Periampullary Dieulafoy's Lesion

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Dieulafoy's lesion is responsible for 1% to 5.8% of acute nonvariceal upper gastrointestinal (GI) bleeding cases. The lesion is most commonly found in the stomach, although it may be encountered in any segment of the GI tract.[1–3] Dieulafoy's lesion was first reported in 1884 by Gallard[4] but was named after a French surgeon who described the lesion more accurately in 1897.[5] The lesion is an unusual clinical finding and is associated with high morbidity and mortality if diagnosis and treatment are delayed. In the setting of an acute upper GI hemorrhage, a high degree of suspicion combined with detailed endoscopic investigation is necessary to identify the lesion, thereby allowing early endoscopic treatment and avoiding surgical intervention. This case report describes an acute GI hemorrhage from a Dieulafoy's lesion found in an uncommon location, the periampullary region.

CASE PRESENTATION

Initial Presentation and History

A 25-year-old man was admitted to the hospital with a 2-day history of lightheadedness and fatigue. Nausea and diaphoresis accompanied the presyncopal symptoms, but he denied syncope, shortness of breath, and chest pains. One day prior to admission, he began to notice melena. He had no complaints of hematemesis, hematochezia, abdominal pain, palpitations, or use of nonsteroidal anti-inflammatory drugs.

The patient's history included hospitalization for a clean-based duodenal bulbar ulcer 2 months prior to the current presentation; he received a total of 4 U of packed erythrocytes over the course of 3 days. During this hospitalization, a rapid urease test revealed that the patient was infected with *Helicobacter pylori,* and he was treated with a 14-day course of appropriate eradication therapy. He had undergone a successful radiofrequency ablation procedure for Wolff-Parkinson-White syndrome 1 year prior to presentation. There was no history of tobacco or alcohol use.

Physical Examination and Laboratory Evaluation

On physical examination, the patient appeared to be in no acute distress. He was afebrile, and his vital signs were stable: heart rate, 91 bpm; blood pressure, 144/77 mm Hg; 99% oxygen saturation on room air via pulse oximetry; and respiratory rate, 16 breaths/min. Pale conjunctiva was noted bilaterally, which is suggestive of anemia. The results of cardiac, pulmonary, and extremity examinations were unremarkable. Abdominal examination, however, was significant for mild epigastric tenderness to palpation. Digital rectal examination confirmed melena. Initial laboratory evaluation revealed hemoglobin level and hematocrit of 6.8 g/dL (normal, 13.7–17.1 g/dL) and 22.8% (normal, 40.9%–51.1%), respectively. Hemoglobin level had been 10.5 g/dL and hematocrit 32.5% 1 month prior to presentation. Electrocardiogram, coagulation studies, liver function tests, and chest and abdominal radiographic studies were normal. Nasogastric lavage did not return coffee ground or bloody aspirate.

Diagnostic Evaluation

Resuscitative measures, including aggressive intravenous hydration and administration of 4 U of packed erythrocytes, were promptly initiated upon admission to the intensive care unit. An upper endoscopy was performed on the first day of hospitalization. On upper endoscopy, fresh blood was noted in the second portion of the duodenum, but the bleeding site could not be localized with the standard forward-viewing endoscope. A side-viewing duodenoscope was then utilized to locate the bleeding site. Brisk active bleeding was seen originating from the periampullary region (**Figure 1**). Irrigation revealed the exact bleeding point to be a small protuberant defect with no endoscopic evidence of associated mucosal ulceration or erosion (**Figure 2**). Based on the endoscopic findings in the setting of an acute GI bleed, the patient was diagnosed with Dieulafoy's lesion.

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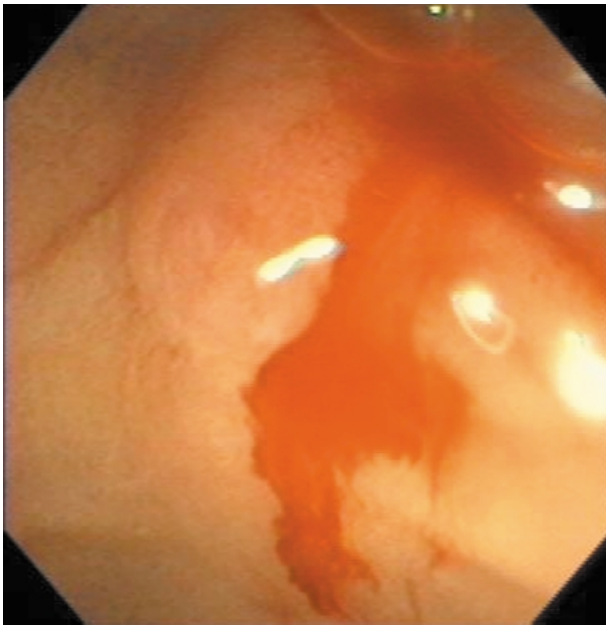

Figure 1. Endoscopy showing brisk active bleeding originating from the periampullary region.

