Single Case

Massive Gastrointestinal Bleeding from a Jejunal Dieulafoy Lesion: An Extraordinary Presentation

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Keywords

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Abstract

We present an atypical and rare case of a previously healthy 27-year-old male who presented with acute onset of abdominal pain, bloody diarrhea, and syncope. At the Emergency Department, vital signs were stable with no signs of shock. Physical examination revealed diffuse tenderness of the abdomen and cherry red blood was noted upon rectal examination. Blood tests showed marked leukocytosis without anemia. Sigmoidoscopy was performed which revealed hematochezia with no obvious site of bleeding. The patient was admitted to the hospital with a working diagnosis of dysentery and received supportive care. During the following days, blood tests revealed an ongoing decline of hemoglobin levels which necessitated a new workup of gastrointestinal bleeding. Investigation modalities including upper and lower endoscopies as well as angiography failed to demonstrate a bleeding site. Scintigraphy, which was performed next, demonstrated an increased radiotracer activity in the right abdomen consistent with small bowel bleeding. Following these results, the patient underwent urgent laparotomy and surgical resection was performed. The histopathological findings were consistent with a Dieulafoy lesion. This case illustrates the importance of the complementary role of various modalities in locating the bleeding site along the gastrointestinal tract.

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Introduction

Dieulafoy lesion (DL), which was first described by Gallard in 1884 and named after Dieulafoy in 1898, is one of the rare etiologies (1.3%) of massive gastrointestinal (GI) bleeding associated with a high mortality rate [1]. The predominant characteristic of DL is a persistently large pulsatile caliber artery, 1–3 mm in size, which runs a tortuous path through the submucosa, with histologically normal surrounding mucosa and increased vascular wall fibrosis [2, 3]. The diagnosis and localization of DL during massive bleeding are both important and difficult. The anatomical distribution of DL includes the stomach (71%), duodenum (15%), esophagus (8%), rectum (2%), colon (2%), and jejunum (1%) [4, 5].

The most frequently used diagnostic modalities are endoscopy, angiography, and scintigraphy. Surgery is the treatment of choice in only 5% of patients and performed when endoscopic and angiographic attempts have failed [5].

We present a rare case of massive lower GI bleeding in a 27-year-old man. After diagnostic failures of endoscopic and angiographic interventions, localization of the bleeding site was achieved using scintigraphy followed by surgical resection. Final histopathological findings revealed a DL of the jejunum.

Case Description

A previously healthy 27-year-old male presented with acute onset of abdominal pain, bloody diarrhea, and syncope. His symptoms had begun 3 h prior to the Emergency Department (ED) visit, after eating at a local restaurant. On examination at the ED, the patient appeared well, vital signs included a temperature of 36.7°C, blood pressure of 118/85 mm Hg, pulse 84 beats per minute, and oxygen saturation was 100% while he was breathing ambient air. Abdominal examination revealed diffuse tenderness of the abdomen and cherry red blood was noted upon rectal examination with no signs of organomegaly, rebound tenderness, or ascites. The remainder of the physical examination including neurological examination was normal. Blood tests revealed a total white blood cell count of 17,000 with 83% neutrophils, hemoglobin levels of 13 g/dL, and a normal platelet count. Other blood tests including levels of glucose, calcium, potassium, sodium, magnesium, phosphorus alanine aminotransferase, aspartate aminotransferase, alkaline phosphatase, total bilirubin, direct bilirubin, vitamin B₁₂, C-reactive protein, and renal-function tests were within the normal range limits.

Sigmoidoscopy was performed and revealed hematochezia with no obvious site of bleeding. The patient was admitted to the internal medicine ward with a working diagnosis of dysentery. The following day, the patient was treated with fluid resuscitation and antibiotics. Hemoglobin level dropped to 11.1 g/dL from a baseline of 13 g/dL, which was attributed to a hemodilution effect. On day 3 of hospitalization, blood tests revealed an ongoing decline of hemoglobin levels, which necessitated a workup of a massive GI bleed. The next morning, the patient underwent urgent upper and lower endoscopies which failed to demonstrate a source of bleeding. Angiography was the next modality to be performed and again there was no evidence of a bleeding site.

In light of a working diagnosis of a life-threatening bleeding, a decision was made to proceed to Tc^{99m} red blood cell (RBC) scintigraphy, which demonstrated an early focus of increased radiotracer activity in the right lower quadrant which appeared to accumulate and ascend the right abdomen in a pattern consistent with small bowel bleeding (online suppl. Video 1, 2; for all online suppl. material, see www.karger.com/doi/10.1159/000495207).

There was also milder uptake in the left abdomen that also accumulated throughout the study. Following these results, the patient underwent emergent laparotomy which demonstrated the bleeding site in the jejunum (Fig. 1). A surgical resection was performed, and the specimen was sent for further pathology investigation. The patient's medical condition improved gradually the following days and he was discharged home on day 5 postoperatively. The histopathological findings were consistent with a DL (Fig. 2).

On follow-up 3 months after surgery, the patient was doing well, with no recurrent bleeding or related symptoms.

