A Challenging Case of Jejunal Dieulafoy’s Lesion: A Rare Cause of Refractory Lower-Gastrointestinal Bleeding

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Patient: Male, 41-year-old
Final Diagnosis: Jejunal Dieulafoy’s lesion
Symptoms: Hematochezia • loss of consciousness • melena • vomiting
Medication: —
Clinical Procedure: Exploratory laparotomy w/partial jejunal resection • lower GI endoscopy • push enteroscopy w/hemoclip application • upper GI endoscopy
Specialty: Gastroenterology and Hepatology • Histology and Embryology • Surgery

Objective: Rare disease

Background: Dieulafoy’s lesion is a rare cause of severe gastrointestinal (GI) bleeding, accounting for approximately 1-2% of all cases of GI hemorrhage. Nevertheless, it can be life-threatening without prompt intervention. Dieulafoy’s lesion of jejunal origin can be particularly challenging to identify due to the inability of conventional endoscopic techniques to visualize the jejunum. This case report emphasizes the difficulties in diagnosing and managing jejunal Dieulafoy’s lesions and highlights the methods by which to approach refractory bleeding.

Case Report: This is a case of a 41-year-old man with a history of uncontrolled hypertension who presented with an episode of syncope and melena associated with low hemoglobin levels requiring multiple packed red blood cell transfusions. This warranted searching for a source of bleeding within the gastrointestinal tract via 2 upper-GI endoscopies, a colonoscopy, and an abdominal computed tomography angiogram, all of which failed to localize the site of bleeding. A push enteroscopy was required to identify the lesion in the jejunum, but the bleeding was not controlled despite the application of hemoclips and epinephrine. Consequently, laparotomy and resection of the jejunal segment containing the Dieulafoy’s lesion was performed and the diagnosis was established histopathologically. The patient recovered well and was discharged 4 days after the procedure.

Conclusions: Suspicion of a jejunal Dieulafoy’s lesion should be raised if both upper- and lower-GI endoscopies yield unremarkable findings. Ideally, a push enteroscopy should be utilized diagnostically and to conservatively manage the bleeding. However, laparotomy should be considered in refractory lesions or in the presence of hemodynamic instability.

Keywords: Endoscopy • Gastrointestinal Hemorrhage • Jejunal Diseases • Laparotomy • Melena • Syncope

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Background

Bleeding from the gastrointestinal tract (GI) can often present as a life-threatening condition. One of the rare causes of major GI bleeding is Dieulafoy’s lesion (DL). It is defined as a submucosal artery characterized by an abnormally large diameter with a tortuous course along the lining of the GI tract. It can protrude through a defect in the overlying mucosa secondary to erosion or trauma and into the lumen, resulting in gastrointestinal bleeding [1,2]. First identified in 1898 by French surgeon Paul Georges Dieulafoy, it has since been found to be involved in 1-2% of all GI hemorrhage cases [2]. DL can occur anywhere throughout the GI tract, with the gastric region being the most common site, found in 72% of cases, followed by the duodenum in 15%, the esophagus in 8%, the colon in 2%, the rectum in 2%, and most rarely, in the jejunum-ileum in only 1% of cases [1]. This case report highlights an elusive jejunal DL identified enteroscopically in a 41-year-old man who first presented with syncope associated with melena and signs of anemia. He subsequently underwent emergency laparotomy with partial resection of the jejunum and primary anastomosis, which was followed by histopathological confirmation of the diagnosis.

Case Report

Our patient was a 41-year-old laborer with a background of uncontrolled hypertension and a 25-pack-year history of smoking. He initially presented to the hospital after an episode of loss of consciousness (LOC) that occurred while he was walking to the washroom. This was associated with 2 episodes of melena and vomiting twice, without hematemesis. He denied any prodromal symptoms preceding his LOC. He denied any fatigue or confusion following the event.

The patient’s vitals on admission were stable, with a heart rate of 60 beats per minute, blood pressure (BP) of 155/85, an oxygen saturation of 99% on room air, and he was apyrexial. His abdomen was soft, lax, and non-tender. Cardiological, respiratory, and neurological examinations were all unremarkable. Laboratory investigations identified severe anemia with a hemoglobin of 6.6 g/dl and severe hypokalemia of 1.8 mmol/L. Given his clinical presentation and initial findings, it was deemed appropriate not to perform a plain CT head as no head injury was suspected, there was no decrease in consciousness level with no focal neurological signs, and severe blood loss per rectum was sufficient to account for the patient’s syncopal attack. He was then admitted under the medical team for treatment and further investigation of his presentation.

