ACG CASE REPORTS JOURNAL

CASE REPORT | SMALL BOWEL

Intravascular Papillary Endothelial Hyperplasia of the Jejunum as a Cause of Obscure-Overt Gastrointestinal Bleeding

Jonathan Gotfried, MD[1], Raul Berio Dorta, MD[2], and Jennifer Maranki, MD[3]

[1]*Department of Gastroenterology, Temple University Hospital, Philadelphia, PA*
[2]*Virtua Medical Group, Cherry Hill, NJ*
[3]*Division of Gastroenterology, PennState Health, Milton S. Hershey Medical Center, Hershey, PA*

ABSTRACT

Intravascular papillary endothelial hyperplasia (IPEH), or Masson's tumor, is a rare, benign vascular lesion characterized by reactive endothelial proliferation with venous stasis and thrombus. Lesions typically arise in vascular regions of the head and neck or extremities, but involvement of other organ systems has been reported. Clinically, IPEH mimics other benign lesions (hemangiomas) and malignant neoplasms (angiosarcoma) depending on the affected site. Diagnosis is essential because local excision of certain IPEH subtypes is curative. IPEH in the gastrointestinal tract is rare with few reported cases. We present a case of obscure-overt gastrointestinal bleeding from IPEH of the jejunum.

INTRODUCTION

Intravascular papillary endothelial hyperplasia (IPEH) is a rare, benign vascular lesion that usually occurs in the head and neck or extremities. IPEH is a reactive process of endothelial proliferation around thrombi in the setting of venous stasis.[1] IPEH can occur in other organ systems, and clinical manifestations depend on the affected site. Six cases of IPEH of the small intestine have been reported.[2-6]

CASE REPORT

A 43-year-old woman presented with 2 days of nausea, vomiting, and abdominal pain. She had a history of endometrial cancer at stage IB in remission for 3 years and provoked pulmonary embolism 3 years prior to presentation, for which she had completed 6 months of oral anticoagulation. Immediately prior to presentation, she had one episode of melena followed by hematochezia. She denied nonsteroidal antiinflammatory drug or alcohol use. Vital signs at presentation were stable, and physical examination was unremarkable for stigmata of liver disease. Initial hemoglobin was 9.8 g/dL and decreased to 6.5 g/dL over 6 hours. An esophagogastroduodenoscopy on the day of admission revealed patchy erythematous mucosa in the gastric fundus without an identified source of bleeding. She remained stable and underwent colonoscopy on hospital day 2 that found old blood throughout the entire examined colon and terminal ileum without a localized source of bleeding.

Early on hospital day 3, the patient experienced an isolated, self-limited episode of hematochezia and hypotension and received 2 units of packed red blood cells. As the patient stabilized following transfusion, a decision was made to proceed with upper double-balloon enteroscopy (DBE) for the initial evaluation of suspected small bowel bleeding. A bleeding, proximal jejunal mass 3 cm in size was identified as the source of bleeding (Figure 1). Biopsies of the lesion were not obtained due to concern of bleeding risk given the vascular appearance of the lesion. The area adjacent to the mass was tattooed in anticipation of surgical removal.

ACG Case Rep J 2017;4:e80. doi:10.14309/crj.2017.80. Published online: June 21, 2017.

Correspondence: Jonathan Gotfried, Department of Gastroenterology, 3401 N. Broad St., Philadelphia, PA 19140 (jonathan.gotfried@tuhs.temple.edu).

