lower gastrointestinal (LGI) bleeding is a frequent and sometimes frustrating clinical entity. Whereas the majority of episodes of LGI bleeding resolve with supportive measures, the source of the bleeding in many patients remains unclear. In general, angiodysplasia and diverticulosis are the most common causes of LGI bleeding. However, other, less common causes should be included in the differential diagnosis of LGI bleeding.

This report presents the case of a 41-year-old man with massive LGI bleeding caused by a Dieulafoy’s lesion of the jejunum. The definition, etiology, pathophysiology, clinical presentation, differential diagnosis, and management of Dieulafoy’s lesion are discussed.

**CASE PRESENTATION**

**Patient Presentation**

A 41-year-old man came to the emergency department because of severe rectal bleeding. His medical history was significant only for hypercholesteremia. He had no history of surgery, use of medications, allergy, or recent travel; his family history was similarly unremarkable. He smoked occasionally but did not drink alcohol or use recreational drugs.

**Evaluation in the Emergency Department**

Examination showed the patient to be hypotensive, with a blood pressure of 80/50 mm Hg. Temperature was 36.2°C (97.1°F), pulse was 78 bpm, and respiratory rate was 18 breaths/min. He appeared pale and weak, with a dry oral mucosa. His abdomen was soft, lax, nontender, and nondistended; bowel sounds were normal. Rectal examination revealed dry blood on the buttocks but no hemorrhoids or masses; however, there was maroon-colored blood in the rectum, and a guaiac test was strongly positive for occult blood. The rest of the examination was unremarkable.

While still in the emergency department, the patient was given saline, intravenously. He subsequently experienced 2 additional episodes of fresh rectal bleeding, losing a total of 800 mL of blood. He received 2 units of packed erythrocytes and was admitted to the intensive care unit.

**Postadmission Testing**

Results of laboratory testing on admission showed a hemoglobin level of 12.2 g/dL, a hematocrit of 35.5%, a prothrombin time of 11.9 seconds, and a partial thromboplastin time of 22.6 seconds. Values obtained on measurement of liver enzymes were within normal limits. Results of nasogastric tube lavage were negative for blood. No abnormalities were found on chest radiography or electrocardiography. A radioisotope scan of the abdomen and pelvis performed to detect bleeding showed accumulation of radioisotope over the lower left abdomen with possible bleeding sites involving the sigmoid colon and jejunum (Figure 1). A sigmoidoscopy performed at the bedside revealed maroon-colored blood over the lumen as the endoscope was advanced 60 cm. Subsequently, the patient underwent colonoscopy and endoscopy of the upper gastrointestinal tract in an unsuccessful attempt to detect the source of active bleeding.

Throughout testing, the patient remained hemodynamically unstable. A central line was placed, and he received 6 units of packed erythrocytes. A subsequent mesenteric angiogram revealed 2 apparent aneurysms on the jejunal branch of the superior mesenteric artery (Figure 2). Intra-arterial infusion of vasopressin at 0.2 U/min was started and resulted in bleeding cessation. Repeat angiography did not reveal any active bleeding. The patient became hemodynamically stable;
his hemoglobin level was now 8.2 g/dL. Because of the angiographic suggestion of aneurysms, a rheumatology consultation was requested to rule out a possible diagnosis of polyarteritis nodosa. The patient was then given corticosteroids intravenously. Results of subsequent measurement of erythrocyte sedimentation rate, serum rheumatoid factor, serum antinuclear antibodies, and serum antineutrophil cytoplasmic antibodies were within normal limits, and serologic tests to detect hepatitis were negative.

**Diagnosis and Treatment**

Three days after admission, the patient developed another massive attack of rectal bleeding and received another transfusion of packed erythrocytes. Because of the severity of the LGI bleeding and the aneurysmal dilatation of the jejenum, a tentative diagnosis of a Dieulafoy’s lesion of the jejenum was made. Exploratory laparotomy was performed, with resection of the bleeding jejenum and excision of a Meckel’s diverticulum that was incidentally discovered. The patient remained stable and had no further bleeding. During his hospital course, including surgery, the patient received a total of 16 units of packed erythrocytes. He was discharged home with a hemoglobin level of 11.8 g/dL.

Pathologic analysis of a surgical specimen of resected jejenum revealed cystic degeneration of the tunica intima and media and an incomplete tunica elastica. Additionally, there were 2 submucosal aneurysmal dilations, each measuring 6 to 7 mm in diameter. The dilatations were filled with clotted blood, eroding through the jejunal mucosa (Figure 3). These findings confirmed the diagnosis of a Dieulafoy’s lesion. There was no evidence of inflammatory components in the jejunal material, a finding that was also consistent with Dieulafoy’s lesion (Figure 4). Analysis of the incidental, nonbleeding Meckel’s diverticulum showed a lining of regular ileal mucosa. No evidence of hemorrhage or neoplasm was present. During 6 months of continuous follow-up visits, the patient had no episodes of active bleeding, and his hemoglobin level remained stable.

