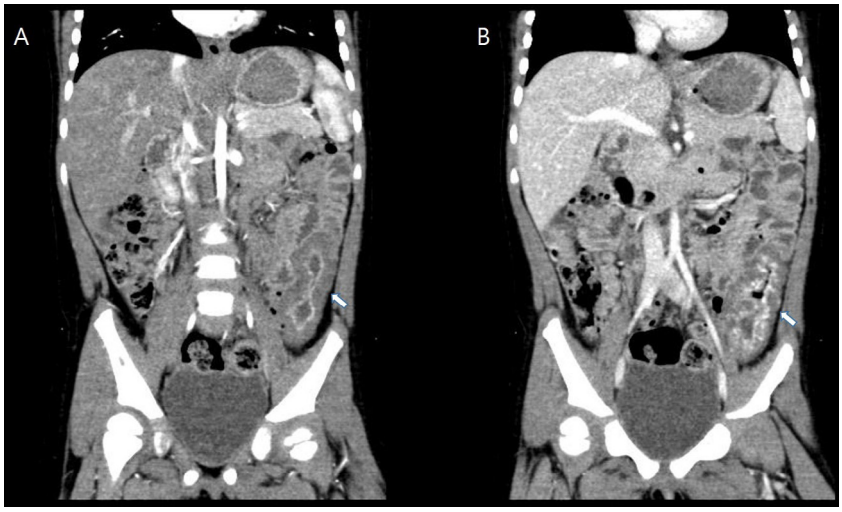

Fig. 4. Enhanced CT angiography of Case 2 shows a segmental, circumferential wall thickening (A) and multifocal punctate enhancement on portal venous phase (B).

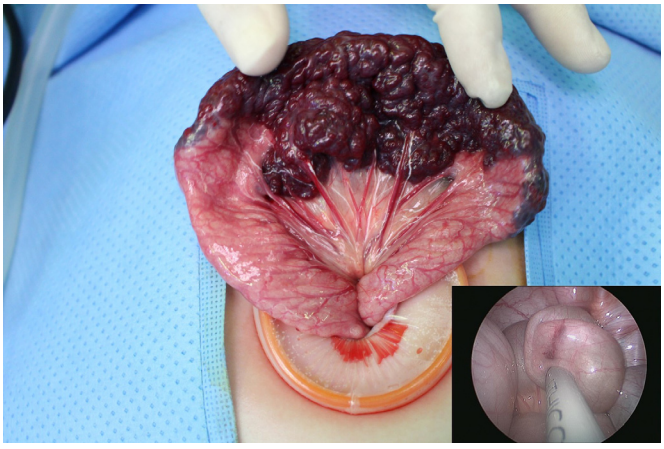

Fig. 5. Gross appearance shows a markedly dilated, engorged vascular structure encircling the wall of small intestine and a small discrete lesion apart from the main lesion in inlet.

adequate. Alternatively, a selective angiography could be more helpful to diagnose and determine the location of the lesions. When the bleeding ceases temporarily, it is recommended that an abdominal vascular enhanced CT or capsule endoscopy be performed [10,11]. In the present study, selective angiography was useful in one case and CT enterogram with angiography was useful in the other case. GI lesions could be associated with the following syndromes: the Klippel-Trenaunay syndrome, and the blue rubber bleb nevus syndrome. These syndrome usually encompass vascular malformations as a skin manifestation, so the possibility of visceral lesion may be suspected. In our cases, however, the malformations were a unique manifestation without any associated syndrome.

The management for symptomatic visceral vascular malformations depends on the specific type of lesion, anatomical location, and presenting symptoms [6,7]. There are many non-operative therapeutic modalities such as pharmacotherapy, intravascular embolization, endoscopic sclerotherapy or banding, however, the treatment of intestinal lesions still involve surgical resection in cases with severe bleeding. When considering surgical management, it can be challenging to localize the lesion intraoperatively, but it might be a good choice to apply the minimal invasive approach like as our cases. Additionally, some lesions cannot be removed completely. However, if lesions are amenable to complete removal, surgical resection is curative. Fortunately, in the present cases, we encountered positive results with complete removal of the lesions following proper intraoperative localization.

In this report, we treated two cases of vascular malformations occurring at the jejunum, one with a single lesion and the other with multiple lesions, with successful minimal invasive procedure and there were no intraoperative complications and no recurrence during follow-ups.

4. Conclusion

It is necessary to consider a vascular malformation as one of the causes when managing chronic anemia with intermittent, unclear GI bleeding in pediatric patients. If possible, a multidisciplinary approach is more appropriate owing to the rarity and complexity of visceral vascular anomalies. Above all, we could suggest a minimal invasive procedure when needs a surgical management for a pediatric patient.

Conflicts of interest

No conflicts of interest.

Funding

No financial relationship to disclose.

Ethical approval

No ethical approval is required to publish this report.

Consent

Informed consent was obtained from the patient's parents and all data were managed with personal information protection.

Author contribution

YH Cho made a conception and design of this study. SH Kim wrote the first draft of the manuscript. YH Cho and HY Kim contributed to the drafting of manuscript and critical revision. All authors reviewed and approved the final version of manuscript.

Guarantor

The guarantor is identical to corresponding author.

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