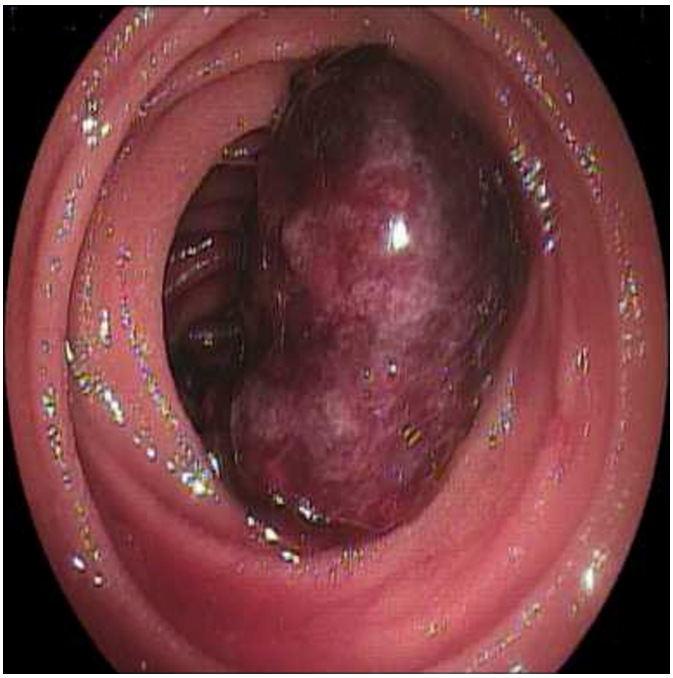

Figure 1. DBE revealing a large, vascular, oozing mass in the jejunum.

The patient remained stable and underwent laparotomy on hospital day 4. A segment of tattooed bowel was identified 30 cm distal to the ligament of Treitz, and successful resection and primary anastomosis was performed. Macroscopic examination of the specimen revealed a single, 3-cm polyp with adjacent mucosal edema and hemorrhage. Examination of a frozen section revealed focal transmural hemorrhage and surrounding histiocytic reaction favoring a vascular etiology. Microscopic examination demonstrated focal transmural hemorrhage with endothelial papillary proliferation and hyalinized fibrous cores, consistent with IPEH. There was no evidence of malignancy (Figure 2). The patient's postoperative course was uneventful, and she was discharged in stable condition with regular primary care follow up.

DISCUSSION

IPEH is a rare, benign, vascular lesion likely resulting from reactive endothelial proliferation in response to vascular stasis or trauma. First described by Masson in 1923, the entity was hypothesized to be a neoplasm with associated thrombi.[7] In 1932, Henschen clarified the disease to be reactive in nature with endothelialization around fragments of thrombi.[8] In 1976, Clearkin and Enzinger termed the condition "intravascular papillary endothelial hyperplasia," reflecting the likely pathophysiology.[9]

IPEH is categorized into three types: pure, mixed, and extravascular. The pure form occurs de novo in a dilated vascular space with no causative etiology; the mixed form

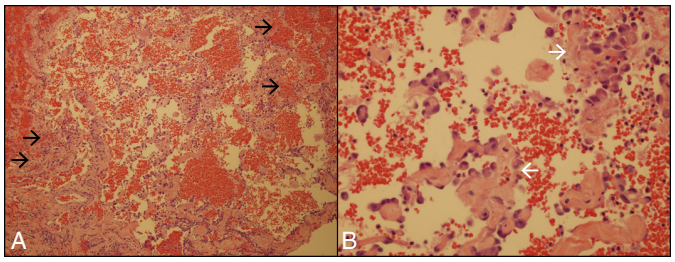

Figure 2. (A) Areas of hemorrhage and thrombus juxtaposed to areas with collagenized structures and ingrowth of endothelium (arrows). (B) Intravascular papillary endothelial hyperplasia showing papillary formation with hyaline stalks covered by plump endothelial cells (arrows). There was no significant nuclear atypia, tumor cell necrosis, or mitotic activity to suggest angiosarcoma.

is a superimposed lesion over a preexisting vascular abnormality (arteriovenous malformation or hemangioma); and the extravascular form, which is least common, occurs with endothelial proliferation at a hematoma after trauma. Depending on the affected site, clinical features of IPEH may mimic other benign (hemangiomas, pyogenic granuloma), infectious (bacillary angiomatosis, Kaposi's sarcoma), or malignant lesions (angiosarcoma).

IPEH usually occurs in the vascular regions of the head and neck or in extremities, although other organ systems have been reported. A literature review revealed 23 reported cases of IPEH arising within the abdominal cavity, including the urogenital tract, retroperitoneum, adrenal glands, and liver.[10-13] Six cases of IPEH within the gastrointestinal lumen have been reported: 2 in the duodenum and 4 in the jejunum.

A possible hormonal etiopathogenesis of IPEH has been proposed in previous reports.[5,14] The higher incidence in females with IPEH further supports this hypothesis. The subtype of IPEH in our case is unknown, as a hypercoagulable workup and exclusion of endometrial cancer recurrence were not done during the patient's stay.

