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

Soo-Hong Kim, Yong-Hoon Cho *, Hae-Young Kim

Division of Pediatric Surgery, Department of Surgery, Pusan National University Yangsan Hospital, Yangsan, Republic of Korea

ARTICLE INFO

Received 26 December 2016
Received in revised form 23 January 2017
Accepted 26 January 2017
Available online 30 January 2017

Keywords:
Vascular malformations
Small intestine
Pediatric
Minimal invasive

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.

© 2017 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

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

* Corresponding author at: Department of Surgery, Pusan National University School of Medicine, Geumo-ro 20, Mulgeum-eup, Yangsan, Gyeongnam-do, 50612, Republic of Korea.

E-mail address: choyh70@pusan.ac.kr (Y.-H. Cho).

http://dx.doi.org/10.1016/j.jijscr.2017.01.057
2210-2612/© 2017 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).
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 man-
aged 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
gastroduodenoscopy, colonoscopy, and RBC scan) revealed no focus
of the GI bleeding. Planned a computed tomographic (CT) angi-
ography after having failed a capsule endoscopy, however, it showed
segmental, circumferential wall thickening of the small bowel with
multisected 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 malforma-
tions of the small intestine in children [1–3]. Vascular malforma-
tions 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 com-
parison. There are four types of vascular malformations involving
the abdominal viscera; lymphatic malformation (LM), capillary
malformation (CM), venous malformation (VM), and arterio-
venous 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 malforma-
tions 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, ap-
propriate 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. 1. Selective angiography of Case 1 shows an early venous drainage with vascular
tangle in distal jejunal branch (white arrow).

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).
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.

References

[1] U.S. Karnam, J.S. Barkin, Vascular malformations of the small intestine, Curr. Treat. Options Gastroenterol. 4 (2001) 173–179.
[2] A. Handra-Luca, E. Montgomery, Vascular malformations and hemangiolymphangiomias of the gastrointestinal tract: morphological features and clinical impact, Int. J. Clin. Exp. Pathol. 4 (2011) 430–443.
[3] B. Fremond, S. Yazbeck, J. Dubois, P. Brochu, L. Garel, A. Ouimet, Intestinal vascular anomalies in children, J. Pediatr. Surg. 32 (1997) 873–877.
[4] J.B. Mulliken, J. Glowacki, Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics, Plast. Reconstr. Surg. 69 (1982) 412–422.
[5] O. Enjolras, J.B. Mulliken, Vascular tumors and vascular malformations (new issues), Adv. Dermatol. 13 (1997) 375–423.
[6] S.J. Fishman, P.E. Burrows, A.M. Leichtner, J.B. Mulliken, Gastrointestinal manifestations of vascular anomalies in childhood: varied etiologies require multiple therapeutic modalities, J. Pediatr. Surg. 33 (1998) 1163–1167.
[7] R. Dasgupta, S.J. Fishman, Management of visceral vascular anomalies, Semin. Pediatr. Surg. 23 (2014) 216–220.
[8] E. Bollinger, D. Raines, P. Sahtta, Distribution of bleeding gastrointestinal angioectasias in a Western population, World J. Gastroenterol. 18 (2012) 6235–6239.
[9] C.E. Dye, R.R. Gaffney, T.M. Dykes, M.T. Moyer, Endoscopic and radiographic evaluation of small bowel in 2012, Am. J. Med. 125 (2012), 1228.e1–1228.e12.
[10] L.K. Onal, M. Akdogan, M. Arhan, Z.M. Yalinkilic, B. Cicek, S. Kacar, et al., Double balloon enteroscopy: a 3-year experience at a tertiary care center, Hepatogastroenterology 59 (2012) 1851–1854.
[11] J.E. Huprich, J.M. Barlow, S.L. Hansel, J.A. Alexander, J.L. Fidler, Multiphase CT enterography evaluation of small-bowel vascular lesions, Am. J. Roentgenol. 201 (2013) 65–72.

Open Access
This article is published Open Access at sciencedirect.com. It is distributed under the IJSCR Supplemental terms and conditions, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.