

# Obscure Overt Gastrointestinal Bleeding Due To Isolated Small Bowel Angiomatosis

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## Abstract

Isolated small bowel angiomatosis is a rare entity with a distinctive endoscopic appearance. A multidisciplinary approach is often required to diagnose and treat these complex lesions. We present 2 cases of isolated small bowel angiomatosis, and illustrate the endoscopic findings that may guide similar diagnoses.

## Introduction

Small bowel angiomatosis is a rare type of vascular malformation, with only few cases reported in the English literature.<sup>1-4</sup> Angiomatosis is incompletely understood, characterized by diffuse excess of dilated, capillary-sized flat endothelial blood vessels in an otherwise normal vasculature.<sup>5</sup> Although vascular malformations are congenital, they may manifest at any time and may be part of clinical syndromes such as Rendu-Osler-Weber syndrome, Klippel-Trenaunay syndrome, or Sturge Weber Syndrome.<sup>5</sup> Isolated intestinal angiomatosis is a very rare entity, and diagnosis is often delayed by years of extensive negative work-up.<sup>1-4</sup>

## Case Report

**Case 1:** A 30-year-old man with Down syndrome presented with recurrent melena since age 10 years. He was maintained on oral, then IV, iron supplementation. Previous work-up included multiple negative esophagogastroduodenoscopies (EGDs), colonoscopies, and enteroscopies. Video capsule endoscopy (VCE) showed a bleeding spot in the proximal small bowel. Push enteroscopy identified multiple, large vascular malformations in the proximal jejunum not amenable to endoscopic therapy (Figure 1). The area was tattooed and clipped, and was easily identified during exploratory laparotomy. Intraoperatively, the small bowel had a very intense hyperemic appearance with large mucosal tortuous vessels encroaching around the serosa. Resection of the involved segment (8 cm of small bowel) and primary end-to-end anastomosis were performed. The rest of the small bowel was visualized, a similar hyperemic appearance was noted, and there was no tumor. Surgical pathology showed diffusely dilated and thick-walled capillaries within the mucosa, submucosa, and inner layer of the muscularis mucosa (Figure 2). The patient improved markedly after surgery and was symptom-free for almost 18 months.

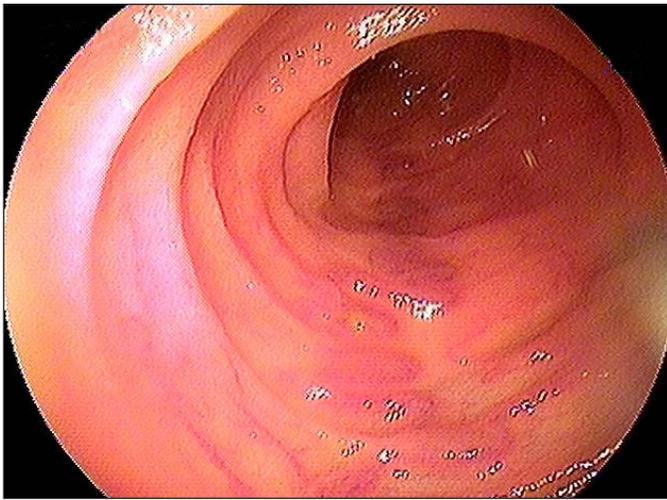
He then presented with increasing right upper quadrant pain with no bleeding. Abdominal computed tomography (CT) and magnetic resonance imaging (MRI) showed an 8 cm x 10 cm mass in the liver. Biopsies showed moderately differentiated angiosarcoma. The patient was started on chemotherapy with Taxol. Bleeding then recurred, and the Taxol was stopped within 1 month. Repeat VCE showed several vascular malformations in

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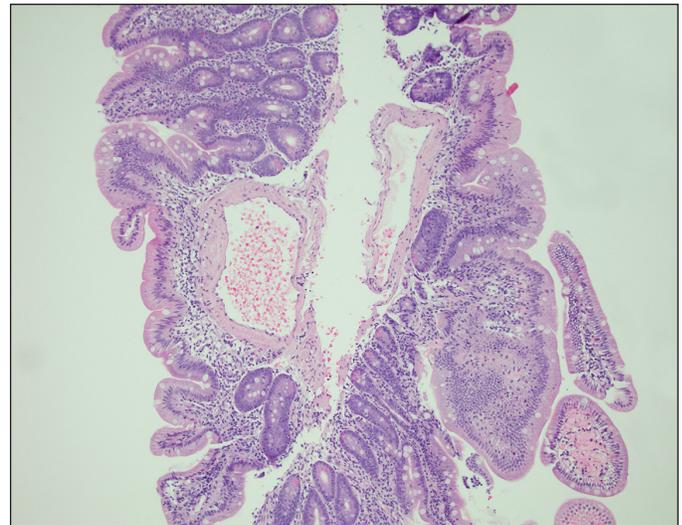
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**Figure 1.** Endoscopic view of the proximal jejunum showing multiple large tortuous ectatic vessels.

