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Necrotizing Fasciitis of the Upper Extremity: Case Reports and Description of the Disease Process

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Cavernous Hemangiomas of the Small Bowel: Case Reports and Literature Review

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ABSTRACT

Hemangiomas represent slightly more than 1% of all benign tumors of the small intestine. The cardinal symptom of a hemangioma is bleeding, which may be massive or occult. Diagnosis is frequently difficult. Treatment depends on the site and extent of the lesion. Surgical intervention may be necessary for recurrent or severe bleeding from a known location. Where possible, resection of the involved segment of intestine is the procedure of choice. Four cases of hemangiomas of the small intestine are described and a brief review of the literature is presented to heighten awareness of this entity as a cause of gastrointestinal tract bleeding of uncertain etiology.

INTRODUCTION

Vascular malformations represent approximately 5-16% of all benign bowel tumors.¹⁻³ Among all benign tumors of the small intestine, slightly more than 1% are hemangiomas. The diagnosis of hemangiomas of the small intestine can be difficult because of their anatomic location.⁴ The most common symptom is gastrointestinal tract bleeding. Hemangiomas should be included in the differential diagnosis of iron deficiency anemia caused by

chronic blood loss or acute gastrointestinal hemorrhage.

Four cases of hemangiomas of the small intestine are described and a review of the literature on this subject is presented.

CASE REPORTS

A review of the patients treated at our institution from 1976 to 1993 revealed eight cases of small bowel hemangiomas. The hemangioma was localized in the jejunum in three patients, in the ileum in two, and in the duodenum in two, and one patient had diffuse gastrointestinal hemangiomas. The eight patients included five males and three females whose mean age was 51.6 years (age range: 8-81 years). Hemangioma was an incidental finding in five patients. Four of these patients were undergoing laparotomy for other reasons: two for neoplastic lesions in the colon, one for a nonhealing gastric ulcer, and one for portal vein aneurysm and thrombosis. In one of the remaining four patients, a hemangioma of the duodenum was discovered during upper endoscopy performed to evaluate epigastric pain. In the remaining three patients, the tumors were symptomatic.

Case 1: The patient was a 73-year-old male with a six-month history of fatigue, weakness, recent syncope, and anemia. Hematologic evaluation performed elsewhere had revealed an iron deficiency anemia. The patient was treated with vitamin B12 and iron supplementation, but his anemia persisted and eventually required transfu-

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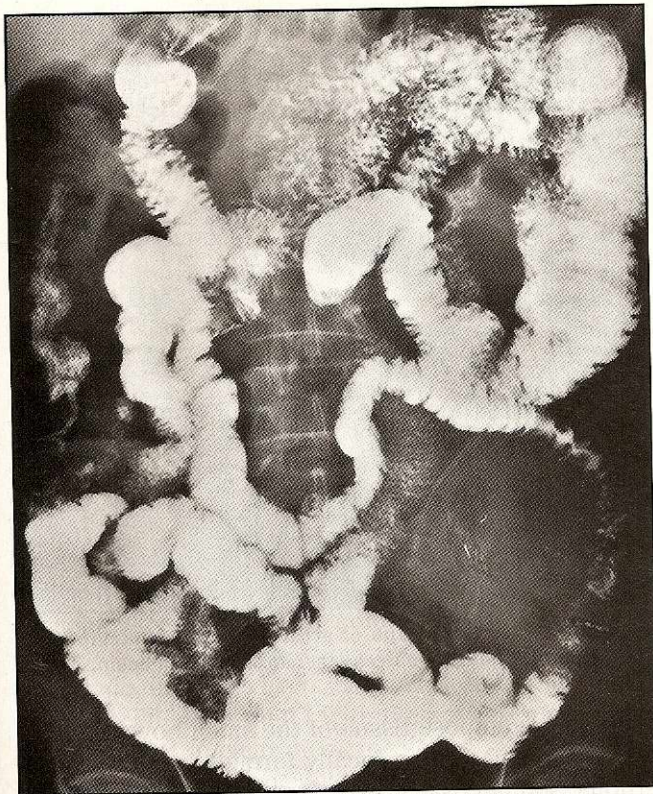


Figure 1: Small bowel study demonstrates a mass in the left lower quadrant within the small bowel mesentery with associated displacement of several small bowel loops.

sions. He was hospitalized because of persisting anemia and episodes of syncope.

