

An Uncommon Cause for a Common Scenario: A Case of Hematochezia Caused by a Massive Cavernous Hemangioma

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Cavernous hemangiomas are among the rarest causes of gastrointestinal bleeding. Here, we report a case of hematochezia resulting from an extensive cavernous hemangioma involving the rectum. We also highlight the difficult therapeutic challenges that can be posed by these lesions.

Case Report

A 20-year-old woman with complaints of hematochezia of two days duration was evaluated. The patient had a history of cavernous hemangioma of the buttocks, left hip, and posterior thigh since the age of 3. Her hemangioma had been treated with approximately 40 percutaneous transcatheter procedures with both sclerosing agents and coil embolizations. These treatments were complicated by damage to the patient's left sciatic nerve and by subsequent left lower extremity paralysis.

Upon presentation to the hospital, the patient was hemodynamically stable and her hemoglobin was normal. On physical examination, it was noted that she had some bluish skin discoloration on the buttocks, as well as maroon blood on digital rectal examination. She subsequently underwent a colonoscopy that showed several polypoid, red, vascular-appearing lesions approximately 10 cm from the anal verge (Figure 1). No active bleeding was noted. An endoscopic ultrasound confirmed an extensive network of blood vessels (Figure 2). Computed tomography scan of the abdomen revealed a low-density 3.4-cm × 3.2-cm mass abutting the left wall of the rectum and extending from the soft tissues of the left pelvic floor to the soft tissues of the left hip (Figure 3). An angiogram

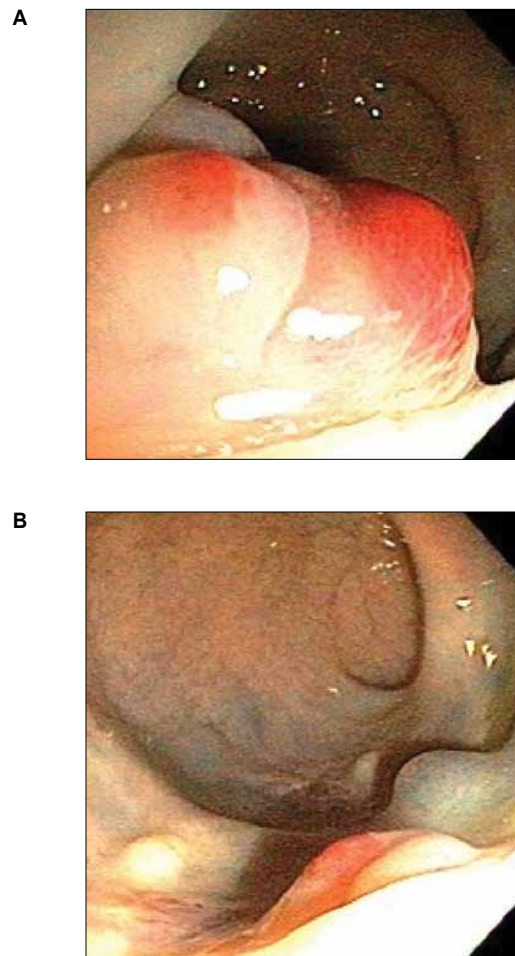


Figure 1. Colonoscopy showing several vascular-appearing lesions approximately 10 cm from the anal verge. Some lesions were polypoid and red (A), whereas other lesions had a bluish hue (B).

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Figure 2. Endoscopic ultrasound showing extensive network of blood vessels.

confirmed the vascular nature of the lesion but did not find any major arterial feeders to the hemangioma amenable to embolization.

The patient's case and potential treatment options were discussed with specialists from interventional radiology, surgery, and gastroenterology. It was agreed that, unfortunately, treatment would be very difficult given that her hemangioma had progressed in size despite 40 prior embolizations. Different surgical options, including a left hemicolectomy with colo-anal anastomosis; an abdominoperineal resection with colostomy; or mucosectomy procedures, were contemplated and discussed with the patient. It was decided that any surgical procedure would be very risky and bloody given the extent of hypervascularity in the area. Therefore, it was decided to follow her expectantly and to proceed with surgery only in the case of significant, recurrent, or life-threatening bleeding. The patient experienced no further bleeding episodes during the 5 days she was hospitalized, and she was discharged. In a follow-up visit 1 year later, the patient reported only intermittent minor bleeding episodes with no significant progression of symptoms and decided to continue to be followed expectantly.