**DISCUSSION**

Despite its rarity, Dieulafoy’s lesion should always be considered in the differential diagnosis of LGI bleeding. Although usually located in the upper portion of the stomach, within 5 cm of the esophagogastric junction, Dieulafoy’s lesions recently have been identified in the duodenal bulb, the jejenum, the right colon, and the rectum.2 LGI bleeding is defined as bleeding distal to the ligament of Treitz. Acute LGI bleeding occurs less frequently than does acute upper gastrointestinal bleeding, although the actual incidence of either is unknown.3 Most cases of LGI bleeding involve the colon (85%), with diverticulosis and angiodysplasia considered to be the most common causes.4 Colonic LGI bleeding also
can result from neoplasms, inflammatory bowel disease, ischemia, colitis (secondary to radiation or infection), and hemorrhoids. Approximately 10% of cases of LGI bleeding are secondary to upper gastrointestinal bleeding, and only 3% to 5% of cases are secondary to bleeding from the small intestine. Vascular lesions, including Dieulafoy’s lesion, are the most common cause of small intestinal bleeding, accounting for 70% of cases. The causes of small intestinal bleeding that comprise its differential diagnosis are presented in Table 1.

**History of Dieulafoy’s Lesion**

Although Gallard’s description in 1884 preceded it, the initial description of a gastric submucosal aneurysm has been attributed to Dieulafoy (in 1898). Dieulafoy, a French surgeon, described 3 cases of fatal gastric hemorrhage that he attributed to a bleeding artery associated with a solitary gastric erosion. He called the condition *exulceratio simplex.*

Enteric Dieulafoy’s lesions are very rare. In 1978, Matuchansky and colleagues reported 2 cases of jejunal Dieulafoy’s lesions diagnosed by angiography. Vetto and colleagues described 2 other cases in the jejunum in 1989 and recognized the similarity between Dieulafoy’s lesions of the stomach and of the jejunum. Additional cases of jejunal Dieulafoy’s lesion were reported in 1998 and 1999.

**Pathology of Dieulafoy’s Lesion**

Grossly, a Dieulafoy lesion consists of a protuberant, tortuous, serpiginous, and abnormally wide artery located in the submucosa of the gastrointestinal tract that approximates a submucosal tumor. Microscopic examination usually shows a thick-walled vessel, without a surrounding inflammatory reaction. Pathologically, this caliber-persistent artery has no associated aneurysm, despite that fact that it has been called a cirrhotic aneurysm and can be ulcerated. There is no histologic distinction between gastric and small intestinal Dieulafoy’s lesions.

**Clinical Presentation of Dieulafoy’s Lesion**

The characteristic clinical presentation of a Dieulafoy’s lesion involves recurrent, painless, and massive gastrointestinal bleeding in a patient who is hypotensive. It can occur in persons of any age, is characterized by a male predominance, and does not have a familial tendency.

The clinical presentation of patients with Dieulafoy’s lesions of the duodenum and the proximal portions of the jejunum is similar to that of patients with gastric Dieulafoy’s lesions. Typical signs include melena and hematemesis. In contrast, patients with lesions in the middle or distal jejunum, as well as in the colon, generally have massive, bloody stools.

**Diagnosis of Dieulafoy’s Lesion**

The diagnostic tools appropriate to identify a Dieulafoy’s lesion depend on the location of the lesion. Dieulafoy’s lesions of the stomach or duodenum can be detected by endoscopy of the upper gastrointestinal tract. However, performing endoscopy often is difficult because of the massive bleeding associated with these lesions (and with Dieulafoy’s lesions of the colon and rectum). Jejunal Dieulafoy’s lesions cannot be detected by conventional endoscopy. If enteroscopy is available (ie, endoscopy of the small intestine), Dieulafoy’s lesions in the proximal part of jejunum might be detectable.
### Table 1. Causes of Small Intestinal Bleeding