the distal small bowel. Anterograde double-balloon enteroscopy (DBE) showed patent enteroenterostomy in the jejunum 60 cm from the pylorus, and multiple bleeding superficial, tortuous ectatic vessels in the jejunum, mostly concentrated around the anastomosis. Coagulation for hemostasis using argon plasma coagulation (APC) was attempted. Biopsies showed large, intramucosal thick-walled vessels. Repeat anterograde DBE 1 month later for recurrent bleeding showed diffuse friable mucosa without active bleeding in the entire examined portion of the jejunum.

**Case 2:** A 25-year-old man with congenital asplenism presented with a 4-year history of recurrent melena requiring multiple admissions and blood transfusions. Previous work-up included multiple negative EGDs, colonoscopies, and enteroscopies. VCE showed multiple small vascular malformations without bleeding in the entire small bowel from the duodenum to ileum, built up in clusters in some areas. These appeared as superficial, ectatic, looping capillaries instead of typical angiodysplasias. Anterograde DBE showed hundreds of dilated, looping intraepithelial vessels with no bleeding in the entire examined portion of jejunum (Figure 3). The examination was discontinued and endoscopic therapy in the distal jejunum was not pursued due to the number and uniformity of the le-



**Figure 2.** Photomicrograph showing diffusely dilated and thick-walled capillaries within the jejunal mucosa (original magnification x 40).

sions. Biopsies showed unclassified vascular anomaly (large-caliber capillary-like vessels) present within the superficial mucosa of the intestine. Each vessel had a large diameter and a walls thicker than the adjacent lacteals. The rest of the examined small bowel mucosa was histologically unremarkable.

The patient later underwent exploratory laparotomy of the entire small bowel for recurrent bleeding. Uniform and diffuse extensive abnormal vascularity was observed and no resection was performed. The mesentery, stomach, liver, and colon appeared normal, and no spleen was found. Intraoperative liver biopsy showed preserved parenchymal architecture. The patient refused hormonal therapy but was referred to the transplant service for consideration for isolated small bowel transplantation. He finished part of the pre-transplant work-up before being incarcerated. No further follow-up is available.

## Discussion

Small bowel angiomas is usually localized in the jejunum and/or ileum, beyond the reach of standard endoscopes. VCE and DBE are the main tools for the evaluation of obscure gastrointestinal bleeding in hemodynamically stable patients



**Figure 3.** Endoscopic views of the jejunum showing multiple non-bleeding, dilated, and looping intra-epithelial vessels.

(after negative second-look endoscopies).<sup>4,6,7</sup> VCE can non-invasively visualize the entire small bowel, making it the first-line tool in many centers. Its limitations include capsule entrapment and the inability to provide endoscopic-guided therapy. DBE allows evaluation of the entire small bowel using a combination of both antegrade and retrograde routes. Current American College of Gastroenterology guidelines recommend performing VCE before deep enteroscopy to increase the diagnostic yield.<sup>7</sup> In acute overt massive gastrointestinal bleeding, conventional angiography should be performed emergently for hemodynamically unstable patients. In hemodynamically stable patients with evidence of active bleeding, multidetector CT (MDCT) angiography can be performed to identify the site of bleeding and guide further management.<sup>8</sup> Finally, intraoperative enteroscopy is a highly sensitive diagnostic and effective therapeutic procedure, but is invasive. Its usage should be limited to scenarios in which enteroscopy cannot be performed, such as patients with prior surgeries and intestinal adhesions.<sup>7</sup>

Given its rarity, the optimal management of obscure overt gastrointestinal bleeding due to intestinal angiomatosis is unknown. One case reported successful treatment of multiple patients with intestinal angiomatosis with thalidomide monotherapy.<sup>9</sup> The same study reported combined therapy with thalidomide and interferon alpha in cases of systemic angiomatosis of the stomach, intestine, mesentery, retroperitoneum, and mediastinum. As there are usually multiple foci throughout the small bowel, the role of endoscopic thermal therapy with APC is limited.<sup>10</sup> Surgical resection can be curative if the lesions are localized.<sup>10</sup> Finally, isolated small bowel transplantation has been reported.<sup>11</sup>

## Disclosures

Author contributions: II El Hajj prepared the manuscript, reviewed the literature, and is the article guarantor. M. Martinez, MV Chiorean, and GA Cote reviewed the manuscript.

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