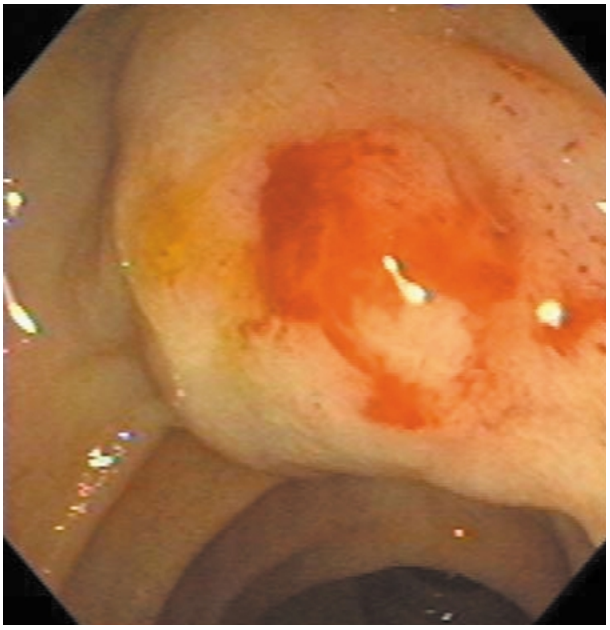

Figure 3. Endoscopy showing hemostasis after epinephrine injection therapy directed at the margins of the bleeding point.

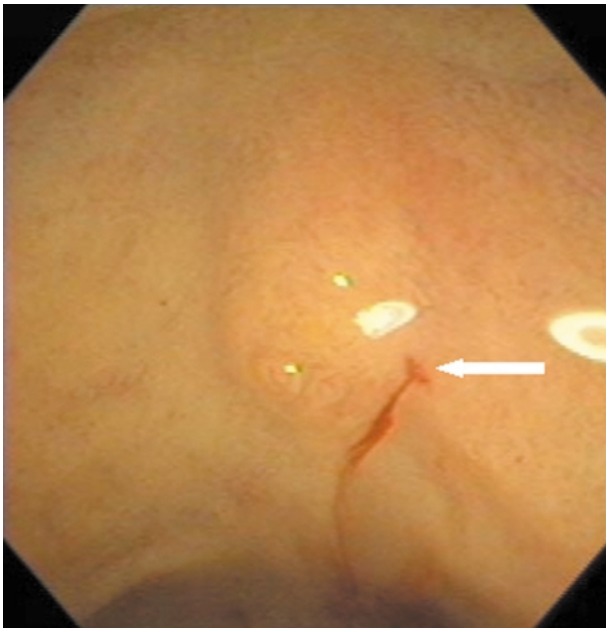

Figure 2. Endoscopy revealing that the exact bleeding point was a small protuberant defect (*arrow*) with no evidence of associated mucosal ulceration or erosion.

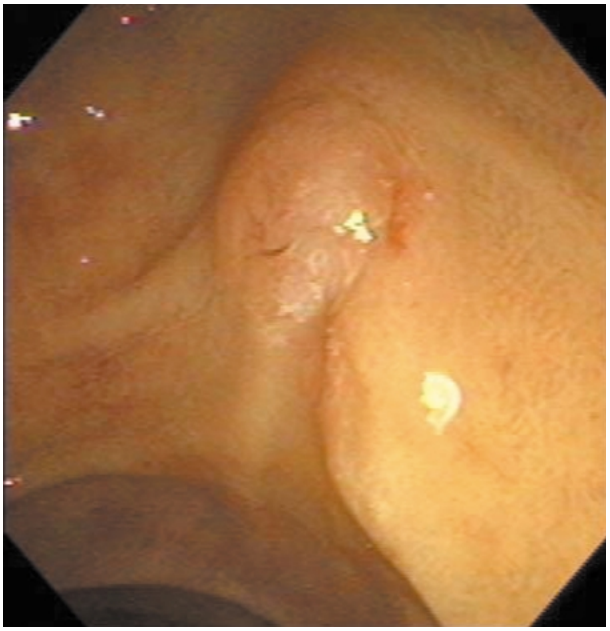

Figure 4. Repeat endoscopy confirming the absence of bleeding, ulceration, or erosion in the periampullary region.