<table>
<thead>
<tr>
<th>Cause</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Vascular lesions</strong></td>
<td></td>
</tr>
<tr>
<td>Angiodysplasia (also known as vascular ectasia)</td>
<td>Dilated vascular complexes that include arterioles, capillaries, and venules but no dysplastic tissue†</td>
</tr>
<tr>
<td>Telangiectasia</td>
<td>Dilated vascular complexes that differ from those of angiodysplasia in their diffuse nature, tendency to recur, and association with cutaneous or mucous membrane lesions (a hereditary form of telangiectasia is most often responsible for intestinal telangiectasia‡)</td>
</tr>
<tr>
<td>Venous ectasia (also known as phlebectasia)</td>
<td>Nonneoplastic venous varicosities (with abnormal endothelial lining) that appear as multiple bluish-red nodules; very uncommon cause of bleeding§</td>
</tr>
<tr>
<td>Hemangioma</td>
<td>Rarely malignant neoplastic growth composed of proliferating blood vessels pathologically divided into capillary, cavernous, and mixed forms§</td>
</tr>
<tr>
<td>Arteriovenous malformation</td>
<td>Congenital lesion found anywhere in the gastrointestinal tract (mostly in children)¶ that is characterized by thick-walled arteries and veins with no intervening capillaries, forming a true arteriovenous fistula</td>
</tr>
<tr>
<td>Dieulafoy’s lesion (also known as caliber-persistent artery)</td>
<td>Arterial lesion that fails to narrow or decrease in diameter as it penetrates the submucosa†</td>
</tr>
<tr>
<td><strong>Tumors of the small intestine</strong></td>
<td></td>
</tr>
<tr>
<td>Leiomomas, leiomyosarcoma#</td>
<td>Growth that collectively comprise 5% of all gastrointestinal tumors and 5% to 10% of all cases of gastrointestinal bleeding</td>
</tr>
<tr>
<td>Carcinoid tumors, adenocarcinoma, lymphoma, adenomatous polyp, metastases**</td>
<td></td>
</tr>
<tr>
<td><strong>Less common causes</strong></td>
<td></td>
</tr>
<tr>
<td>Ulcerations of the small intestine</td>
<td>Abnormalities associated with Crohn’s disease, Meckel’s diverticulum, and Zollinger-Ellison syndrome</td>
</tr>
<tr>
<td>Large-artery vasculitis (eg, aortitis)</td>
<td>Inflammation that can lead to arterial occlusion with subsequent gangrene and perforation of the small intestine</td>
</tr>
<tr>
<td>Medium-sized artery vasculitis</td>
<td>Inflammation that can lead to the development of aneurysms (as in polyarteritis nodosa, rheumatoid arthritis, and systemic lupus erythematosus)</td>
</tr>
<tr>
<td>Small-sized artery vasculitis</td>
<td>Inflammation that can cause pain, fever, and occult bleeding (as in Henoch-Schönlein purpura††)</td>
</tr>
<tr>
<td>Radiation enteritis</td>
<td>Inflammation that can cause vasculitis and subsequent bleeding; radiation initially affects the intestinal mucosa directly, causing ulceration and bleeding, with late injury occurring 6 to 24 months after radiation treatment because of progressive occlusive vasculitis‡‡</td>
</tr>
</tbody>
</table>

*Arranged in order of decreasing frequency.
#High bleeding tendency.
Sonde enteroscopy can examine the entire small intestine. An alternative diagnostic tool is angiography, which is helpful in the localization of the bleeding site. In patients with recurrent massive gastrointestinal bleeding, repeat angiography might be necessary if the first examination fails to find the bleeding site because of inactive bleeding. If a diagnosis remains uncertain after angiography, portable intraoperative technetium Tc 99m scintigraphy should be considered in patients with problematic gastrointestinal bleeding.

Management of Dieulafoy’s Lesion

Enteroscopy can be therapeutic as well as diagnostic in cases of lesions of the small intestine. However, the optimal treatment of jejunal Dieulafoy’s lesions is surgical wedge resection. In fact, surgery might be the only way to treat distal jejunal Dieulafoy’s lesions. Once lesions are surgically resected, bleeding should not recur; the occurrence of more than 1 of these congenital lesions has not been described in the literature.

The case patient was initially treated with intraarterial vasopressin therapy, but bleeding recurred, most likely because of the relative mobility of the jejunum and the thinness of its wall, both of which made it difficult to obtain the desired compression of the vessel by injection sclerotherapy. However, sclerotherapy or sclerocautery of Dieulafoy’s lesions of the stomach or colon can be successful.

The mortality rate associated with Dieulafoy’s lesions ranges from 23% to 35%. Death most often occurs as a result of the massive bleeding.

CONCLUSION

The majority of cases of LGI bleeding pose no diagnostic or therapeutic challenge; either bleeding resolves spontaneously, or a bleeding site is easily identified. The remaining cases of LGI hemorrhaging represent a minority of cases in which localizing procedures are equivocal, despite ongoing bleeding. Dieulafoy’s lesions of the jejunum fall into the latter category. These lesions represent a rare vascular anomaly that can be potentially life-threatening. Because of the relatively high mortality rate of these lesions, they should be part of the differential diagnosis of any occult lower gastrointestinal hemorrhage. Many of the cases in the literature were diagnosed by angiography, and nearly all required surgical excision, as was true with the case patient. Surgery is usually curative, with no further bleeding occurring. This report is the ninth case in the English literature of jejunal Dieulafoy’s disease.

ACKNOWLEDGMENTS

The authors thank Dr. Jacklyn Mansour for her useful editing suggestions and Dr. Michael Getachew for his significant radiologic expertise.

REFERENCES