Our patient's lesion was directly visualized during upper DBE. A case of IPEH diagnosed with video capsule endoscopy was recently reported.[4] While video capsule endoscopy and angiography are available at our institution, DBE was utilized given its immediate availability at the time of the patient's bleeding episode and response to resuscitation. DBE allowed us to mark the lesion in anticipation of surgical removal and exclude synchronous lesions.

Prognosis varies between IPEH types, with the pure IPEH type considered curative after complete removal whereas recurrence has been reported with mixed types.[1] As this case illustrates, clinicians should be careful during the workup of obscure-overt gastrointestinal bleeds and consider IPEH when treating those patients. Prompt identification and

resection is curative, with low rates of recurrence and excellent patient outcomes in most cases.

DISCLOSURES

Author contributions: All authors contributed equally to the manuscript. J. Maranki is the article guarantor.

Financial disclosure: None to report.

Attempts to contact the patient for consent were unsuccessful. All identifying information has been removed to protect patient privacy.

Acknowledgements: The authors wish to thank Gary Kirshenbaum, MD, Clinical Professor of Pathology at Advocate Illinois Masonic Medical Center for pathology support.

Received December 7, 2016; Accepted April 4, 2017

REFERENCES

1. Hashimoto H, Daimaru Y, Enjoji M. Intravascular papillary endothelial hyperplasia: A clinicopathologic study of 91 cases. *Am J Dermatopathol*. 1983;5(6):539–46.
2. Okauchi S, Kurooka N, Nagata J, et al. Arteriovenous malformation associated with intravascular papillary endothelial hyperplasia in jejunal region. A case report (author' transl). *Rinsho Hoshasen*. 1982;27(1):139–42.
3. Dunphy CH, Sotelo-Avila C, Luisiri A, Chu JY. Paroxysmal nocturnal hemoglobinuria associated with venous thrombosis and papillary endothelial hyperplasia presenting as ulcerated duodenal mass. *Arch Pathol Lab Med*. 1994;118(8):837–9.
4. Mestiri S, Karoui M, Charachon A, Deux JF, Chaumette MT. Intravascular papillary endothelial hyperplasia of the jejunum: An unusual cause of melena. *Int J Surg Pathol*. 2007;15(2):192–5.
5. Meadows MC, Sun X, Dardik M, Tarantino DR, Chamberlain RS. Intraabdominal intravascular papillary endothelial hyperplasia (Masson's tumor): A rare and novel cause of gastrointestinal bleeding. *Case Rep Gastroenterol*. 2010;4(1):124–32.
6. Lippai D, Madácsy L, Baló T, et al. Mass-like lesions including a rare case of melena causing Masson's hemangioma among the capsule endoscopy cases of the Semmelweis University-2nd department of internal medicine during 2014. *Z Gastroenterol*. 2015;53(05):A30.
7. Masson P. Hemangioendothelioma vegetant intravasculaire. *Bull Soc Anat*. 1923;93:517–23.
8. Henschen F. L'endovasculite proliferante thrombopoietique dans la lesion vasculaire locale. *Ann Anat Pathol*. 1932;9:113–21.
9. Clearkin KP, Enzinger FM. Intravascular papillary endothelial hyperplasia. *Arch Pathol Lab Med*. 1976;100(8):441–4.
10. Garber BB, Prestipino AJ, Pollack HM, Levine SR, Whitmore KE. Masson's tumor of the kidney: A new renal lesion. *J Urol*. 1990;143 (2):344–6.
11. Petry M, Brown MA, Hesselink JR, Imbesi SG. Multifocal intravascular papillary endothelial hyperplasia in the retroperitoneum and spine: A case report and review of the literature. *J Magn Reson Imaging*. 2009;29 (4):957–61.
12. Kawashima A, Johsen T, Murayama S, Russell WJ. Intravascular papillary endothelial hyperplasia of the adrenal gland. *Br J Radiol*. 1986;59 (702):610–3.
13. Hong SG, Cho HM, Chin HM, et al. Intravascular papillary endothelial hyperplasia (Masson's hemangioma) of the liver: A new hepatic lesion. *J Korean Med Sci*. 2004;19(2):305–8.
14. Pins MR, Rosenthal DI, Springfield DS, Rosenberg AE. Florid extravascular papillary endothelial hyperplasia (Masson's pseudoangiosarcoma) presenting as a soft-tissue sarcoma. *Arch Pathol Lab Med*. 1993;117 (3):259–63.