The patient received several transfusions of packed red blood cells (PRBC) as well as potassium chloride intravenously (i.v.) throughout his hospital stay for correction of his anemia and hypokalemia, respectively. The underlying cause of the latter was suspected to be Conn’s syndrome, as a left suprarenal lip- id rich adenoma was identified on computed tomography (CT) of the abdomen and pelvis on day 4 of admission.

After his potassium levels were corrected, the patient underwent multiple endoscopic tests, the first of which was an upper-GI endoscopy conducted on day 4 of admission. This demonstrated gastric antral erosions, but no signs of active or recent bleeding despite continuing to experience melena, with hemoglobin levels of 7-8 g/dl leading up to the investigation.

He underwent another upper-GI endoscopy in addition to a colonoscopy on day 5 of admission, both of which failed to identify any sources of bleeding. In light of this development, a Meckel’s scan was performed on day 6 of admission but was unremarkable. The next day, an abdominal CT angiogram was also unsuccessful in identifying a source of bleeding. Nevertheless, the patient remained under observation as he continued to experience melena.

On day 10 of admission, the patient reported passing a massive amount of melena and hematochezia, resulting in a severe drop in hemoglobin down to 5.8 g/dl. He subsequently underwent an urgent push enteroscopy with a Fujinon colonoscope which was advanced to the mid-jejunum. This revealed a bleeding submucosal lesion protruding into the jejunal mucosa. It was approximately 1.2 cm in diameter, with a central ulcer, which was covered by an adherent clot (Figure 1). Diluted epinephrine 6 ml was administered and 2 hemoclips clips were applied. Despite this intervention, the lesion continued to bleed. The surgical team was consulted regarding the case, and it was determined that urgent surgical intervention was necessary to prevent any further blood loss.

The patient underwent an exploratory laparotomy, during which the jejunal mass was identified 40 cm from the duodenojejunal flexure. Partial resection of the jejunal segment containing the lesion was performed, followed by a hand-sewn primary anastomosis, for a total operative time of approximately 90 minutes. The resected sample measured 5.7 cm in length and 4.0 cm in width. It was sent for further histopathological assessment, which later confirmed the diagnosis of a jejunal DL (Figures 2, 3).

On day 1 post-operatively, his blood pressure (BP) was difficult to control, with constant systolic BP readings of <170 mmHg. This was acutely managed via i.v. furosemide and labetalol. The patient’s BP gradually normalized in the following days. Moreover, the patient received 1 unit of PRBC transfused on post-operative day 2 to treat an asymptomatic hemoglobin drop from 9.6 g/dl down to 8.4 g/dl.
FIGURE 1. Enteroscopic images of the jejunal Dieulafoy’s lesion in-situ. The lesion is being viewed from a proximal perspective protruding into the jejunal lumen where an adherent clot is noted on its surface (A) and streaks of blood can be seen heading distally (B).

FIGURE 2. Macroscopic images of the jejunal Dieulafoy’s lesion after resection. Grossly, the jejunal segment featured a small defect in the mucosa through which a protruding blood clot was seen (A). Serial sectioning of this area revealed that the blood clot was centered in the submucosa and measured 1.1×1.0×0.6 cm in size (B).

On post-operative day 4, he was vitally stable. He had experienced no episodes of melena and his abdominal pain at the surgical sites had markedly improved. His urine and bowel functions had returned to normal. He was tolerating oral intake, with no nausea or vomiting. His potassium levels had also stabilized, and he was scheduled for outpatient follow-up with the Endocrinology Department with regards to his Conn’s syndrome diagnosis. Thus, he was deemed fit for discharge with advice to follow up with the surgical team as an outpatient in 2 weeks’ time, but was lost to follow-up.

Discussion

Clinical Presentation

An actively bleeding jejunal DL usually manifests with either melena or hematochezia. It can also present with hypovolemic syncope, as in our patient, a symptom which should prompt immediate investigation as it signifies severe hemorrhage. Therein lies the diagnostic challenge, as this presentation can mimic other more common causes of acute GI bleeding, including peptic ulcer disease, diverticulitis, and inflammatory bowel disease [2]. Our patient also presented...
with uncontrolled hypertension and had considerable spikes in his BP, which was likely due to suspected Conn’s syndrome. This hypertensive state produces a constant pulsatile force, which can increase the risk of the protrusion of a DL through the jejunal mucosa, making it more susceptible to rupture via erosion or minor trauma [2]. The presence of this risk factor could have been associated with the relatively younger age of presentation for our patient, as a study observed <88% of all DLs were found in patients over the age of 50 years, possibly due to a general increase of co-morbidities with age [3]. However, the association between hypertension and the formation of a DL remains unclear. It has been proposed that

Figure 3. Microscopic images of the jejunal Dieulafoy’s lesion after resection stained with hematoxylin and eosin. Examination showed a large-caliber submucosal artery which feeds into the mucosa through a minute discontinuation (A). Thrombosis of the artery was noted with organization and focal papillary endothelial hyperplasia (B).
systemic co-morbidities such as cardiovascular disease and renal failure can impair the normal process of angiogenesis, resulting in formation of abnormal blood vessels [3,4]. This may explain the high incidence rate of hypertension, renal failure, and congestive heart failure in patients admitted with DL, at 58%, 42.5%, and 19.7%, respectively.