Discussion

Described as early as the 1800s, hemangiomas have been known to be among the rarest causes of gastrointestinal bleeding.^{1,2} Hemangiomas may be classified as capillary, cavernous, or mixed types³ and are thought to be benign masses that have been present since birth.

Colonic hemangiomas usually present with occult bleeding or with slow painless hematochezia. However, massive gastrointestinal hemorrhages have also been known to occur, and a case report of cavernous hemangiomas presenting as bowel obstruction has also been reported.⁴ Most colonic hemangiomas occur in the left colon, though some may occur in the right colon.⁵



Figure 3. Computed tomography scan of the abdomen and pelvis demonstrating an ill-defined, low-density 3.4-cm × 3.2-cm mass abutting the left wall of the rectum. The lesion extended from the soft tissues of the left pelvic floor to the soft tissues of the left hip (A and B) and also showed some phlebolith formation (B).

The diagnosis is best established by endoscopic visualization of a blood-filled hemangioma that has an appearance of plum-red nodules or vascular congestion. Abdominal radiographs may show phlebolith formation. It has been proposed that phlebolith formation may be related to thrombosis in a cavernous hemangioma due to perivascular inflammation and stasis of blood flow.⁵⁻⁷ Computed tomography scans and magnetic resonance imaging can also be used for diagnosis and evaluation of the extent of the lesion. Computed tomography scans may reveal thickened colonic wall and pelvic phleboliths. Magnetic resonance imaging may show colon hemangiomas as bright heterogeneous signal intensity on T2 images.⁴

Small hemangiomas that are solitary can potentially be treated by endoscopic means. Amano and associates reported successful treatment of a 33-mm × 22-mm × 14-mm pedunculated, solitary cavernous hemangioma using endoscopic polypectomy.⁸ However, this is not effective on larger lesions. Embolization of the blood vessels has variable success and may not be effective in some cases. Surgical resection is generally required for larger lesions.^{9,10} Among the possible surgical approaches, abdominoperineal resection has been used when the perineum is involved and low anterior resection has been used when there is no perineal involvement.^{11,12} Several sphincter-saving mucosal resection techniques have also been proposed.¹³ Some have suggested that removal of the involved engorged friable rectal mucosa and use of a colo-anal sleeve anastomosis may control bleeding from rectal cavernous hemangiomas.^{13,14} The type of surgery performed should depend upon the surgeon's expertise and the extent and location of the lesion.

Our case presentation demonstrates a rare occasion of a very large hemangioma involving a significant portion of the body. Its large size made surgical resection difficult and risky, and the presence of multiple collateral vessels with limited arterial supply made embolization an option of limited value. Large hemangioma lesions represent a difficult therapeutic challenge.

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Review

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Hemangiomas of the large intestine are rare congenital lesions that come to clinical attention on even rarer occasions. These benign vascular hamartomas have been divided into two main types: capillary and cavernous hemangiomas. Capillary hemangiomas consist of a collection of submucosal vessels, similar in size to other capillaries, that are lined by a well-differentiated but hyperplastic endothelium. Most commonly found in the small intestine, appendix, and perianal skin,¹ they appear as bright-red, well-circumscribed, minimally compressible nodules, as they contain many closely packed, submucosal capillaries. These capillary hemangiomas tend to be clinically silent and typically represent incidental findings at surgery, endoscopy, or autopsy. However, slow oozing, possibly resulting in occult or obscure bleeding, can sometimes occur.²⁻⁴

Cavernous hemangiomas, on the other hand, consist of large, thin-walled, vascular channels arising from larger, submucosal vascular plexuses. They tend to have a loose, connective-tissue stroma with smooth-muscle fibers. These vascular networks also include blood-filled sinuses lined by single or multiple layers of endothelial cells. Bleeding from these lesions is often brisk, typically recurrent, and generally starts early in life, which is consistent with their congenital origin. Sometimes, bleeding diathesis can be worsened by consumption coagulopathy (Kasabach-Merritt syndrome) resulting from intravascular clotting within venous sinusoids of the hemangioma.⁵ The high risk of recurrent bleeding results from the lack of muscular and supportive elastic tissue in the walls of these abnormal vessels.⁶ However, small cavernous hemangiomas can also occur and sometimes a large number may occur throughout the gastrointestinal (GI) tract, a condition called multiple phlebectasia.^{6,7} These phlebectasias tend to be under 5 mm in size and are, thus, generally of no clinical importance. Mixed capillary-cavernous hemangiomas have also been described in the literature and may represent a transitional stage between the two more common types of hemangiomas.⁷