The physical examination was unremarkable except for an irregular cardiac rhythm. Laboratory evaluation included the following: hemoglobin 7.9g/dL, hematocrit 24.4%, with normal white blood cell and platelet counts. A chest radiograph was normal. An electrocardiogram showed atrial fibrillation. An upper gastrointestinal tract barium contrast series with small bowel follow-through showed an area of irregularity in the proximal ileum, suggesting possible tumor (Figure 1). A CT scan of the abdomen disclosed a similar irregularity in the distal small intestine (Figure 2). Esophagogastroduodenoscopy revealed a hiatal hernia and mild antral and duodenal scarring and ulceration. No active bleeding sources were noted. Colonoscopy demonstrated a few small benign adenomatous polyps, which were removed. Arteriography was not performed.

Based on the preoperative suspicion of small intestine neoplasia (lymphoma versus carcinoid tumor), exploratory laparotomy was undertaken. The source of the patient's anemia was identified as a cavernous hemangioma in the proximal ileal mesentery that had

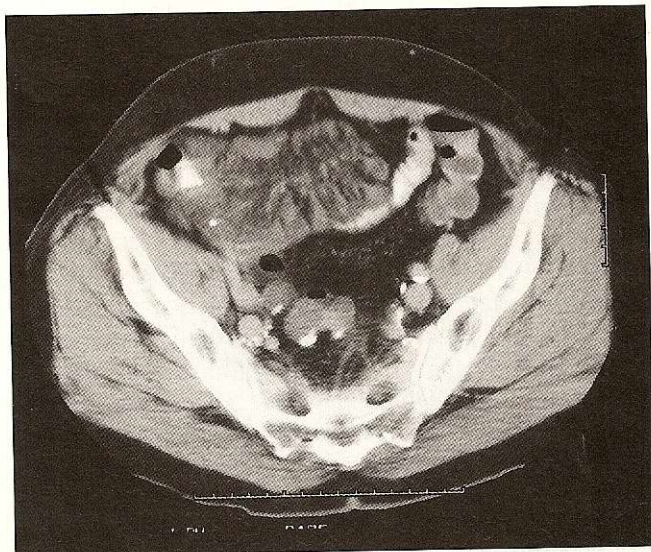


Figure 2: Mass consisting of soft tissue and fat within the small bowel mesentery displaces a small bowel loop inferiorly. The appearance is nonspecific and could represent a neoplasm or an inflammatory condition.

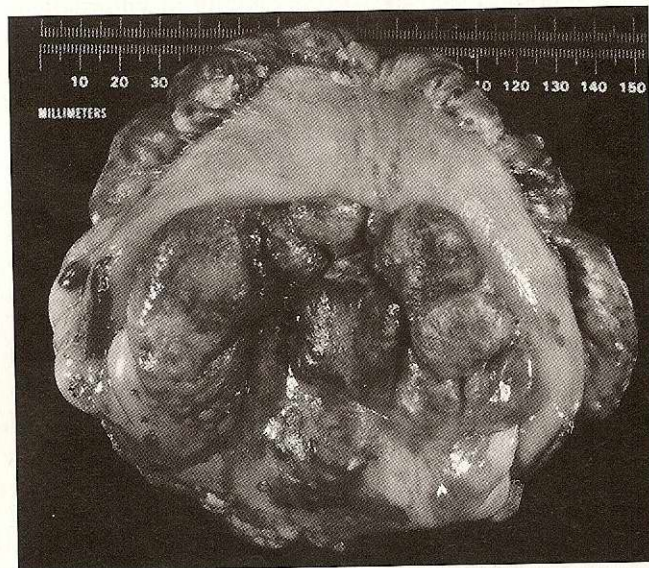


Figure 3: Gross pathology specimen shows a segment of proximal ileum and surrounding mesenteric vascular tumor.

eroded into the ileum. This portion of the small intestine, including the cavernous hemangioma, was resected. The patient's postoperative course was uneventful, and he was discharged on postoperative day six.

Gross pathologic examination revealed a well-circumscribed 9.0cm vascular tumor of the small bowel mesentery (Figure 3). Microscopy showed benign, closely packed anastomosing cavernous vascular spaces with thick, smooth muscle and thin, fibrous