**Diagnosis**

This patient had a bleeding DL in the middle-GI tract. While DL is already categorized as a rare occurrence, lesions occurring in the jejunum or ileum are the rarest form of the disease, identified in only 1% of all reported cases [1]. Owing to its rare occurrence and the intermittency of the bleeding events, this DL posed a significant diagnostic challenge and was not visualized using a standard endoscope. Indeed, DL can often be difficult to diagnose and is undetected in up to 30% of initial GI endoscopies [1]. This could explain why up to 6% of patients undergo a diagnostic odyssey, requiring 3 or more endoscopic investigations to establish a diagnosis [1]. Moreover, the non-specific clinical and endoscopic features of a DL may render it particularly difficult to establish a definitive diagnosis based on endoscopic evaluation alone [2].

In this case, the patient underwent 2 upper-GI endoscopies, 1 colonoscopy, and 1 CT angiography of the abdomen, all of which failed to detect the source of hemorrhage. After finally undergoing a push enteroscopy, the bleeding lesion was found in the jejunum. This emphasizes the importance of investigating the middle-GI tract for a source of bleeding if both upper- and lower-GI tract endoscopies are inconclusive. Push enteroscopy has been found to have a diagnostic yield of up to 73% in the detection of jejunal DLs [2,5]. Although the lesion was visualized in this case by push enteroscopy, the final diagnosis was made only after histopathological assessment of the resected lesion following an exploratory laparotomy.

Other potential investigations for examining the middle-GI tract include single- and double-balloon-assisted enteroscopy, which have proven to be the most effective investigations in identifying jejunal DLs. These investigations successfully detect up to 96% and 98% of DLs, respectively [2].

Capsule endoscopy is another minimally invasive method and can be used in the detection of jejunal DLs [1,2]. However, it should be reserved for stable patients due to its potentially low diagnostic yield of 57% and the fact that it cannot provide any form of intervention. Nonetheless, it can be useful in localizing the lesion to determine which approach would be best prior to the use of balloon-assisted enteroscopy [2,6]. Furthermore, despite its relatively low jejunal DL detection rate, 1 algorithm promotes the use of capsule endoscopy in GI bleeds of unknown origin that have already undergone adequate upper- and lower-GI endoscopies, as its findings would aid decision-making with regards to more invasive investigations and treatments [7].

Radiological modalities such as CT angiogram of the abdomen may be considered, especially in hemodynamically unstable patients. However, it was unremarkable in this case, likely due to the intermittency of the bleeding events, a common occurrence in DLs [1]. Surgical methods, typically exploratory laparotomy, can be used to diagnose the lesion when other modalities have failed [1].

**Treatment**

Of the 136 jejunal DLs identified in a recent systematic review of the literature, 64% were treated endoscopically, 4% via angiographic embolization, and 32% surgically [2]. Endoscopic techniques can be broadly categorized into injection-based, thermal, and mechanical, the last of which being best in terms of success rates [2,8]. The aforementioned techniques have been employed as monotherapy but can also be used in combination with one another, as this has been shown to increase the rate of achieving primary hemostasis [2,8,9]. Several approaches have been found to be effective with an overall success rate of 85%. However, as was applied in this case, the most frequently tested and proven treatment is the use of hemoclipping, either as monotherapy or concurrently with epinephrine injections at the site of bleeding. This approach was used in 25 individual cases of jejunal DL, with a 92% primary hemostasis rate [2,10]. Hemoclipping has also been shown to be safer than thermocoagulation, as it is associated with a relatively lower risk of bowel perforation [11].

Surprisingly, although this highly reliable treatment regimen was used for our patient, laparotomy was still deemed necessary to adequately control a bleeding jejunal DL, making it one of the few instances of unsuccessful hemoclipping and epinephrine combination therapy. Adding injection sclerotherapy as an adjunct may be beneficial in such cases, as it has been reported to be 100% successful when supplementing hemoclipping, with no incidents of rebleeding in the 18 jejunal DLs in which this approach was implemented [2]. Novel endoscopic treatment modalities could be of further use, as was applied in 1 case study, with the successful use of Hemospray® following failure to control 3 bleeding DLs with epinephrine injections and endoclips [12]. A randomized controlled trial later supported this approach, as Hemospray® was found to be just as effective as the hemoclip, with a non-significant increased rebleeding rate, although it is typically used as a temporary measure rather than a definitive one in the management of DLs [13,14].