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Cavernous hemangiomas of the large intestine can also coexist with similar lesions in other parts of the GI tract, and occasionally there may be lesions at multiple levels of the alimentary canal, a condition called intestinal hemangiomatosis.⁶⁻⁸ Similarly, intestinal hemangiomas can also be associated with hemangiomas of other GI organs such as the gallbladder, liver, or pancreas, as well as non-GI viscera such as the kidney, spleen, or brain.^{7,9} However, the most commonly involved organ is the skin. In one study, half of the patients with intestinal hemangiomas had cutaneous involvement.¹⁰ Furthermore, there are syndromes in which GI and cutaneous hemangiomas coincide, including blue rubber bleb nevus syndrome and Klippel-Trenaunay-Weber syndrome.¹¹⁻¹³ Lastly, intestinal hemangiomas can be familial and frequently coexist with other congenital malformations.¹

Up to 80% of hemangiomas involving the colon and rectum are of the cavernous type,¹ and larger diffuse lesions may focally replace all enteric wall layers, possibly narrow the lumen, and even extend into other local structures. The majority, over 70%, of large intestinal hemangiomas occur in the rectum.^{7,14-16} Diagnosis often involves endoscopic imaging and angiography. Inconsistent awareness of the condition has also led to many delayed diagnoses. Specifically, recurrent bleeding may be attributed to hemorrhoids or inflammatory bowel disease, as proctoscopy often shows engorged veins and/or submucosal congestion causing nodularity or mucosal erosions.¹⁷

Treatment strategies for cavernous hemangiomas have generally relied on surgical intervention. On rare occasions, focal, discrete, polypoid-type hemangiomas can be treated with endoscopic polypectomy or injectable sclerotherapy. More often, surgical resection is required. Hemangioma size, location, and involvement of adjacent structures tend to determine the approach, extent of resection, sphincter-sparing, and need for intestinal diversion. As most cases involve rectal hemangiomas, difficult pelvic dissections are a common concern, as is the potential for abdominoperineal resection, which can be necessary when hemangiomas also involve perirectal pelvic structures. Luckily, most lesions are amenable to complete resection using sphincter-sparing anterior resection with colo-anal anastomosis.¹⁸ Non-resectional treatments have included injected sclerotherapy, cryosurgery, and local radiotherapy, as well as intravascular embolizations, operative vascular ligations, and even diverting colostomies. However, none of these techniques can reliably eradicate all of the vascular channels of a large cavernous hemangioma, and so the risk of rebleeding remains. Therefore, the use of these non-resectional therapies should be considered more anecdotal and/or as potential individualized options, not standard-of-care therapies.

The case reported by Kasher and colleagues highlights several of the common features of cavernous hemangiomas, as well as some interesting nuances.¹⁹ Their patient, who had lifelong symptoms related to her massive cavernous hemangioma, presented with acute hemorrhage per rectum and spontaneous resolution. The patient had already suffered adverse events related to prior percutaneous interventions, despite noble intentions of minimizing therapeutic morbidity. Having failed her many arterial embolizations, a difficult decision regarding potential extensive surgery was rendered. Although this commentary has focused on primary colorectal hemangiomas, which are more commonly encountered by gastroenterologists, this patient's hemangioma arose not from the rectum, but from the left pelvis, and incorporated multiple tissues. Unfortunately, palliation of such diffuse lesions almost invariably requires resection, and for more extensive lesions such as the one described, the morbidity can be great. Of course, although some patients will not be able to tolerate curative therapy, for many others, awareness, early diagnosis, and complete resection will prevent further bleeding events and yield improved quality of life.

Although there is no standard algorithm for the management of colorectal hemangiomas, specifically of the cavernous type, recognizing these syndromes is the most important step. Clues may come from the patient's clinical history, as well as from endoscopic visualization of varicosities or a wine- or plum-colored submucosal fullness. Biopsy should be avoided in this scenario given the risk for hard-to-control bleeding. The most useful step after preliminary diagnosis is complete mesenteric angiography, both to evaluate the extent of the primary lesion and to exclude concomitant hemangiomas elsewhere in the GI tract. Involvement of a surgeon is essential at that juncture to facilitate treatment planning. As stated above, the primary goal should be resection for cure, but various endoscopic, percutaneous, or surgical options may deserve consideration in complex cases.

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