Treatment and Outcome

Combination treatment of hemoclip application and epinephrine injection was initially planned. Attempts to hemoclip the lesion were unsuccessful; the device could not be deployed at the tip of the side-viewing duodenoscope. The bend and deflection created by the elevator mechanism of the duodenoscope likely contributed to this mechanical difficulty. However, epinephrine injection therapy (1:10,000 dilution) directed at the margins of the bleeding point was performed successfully, with

the patient receiving a total of 5 mL of epinephrine, and complete hemostasis was achieved (**Figure 3**). At this point the patient became increasingly intolerant of the procedure, and having achieved hemostasis, the procedure was considered successful and completed. No other lesions were found. Repeat endoscopic evaluation 3 days later showed no evidence of bleeding and confirmed the absence of periampullary ulceration or erosion (**Figure 4**).

A repeat rapid urease test to detect *H. pylori* was negative. The patient's hospital course was otherwise unremarkable, and he was discharged on hospital day 4. At 6 months' follow-up, he was doing well and had not experienced recurrent bleeding. His hemoglobin level and hematocrit had normalized to 14.8 g/dL and 45.1%, respectively.

DISCUSSION

Pathophysiology

Dieulafoy's lesion may be an unfamiliar etiology of acute GI hemorrhage to many nongastroenterologists. However, it is a high-risk lesion and has the potential to cause profuse and life-threatening hemorrhage. Dieulafoy's lesion is a large (1–3 mm), "caliber-persistent" submucosal tortuous artery that protrudes through a minute mucosal defect, typically 2 mm to 5 mm, without associated inflammation or ulceration.[2,6,7] The submucosal artery fails to diminish to the minute size of the typical mucosal microvasculature.[8,9] This anomalous vessel is thought to be abnormally fixed to the muscularis mucosa rather than being mobile in the submucosa, thereby causing abnormal stresses and pressure erosion of the overlying mucosa during peristalsis.[8,10] Histologic evidence supports the theory that the final pathologic process is a progressive weakening of the vessel wall, which in combination with the overlying mucosal defect leads to hemorrhage.[8,11]

Epidemiology and Most Common Defect Sites

Dieulafoy's lesion is found twice as often in males as in females, with a median age at presentation of 54 years.[12] Children of any age can also be affected, but it is considered extremely rare in this population.[2,13,14] Dieulafoy's lesion has been described in all segments of the GI tract, but the stomach remains the most common site; in a review of cases published since 1993, 74% of Dieulafoy's lesions were found in the stomach.[2] The majority of stomach lesions are found within 6 cm of the gastroesophageal junction along the lesser curvature.[2,15] The predilection for the lesser curve is likely related to the local blood supply: the lesser curve is an area of the stomach that is not perfused by a submucosal plexus but instead derives its blood supply directly from tributaries of the right and left gastric artery.[16] The duodenum is the second most common site for Dieulafoy's lesion, which may be due to its unique blood supply that often consists of end arteries.[17] Approximately 53% of the Dieulafoy's lesions encountered in the duodenum have been found in the bulb.[2] Periampullary location is an extremely uncommon site for reported Dieulafoy's lesions. Schmulewitz and Baillie[18] described their experience with 40 cases of Dieulafoy's lesions found on upper endoscopy over a 6-year period, but no lesions were identified next to the ampulla and only one was located next to the minor papilla.

Diagnosis

Presenting complaints of patients with Dieulafoy's lesion include hematemesis, melena, large volume hematochezia, presyncope, and syncope. Resuscitative measures and monitoring should follow once GI bleeding is suspected. The mean number of units of blood transfused ranges from 3 to 8 U, and it is estimated that life-threatening acute GI hemorrhage due to Dieulafoy's lesion occurs in approximately 10% of cases.[2] The following endoscopic criteria are generally accepted for the diagnosis of Dieulafoy's lesion: (1) active arterial spurting or micropulsatile streaming of blood from a minute (< 3 mm) mucosal defect or through normal surrounding mucosa; (2) visualization of a protruding vessel, with or without active bleeding, within a minute mucosal defect or through normal surrounding mucosa; or (3) fresh, densely adherent clot with a narrow point of attachment to a minute mucosal defect or to normal-appearing mucosa.[6] The lesion can be difficult to identify in the setting of a massive GI hemorrhage, and the true incidence is difficult to determine. Dieulafoy's lesions are likely underdiagnosed, especially in the setting of blood clots in the stomach that hinder optimal endoscopy or in the absence of active bleeding.[19] A high degree of clinical suspicion as well as an experienced endoscopist is necessary for timely diagnosis and treatment of the lesion. Diagnosis at initial endoscopy ranges from 49% to 92%, and it may require 3 or more endoscopic examinations in as much as 6% of cases to identify the Dieulafoy's lesion as the bleeding source.[1–3,7] As with other etiologies of acute GI hemorrhage, surgical consultation is necessary in difficult cases. Angiography can also be a diagnostic tool as well as a therapeutic tool in endoscopically difficult cases.[20]