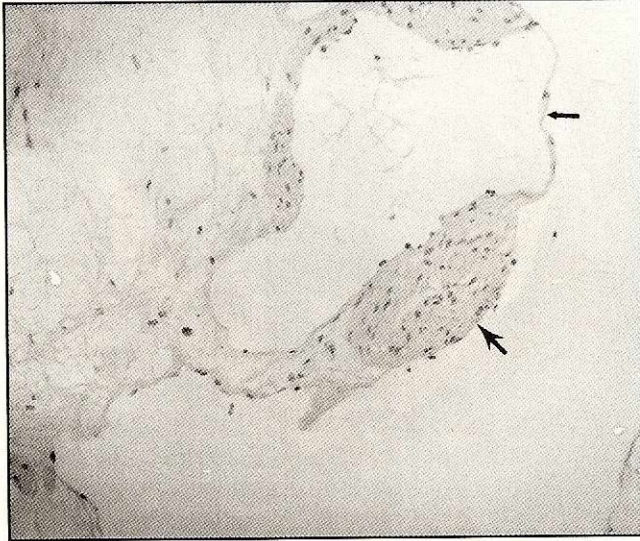


Figure 4: Photomicrograph of arteriovenous hemangioma or malformation, small bowel mesentery. Large arrow points to thick, smooth muscle. Small arrow points to thin, fibrous walls [hematoxylin and eosin, X200].



Figure 5: Photomicrograph of arteriovenous hemangioma or malformation, ileal submucosa [hematoxylin and eosin, X40].

TABLE 1

**Classification of Hemangiomas
According to Kaijser⁵**

- Type 1:** Multiple, submucosal venous varicosities (phleboectasia) consisting of dark bluish-red nodules that vary in size from a pinhead to a pea or serpiginous vessels that are scattered over a limited portion of the intestine.
- Type 2:** Cavernous hemangiomas in which the cavernous tissue usually extends to involve all layers of the bowel. Can occur as localized polypoid masses or multifocally throughout the bowel. Patients often present as a result of hemorrhage.
- Type 3:** Capillary hemangiomas (hemangioma simplex). Usually single, discrete, round, localized submucosal tumors up to 5cm in diameter.
- Type 4:** Diffuse angiomatosis (venous, capillary, or cavernous components histologically). Involves large segments of the body in a contiguous manner.

walls in the adipose tissue of the small bowel mesentery (Figure 4), which focally formed a polypoid cavernous submucosal component in the distal ileum (Figure 5).

Case 2: An eight-year-old boy who had iron deficiency anemia was found to have diffuse hemangiomas of the cerebral area, head, neck, extremities, glans penis, perianal area, and trunk. Upper endoscopy disclosed multiple cavernous hemangiomas of the stomach and duodenum. Colonoscopy showed multiple colonic hemangiomas. The iron deficiency resolved with supplemental iron. This case represents a Type 4 hemangioma (Table 1).

Case 3: A 44-year-old man was evaluated for colicky abdominal discomfort. It was thought that his symptoms were caused by episodic partial intestinal obstruction. CT scan confirmed the physical examination finding of an intraabdominal mass in the small intestine. The patient did not have anemia. Laparotomy revealed a white mass in the jejunum that clinically was thought to represent a lymphangioma. The mass was excised, and histologic examination showed a solitary polypoid cavernous hemangioma with multiple phleboliths. This case represents a Type 2 hemangioma. Postoperatively, the patient's symptoms resolved, and he had an uneventful recovery with no recurrent problems.

Case 4: A 72-year-old man presented with iron deficien-

cy anemia caused by occult gastrointestinal bleeding. Upper endoscopy results were normal. An angiographic study showed hypervascularity in the left colon consistent with angiodysplasia and a 5mm vascular lesion in the terminal ileum that appeared to be the site of gastrointestinal bleeding. Resection of this ileal segment was performed. Histologic examination revealed a Type 3 capillary hemangioma.

DISCUSSION

The diagnosis and treatment of gastrointestinal bleeding can be both frustrating and difficult. In 50-75% of cases of intestinal tract bleeding, the diagnosis is established with endoscopy, radiographic studies, and arteriography. However, some cases remain undiagnosed despite intense evaluation. Although vascular lesions may be suspected in the differential diagnosis of gastrointestinal tract bleeding of uncertain etiology, their demonstration often eludes the most sophisticated endoscopic and radiologic techniques.² A small subset of patients with gastrointestinal tract bleeding (1-27%)² may elude diagnosis even when exploratory laparotomy has been performed.⁶ With the advent of selective arteriography, nuclear medicine scans, and preoperative and intraoperative endoscopy, vascular abnormalities have been identified as the source of intestinal bleeding in an increasing number of patients.

