

Giant Cystic Arteriovenous Malformation of the Mesentery and the Role of Cross-Sectional Imaging in Occult Gastrointestinal Bleeding

Adarsh M. Thaker, MD¹, Felicia Allard, MD², Jeffrey Goldsmith, MD², Martin Smith, MD³, Douglas Horst, MD⁴, and Elliot B. Tapper, MD⁴

¹Department of Medicine, Beth Israel Deaconess Medical Center, Boston, MA

²Department of Pathology, Beth Israel Deaconess Medical Center, Boston, MA

³Department of Radiology, Beth Israel Deaconess Medical Center, Boston, MA

⁴Division of Gastroenterology and Hepatology, Beth Israel Deaconess Medical Center, Boston, MA

Abstract

A 29-year-old woman presented with profound iron-deficiency anemia. Cross-sectional imaging identified a mass in the ileal mesentery. Surgical resection was curative and revealed a giant cystic arteriovenous malformation. Our report highlights the role of cross-sectional imaging in the evaluation and management of iron-deficiency anemia and obscure gastrointestinal hemorrhage.

Introduction

Occult gastrointestinal (GI) bleeding, or GI bleeding without an identifiable cause after initial endoscopic evaluation, is an important and often frustrating condition for gastroenterologists and patients alike. Repeat esophagogastroduodenoscopy (EGD) and colonoscopy are indicated to ensure that proximal and distal lesions are not missed.¹ However, many causes of obscure bleeding arise out of reach from these modalities, between the ligament of Treitz and the ileocecal valve. We demonstrate that cross-sectional imaging is a useful tool to evaluate occult bleeding.

Case Report

A 29-year-old pregnant woman presented with subacute abdominal pain, weakness, and nausea. Laboratory tests revealed hemoglobin 4.5 g/dL (mean corpuscular volume 68 fL) and ferritin 1.6 ng/mL. She had no history of menorrhagia or overt GI bleeding. She was believed to have chronic iron losses due to prior vaginal bleeding exacerbated by physiologic anemia of pregnancy. Her anemia was responsive to oral and intravenous iron supplementation and stabilized at 11.8 g/dL. Sixteen months after an uncomplicated delivery, she presented again with symptomatic anemia. She had an unremarkable EGD with normal gastric and duodenal biopsies. Colonoscopy with visualization of the terminal ileum found old blood throughout the colon. She was lost to follow-up for 8 months, then presented again with hemoglobin 5.8 g/dL and melena. Capsule endoscopy revealed old blood in the distal small bowel but did not identify a source. Abdominal computed tomography (CT) showed multifocal fluid filled lesions within the ileal mesentery with thin, faintly enhancing walls (Figure 1). Some lesions involved the ileal wall, extending to the lumen without perceptible overlying mucosa. Retrograde single-balloon enteroscopy was considered; however, in view of the worrisome mass lesions, the patient opted to pursue surgical resection.

ACG Case Rep J 2015;2(3):158-160. doi:10.14309/crj.2015.40. Published online: April 10, 2015.

Correspondence: Elliot Tapper, Beth Israel Deaconess Medical Center, Department of Medicine, 330 Brookline Avenue, Boston, MA 02115 (etapper@bidmc.harvard.edu).



Copyright: © 2015 Thaker et al. This work is licensed under a Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License. To view a copy of this license, visit <http://creativecommons.org/licenses/by-nc-nd/4.0>.



Figure 1. (A and B) Axial and (C and D) coronal abdominal CT showing multifocal fluid attenuation lesions within the ileal mesentery. Very thin, faintly enhancing walls (solid white arrows) do not appear to have mass effect upon adjacent vessels. In several areas (open white arrows), these lesions involve the wall of the ileum and in some cases appear to extend to the lumen with no perceptible overlying mucosa. A single punctate calcification is noted within one of the lesions (grey arrowhead).

Laparotomy with interoperative enteroscopy revealed a diffuse area of mucosal bleeding near masses 65 cm proximal to the ileocecal valve. An ileal segment including the masses was resected (Figure 2). Histology revealed an abnormal proliferation of dilated, smooth-walled, benign lymphovascular spaces of variable size extending from the mesentery to the intestinal epithelium (Figure 3). The walls varied from thin lymphatic channels to thick, muscular veins and arteries, consistent with a massive arteriovenous malformation (AVM). Evidence of malignant vascular proliferation (e.g., angiosarcoma) was not identified. One year after surgery, she was well with resolved anemia and no evidence of bleeding.

Discussion

The recommended evaluation of obscure gastrointestinal bleeding and iron deficiency begins with upper endoscopy

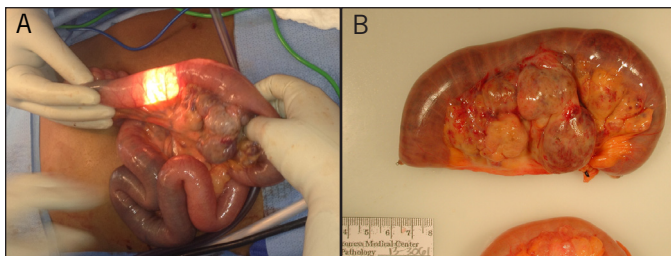


Figure 2. (A) The massive AVM is shown in the center during laparotomy with enteroscopy. (B) Gross specimen of the removed massive AVM.