Discussion

DL can be associated with massive, life-threatening hemorrhage and accounts for 1–2% of cases of major GI bleedings [3–5]. Due to the possibility of massive recurrent bleeding, the mortality rate is high if not managed properly. The mechanism of DL's rupture is unclear. One theory suggests that the path of a DL runs in close proximity to the overlying mucosa; thus, DL's pulsations may disrupt said epithelium, leading to localized ischemia, erosion of the mucosa, and rupture of the artery [4]. DL of the jejunum is extraordinarily rare, with only a small number of cases reported to date [6–8]. This lesion is often seen in elderly, male patients with multiple comorbidities who are treated with non-steroidal anti-inflammatory drugs (NSAIDs), aspirin, or warfarin [5, 6]. The mean age at presentation is over 50 years [6, 7]. DL is more prevalent in men than women. Other etiologies of DL include alcoholism, stress, cardiac or pulmonary failure, and fecalomas. According to previously published studies, over 80% of patients with DL had associated diseases [8].

In our case, the patient did not have any of these mentioned characteristics, which rendered DL quite low in the differential diagnosis at ED presentation. The leading presenting symptom is recurrent massive bleeding, including hematemesis, melena, and hematochezia and resulting hemorrhagic shock [7, 9].

The small size of the lesion and extensive blood and/or clot formation in the lumen make the diagnosis challenging. Diagnosis is even more complex as the bleeding stops. Thus, a repeated endoscopy, which is the most sensitive and cost-effective method, may be required [8]. If upper and lower endoscopies fail to locate the site of bleeding, or the bleeding is beyond the reach of therapeutic endoscopy, angiography is usually performed to localize the bleeding. It might need to be repeated as DL bleeds intermittently and sometimes can be missed as in our case [6, 9].

The relatively stable persistence of Tc^{99m} RBCs in the blood pool enables intermittent (and venous) bleeding to be detected, making RBC scintigraphy unique among diagnostic imaging methods in that it allows for monitoring of patients with an intermittent pattern of bleeding [10].

Tc^{99m} RBCs accurately localize the site of bleeding in 88–97% of patients, with positive findings resulting in a 5-fold greater likelihood that the patient will require surgery [11]. RBC scintigraphy is often performed with the aim of determining a sufficient rate of bleeding to facilitate successful angiography and possible angiographic intervention [12]. A positive RBC scintigraphy study increases the likelihood of a positive angiogram from 22–53%. Among the advantages of RBC scintigraphy versus angiography are the 10-fold greater sensitivity for detection of slow bleeding rates or chronic bleeding lesions and the ability to examine the entire lower GI tract simultaneously and continuously over an extended period of time (60–90 min). Many angiographers prefer to have RBC scintigraphy prior to the contrast study to ensure that

bleeding is active, i.e., the contrast study is more likely to identify the bleeding site if scintigraphy is positive. RBC scintigraphy plays an important role in the diagnosis of small intestinal bleeding, when conventional endoscopy (EGD, push enteroscopy, and colonoscopy) has limited value, and costly innovative methods, such as capsule endoscopy and double-balloon enteroscopy, are not readily available [13].

In our case, evaluation by esophagogastroduodenoscopy, colonoscopy as well as angiography all failed to show any abnormal bleeding site; thus, a decision was made to proceed to RBC scintigraphy prior to proceeding to the surgical treatment which is considered the last resort after failure of the aforementioned techniques [14]. Minimally invasive surgery is an option for DL, especially in the jejunum, but it needs preoperative localization. In our case, it was not possible, as the patient deteriorated hemodynamically. Despite the advantages of low rebleeding rates and definitive treatment option, surgery should be considered only after failure of therapeutic endoscopy and angiographic interventions and should be guided by peri- or preoperative localizations.

Therapeutic options for DL include endoscopic, angioembolization, and surgical resection. Deep enteroscopy is the preferred technique to treat hemodynamically unstable patients suspected of small-bowel DL [15, 16]. The low incidence of DL makes it difficult to conduct large randomized control studies; thus, there are no clear guidelines on the best endoscopic techniques to treat DL. Epinephrine + heater probe (HP), argon plasma coagulation (APC), hemoclips (HCP), and epinephrine + HCP have all been used to treat DL in the small bowel [17, 18]. A retrospective study by Dulic-Lakovic et al. [15] demonstrated 100% initial hemostasis, 20% rebleeding, 2 resections, and no mortality [18]. Even though this study was small, HCP, Epinephrine + thermal methods, or Epinephrine + HCP had similarly effective hemostatic rates and safety profiles [19].

In summary, DL is an important cause of life-threatening bleeding. It is part of the differential diagnosis of upper or obscure GI bleeding, especially in patients who present with acute, massive, and intermittent upper GI bleeding [20]. Diagnosis and treatment options could pose a challenge – especially in young patients without known comorbidities, in whom the clinical presentation could mimic other more common medical conditions, thus necessitating the use of complementary diagnostic tools to accurately diagnose and localize the lesion.

Statement of Ethics

On behalf of all authors, I state hereby our commitment to integrity and honesty while writing the manuscript.

Disclosure Statement

The authors have no conflicts of interest to declare.

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Fig. 1. Jejunal DL during urgent laparotomy: An intraoperative specimen from the jejunum demonstrating the source of bleeding.

Fig. 2. Histopathological findings. Low magnification (objective lens $\times 2$) of hematoxylin and eosin-stained slides shows the presence of blood clot within the large tortuous, caliber-persistent artery through the overlying normal-appearing surrounding mucosa.