Vascular lesions involving the gastrointestinal tract have been reported since 1839, but hemangioma of the small intestine was not described until 1860.⁷ In a comprehensive survey of the literature from 1839 to 1945, Gentry et al⁸ identified 189 cases of benign vascular malformations and vascular tumors of the gastrointestinal tract and added to that 94 Mayo Clinic cases. Hemangiomas can occur as either diffuse tumors or as circumscribed polypoid lesions. They tend to be multiple, and they occur more frequently in the jejunum and ileum (99%) than in the duodenum.⁹

Hemangiomas with diffuse involvement of the gastrointestinal tract are rare.¹⁰ Although the majority of benign tumors of the small intestine are asymptomatic, 66-80% of benign vascular tumors produce symptoms during the patient's lifetime. The cardinal symptom of an intestinal hemangioma is bleeding, which may be either massive or occult. The next most frequent symptom is obstruction secondary to intussusception. Patients with cavernous hemangioma of the small bowel may present with recurrent iron deficiency anemia,¹¹ abdominal pain,⁴ obstruction, and, rarely, intussusception with perforation and peritonitis.¹²

Based on the classification provided by Kaijser⁵ in which intestinal hemangiomas are divided into four dif-

ferent types (Table 1), a Type 2 hemangioma was identified in one of our patients. The lesions associated with hemangiomas outside the bowel often correlate with well-described syndromes such as the Osler-Rendu disease of multiple telangiectasis, the Parkes-Weber-Klippel syndrome of hemangiectatic hypertrophy of limbs, and the Bean syndrome of viscerocutaneous hemangiomas.^{13,14} These syndromes represent less than 10% of all hemangiomas.

The diagnosis of hemangioma usually is difficult to make and sometimes can be established only at exploratory laparotomy. Mucocutaneous hemangiomas may suggest an intestinal hemangioma in a small portion of cases. After a detailed history, physical examination, and routine complete blood cell count and stool guaiac tests, a plain abdominal radiograph should be taken. Hellström et al¹⁵ emphasized that phleboliths that occur outside the usual pelvic venous plexuses may be a clue to the presence of vascular malformation.

The definitive diagnosis can be made by surgical exploration with gross observation of the abnormalities, angiography, or endoscopy.⁶ Gross observation is useful only for lesions large enough to be visible at surgery. Our index case had gross abnormalities consistent with neoplasia that led to surgical exploration.

Visualization of the small intestine by detailed double-contrast technique provides a more satisfactory delineation of lesions of the small bowel mucosa.^{9,16} Superficial mucosal lesions usually are not evident with conventional small bowel barium studies, which are rarely helpful in establishing the diagnosis.

Selective angiography can be valuable in localizing bleeding sites.¹⁷ A bleeding rate of at least 0.5mL/min is necessary.¹⁸ Although technetium 99m-labeled red blood cell scans¹⁹ and technetium 99m sulfur colloid scans²⁰ can detect a slower rate of bleeding than angiography, active bleeding is still required to localize the bleeding source.¹⁷ Both techniques are valuable in the identification of superficial mucosal lesions that are difficult to visualize by other studies.

Intraoperative methods of localization of gastrointestinal hemangiomas include gross examination, palpation, transillumination, enterotomy with mucosal eversion, injection of methylene blue through an angiographic catheter, and endoscopy. A search for the bleeding site by direct examination of the intestine usually is unreliable unless a large hemangioma is present. Transillumination can reveal the lesion if the intestines are held before intense light and the contents are gently milked aside between the examiner's fingers. This test is difficult to perform and may be unproductive when the intestines are filled with blood.²¹ Intraoperative

fiberoptic endoscopy has been used to identify a real or potential source of bleeding in as many as 90% of patients. The endoscope can be introduced orally and passed downward to the ileocecal valve or it can be passed per rectum upward into the small intestine. Another technique is insertion of a flexible fiberoptic scope via a proximal small bowel enterotomy.

Treatment depends on the site and extent of the lesion. Where possible, resection of the involved area of intestine is curative. When hemangiomas give rise to massive involvement of the intestine, a conservative approach is best adopted, but resection may be necessary for recurrent, often severe, bleeding from a known area.¹⁴

CONCLUSION

Although hemangiomas of the small intestine are infrequently encountered, they should be included in the differential diagnosis of gastrointestinal tract bleeding of uncertain etiology. The diagnosis of hemangioma is difficult to make, often eluding the most sophisticated diagnostic techniques, and it sometimes is established only at exploratory laparotomy. The definitive diagnosis can be made by angiography, endoscopy, or a surgical procedure with gross observation of the abnormalities. Good results also have been achieved with fiberoptic endoscopy. Treatment depends on the site and extent of the lesion. Where possible, resection of the involved length of intestine is the procedure of choice.

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