Treatment

As recently as 1986, surgery was being advocated as the treatment of choice in Dieulafoy's lesion cases;

surgery involved ligating the offending vessel or performing a subtotal and even a total gastrectomy.[2,21] Since then, endoscopic therapy has clearly become the intervention of choice for endoscopically accessible Dieulafoy's lesion, achieving up to a 95% success rate.[1,3,19] Today, the mortality rate associated with Dieulafoy's lesion, which in most cases is not directly related to continued GI hemorrhage, is less than 20% compared with rates as high as 80% during periods prior to the prominence of endoscopic management.[2,10] Surgical intervention is now the last resort, reserved for unknown source of GI bleeding or endoscopically uncontrollable bleeding from Dieulafoy's lesion.

Endoscopic treatment of Dieulafoy's lesion includes a variety of modalities, including injection therapy, thermal probe coagulation, band ligation, hemoclipping, endoloop deployment, laser treatment, and a combination of these therapies.[1-3,6,7,15,18,19,22–26] Combination therapy is more effective than single modality in preventing recurrent bleeding, and mechanical therapies, including hemoclip application, have been demonstrated to be at least as effective in achieving hemostasis and preventing recurrent bleeding.[2] It is believed that epinephrine injection and thermal probe coagulation are the 2 most commonly used endoscopic therapies, mainly due to their wide availability and low cost.[18] Furthermore, factors such as the endoscopist's expertise and familiarity with available therapeutic modalities, the location of the lesion, the urgency of therapy, and the experience of the support staff are all considered when deciding on a specific therapy. Recurrent bleeding can be seen in up to 28% of treated Dieulafoy's lesion cases, and repeated endoscopic treatment is recommended because successful hemostasis can be achieved in the majority of cases.[2,20] Prognosis is excellent once the Dieulafoy's lesion is successfully treated; Kasapidis and colleagues[7] reported no recurrence during a mean follow-up of 32 months.

As in the case reported here, the use of a side-viewing duodenoscope can assist in achieving optimal endoscopic visualization and in delivery of therapy. A side-viewing duodenoscope is usually used in performing endoscopic retrograde cholangiopancreatography. This instrument differs from the standard forward-viewing endoscope used in performing esophagogastroduodenoscopy in that it provides imaging along the lateral aspect of the tip rather than from the end of the endoscope, allowing the endoscopist to obtain images of the medial wall of the duodenum.[27] An instrument channel, along with an elevator mechanism at the tip, allows usage of accessory catheters and devices. The value of side-viewing endoscopy has been well established in the setting of GI hemorrhage from lesions such as angiodysplasia of the minor papilla, duodenal diverticula, gastric and duodenal bulbar ulcers, gastric and duodenal bulbar Dieulafoy's lesions, and duodenal aortoenteric fistula.[28–32] The major papilla and the periampullary region are particularly difficult areas to adequately visualize with a forward-viewing endoscope alone; thus, side-viewing duodenoscopy has become the standard for evaluating lesions in this location.[33,34]

CONCLUSION

GI bleeding from a Dieulafoy's lesion is rare but can be potentially life-threatening. Early recognition of the lesion and treatment are imperative in the setting of acute GI bleeding. This case illustrates the importance of including Dieulafoy's lesion in the differential diagnosis of acute GI bleeding and performing a meticulous endoscopic evaluation in establishing the diagnosis. Endoscopic treatment modalities have replaced surgical intervention as first-line therapy for Dieulafoy's lesion. The periampullary region is an uncommon location for a Dieulafoy's lesion. A side-viewing duodenoscope optimizes visualization of the periampullary region and allows delivery of endoscopic therapy. **HP**

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