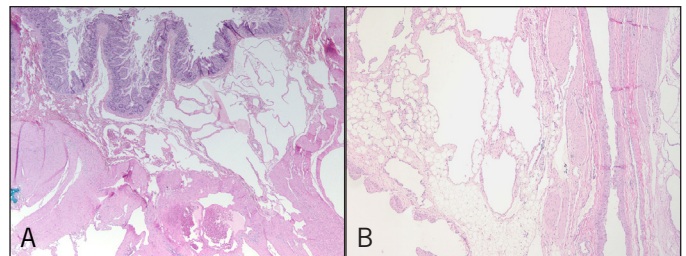


Figure 3. (A) Histologically unremarkable ileal epithelium is seen overlying a mass of confluent, benign lymphovascular channels. (B) The lesion is composed of lymphovascular channels of various caliber and various wall composition, ranging from thin-walled lymphatic spaces (confirmed by D2-40 immunostain) to thick, muscular walls demonstrated in the deeper subserosal portion of the ileal wall.

and colonoscopy.¹ A negative evaluation warrants subsequent investigation of the small bowel. Contemporary options include capsule endoscopy, deep enteroscopy, and advanced cross-sectional enterography. Deep enteroscopy offers interventions, but is time-consuming and highly specialized. Capsule endoscopy offers reliable small bowel visualization, but lacks therapeutic potential. Cross-sectional imaging contributes to the evaluation of occult bleeding because it can localize active bleeding lesions through angiography and can identify submucosal mass-lesions and transmural processes that may be missed by capsule endoscopy.² In our case, retrograde balloon-assisted enteroscopy may have detected a submucosal process following the failure of the capsule endoscopy. However, an operation was planned for the masses and enteroscopy would not have allowed our patient to avoid resection.

Our patient's iron deficiency was caused by and resolved after resection of a massive mesenteric AVM. GI AVMs comprise a spectrum of vascular lesions ranging from flat mucosal lesions that contain thick-walled arteries to masses of thin-walled capillaries that are best described as hemangiomas. The latter appear in the lumen with a bluish discoloration and rarely bleed. The literature regarding massive mesenteric AVMs presenting as GI bleeding is comprised of 20 intestinal hemangiomas.³⁻⁴ Our patient's lesion possessed thick-walled arteries, distinguishing it from a hemangioma as a true AVM. There have been reports of arterial-portal fistulas that are low-flow, low-pressure arteriovenous malformations that form in response to injury (penetrating trauma or postoperative changes).⁵ These lesions present with GI bleeding with crampy abdominal pain and an audible abdominal bruit. Our case is unique because it was a spontaneous, massive mesenteric true AVM discovered in the work-up of anemia. Mass lesions are a critical part of the obscure bleeding differential after negative endoscopy. Our patient's cross-sectional imaging discovered a mesenteric arteriovenous malformation for which resection was curative of her iron deficiency.

Disclosures

Author contributions: AM Thaker wrote the manuscript. F. Allard and J. Goldsmith performed the pathology analysis and revised the manuscript. M. Smith performed the radiology analysis and revised the manuscript. D. Horst revised the manuscript. EB Tapper oversaw the study, revised the manuscript, and is the article guarantor.

Financial disclosure: None to report.

Informed consent was obtained for this case report.

Received: November 18, 2014; Accepted: March 19, 2015

References

1. Fisher L, Lee Krinsky M, Anderson MA, et al; ASGE Standards of Practice Committee. The role of endoscopy in the management of obscure GI bleeding. *Gastrointest Endosc*. 2010;72(3):471–9.
2. Milano A, Balatsinou C, Filippone A, et al. A prospective evaluation of iron deficiency anemia in the GI endoscopy setting: Role of standard endoscopy, videocapsule endoscopy, and CT-enteroclysis. *Gastrointest Endosc*. 2011;73(5):1002–1008.
3. Sylla P, Deutsch G, Luo J, et al. Cavernous, arteriovenous, and mixed hemangioma-lymphangioma of the rectosigmoid: Rare causes of rectal bleeding—Case series and review of the literature. *Int J Colorectal Dis*. 2008;23(7):653–8.
4. Yang GZ, Li J, Jin H. Giant mesenteric hemangioma of cavernous and venous mixed type: A rare case report. *BMC Surg*. 2013;13(1):50.
5. Anderson RD, Liebeskind A, Lowman RM. Arteriovenous fistula of the mesentery. *Am J Gastroenterol*. 1972;57(5):453–8.

Publish your work in ACG Case Reports Journal

ACG Case Reports Journal is a peer-reviewed, open-access publication that provides GI fellows, private practice clinicians, and other members of the health care team an opportunity to share interesting case reports with their peers and with leaders in the field. Visit <http://acgcasereports.gi.org> for submission guidelines. Submit your manuscript online at <http://mc.manuscriptcentral.com/acgcr>.