Moreover, this case may warrant taking other mechanical alternatives into consideration when endeavoring endoscopic management of DLs. Endoscopic band ligation may be a viable option, with comparable success rates to hemoclipping according to a recent meta-analysis [15]. Nonetheless, endoscopic banding
has its own limitations such as poor visual fields and carries a small risk of bowel perforation [8,9]. Promising results have also been reported with over-the-scope clipping in the management of DLs [2,16].

Following the advancement and high success rates of endoscopic techniques, there was a consensus in the literature that surgery is only indicated in the management of refractory DLs or in cases of hemodynamic instability, with segmental resection and anastomosis identified as the most commonly implemented surgical technique [1,2,8,10]. In this case, the jejunal DL was refractory to endoscopic management; therefore, surgical intervention was deemed necessary.

Because DLs often lack pathological changes such as visible fibrosis or inflammation, it would be very difficult to accurately determine its location intra-operatively; as such, pre-operative localization is considered essential for successful resection. Perhaps not a novelty but an anecdote worth mentioning during this particular surgery was the use of enteroscopically-applied hemoclips as a landmark for accurate resection of the lesion. In cases where hemoclips are not used, pre-operative localization can be achieved using endoscopic tattooing, methylene blue dye, or Tc-99m red blood cells [2,17].

In total, 43 patients with jejunal DLs were managed with immediate surgery and 12 following the failure of less invasive treatment modalities with a 100% success rate, underscoring the advantage of surgery in eliminating the risk of rebleeding [2].

Another treatment modality mentioned in the literature is transarterial embolization. Despite its value in the management of gastric DLs, lesions of jejunal origin were very difficult to definitively treat using this approach, for it was only attempted in 5 cases, with no success [2]. This observation could be attributed to the relatively higher mobility of the jejenum and its reduced wall thickness, which can hinder optimal embolization [2,18].

Outcomes and Follow-Up

Even though jejunal DLs can be very difficult to identify initially, once treated, the prognosis is excellent. Our patient was no exception, as he was able to make a complete post-operative recovery and returned to a normal degree of daily functioning within 4 days. This was not the case in the pre-endoscopy era, during which mortality rates ranged between 23% and 35%. It has since been reduced to 4.4-8% with the advancement of non-invasive investigation modalities in addition to increased clinical awareness and surgical expertise [2,9,10]. Interestingly, 1 study examined the clinical outcomes of patients with hypertension who had a GI bleed due to DL, and concluded that there was an increase in mortality and morbidity rates in patients with hypertension compared to normotensive patients [19].

In terms of complications, rebleeding was the most common, at 13.4% in all cases managed non-surgically, but this risk can be significantly reduced by using mechanical endoscopic hemoclipping, especially in combination with injection sclerotherapy [2,10]. It is worth highlighting that neither rebleeding nor recurrence were found to be associated with age, sex, or the site of the lesion [2,10].

No follow-up data were obtained from the patient as he did not attend his follow-up appointment and attempts to contact him via telephone failed. In the literature, some studies have capped follow-up at 6 months after discharge, although the length of intervals between visits is debated, likely due to the variation in the times of rebleeding [2,10].

Conclusions

Although several investigations and interventions were required to treat this patient, he was successfully managed through a multi-disciplinary effort to identify the underlying cause of his presentation, and several conclusions can be drawn from the case. Most importantly, the middle-GI tract must be investigated enteroscopically as a potential source of bleeding if upper- and lower-GI endoscopies are inconclusive. Despite a lack of consensus, the evidence appears to indicate that enteroscopically-applied mechanical hemoclips with or without injection sclerotherapy is the criterion standard in treating Dieulafoy’s lesions of jejunal origin, although combination therapy should be considered when monotherapy fails to achieve primary hemostasis. More research is warranted to establish the efficacy of novel treatments. Moreover, DLs can be difficult to definitively diagnose and manage by standard endoscopic methods and may require surgical intervention, particularly in refractory lesions, hemodynamic instability, and in resource-poor settings where trained personnel and device-assisted endoscopies are not readily accessible. Further analysis is needed to assess the possible association between hypertension and the formation of DLs. In conclusion, jejunal DL is a treatable cause of GI bleeding but requires a high degree of clinical suspicion and expertise to diagnose and treat.

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Declaration of Figures’ Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.
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