CASE REPORT

Dieulafoy's lesion of the oesophagus: a case series and literature review

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SUMMARY

Dieulafoy's lesion is a vascular abnormality consisting of a tortuous, dilated aberrant submucosal vessel that erodes the overlying mucosal layer without ulceration. The oesophagus is a very rare location for the lesion. Herein, we report a case series of 3 patients who presented with upper gastrointestinal bleeding. Endoscopy was performed, which showed a large calibre-persistent tortuous submucosal artery protruding into the oesophageal lumen. All the patients were successfully managed with endoscopic intervention using epinephrine injection around the lesion followed by hemoclip application. The present paper highlights the remarkably rare occurrence of oesophageal Dieulafoy's lesion and reviews the pathophysiology, epidemiology and management with a focus on diagnostic challenge encountered in such patients.

BACKGROUND

Even though M.T. Gallard was the first to identify the Dieulafoy's lesion in 1884, it was named after Paul Georges Dieulafoy since his publication in 1898.¹ He believed that the lesion was due to the erosion of the gastric mucosa, therefore, he called it 'Exulceratio simplex'. Over time, it was believed that aneurysm of the gastric wall in combination with atherosclerosis cause such lesions.^{1 2} Acquired and congenital malformations of the gastrointestinal vessels were also considered as the possible aetiologies of the lesion.³ However, it is now agreed that the lesion is caused by an abnormally dilated vessel, up to 1–3 mm in diameter that runs under the mucosa of the gastrointestinal tract.³ These aberrant blood vessels when reach to the mucosa cause life-threatening bleeding due to the erosion of the vessels.

CASE PRESENTATION

Case 1

A 53-year-old Caucasian man was brought to the emergency department with melena for the last 1 week. Melena was initially a teaspoon in amount, but it was progressively worsening from the last 2 days. He denied fever, abdominal pain, fatigue, abdominal distension, weight changes or haematemesis.

On presentation, the patient was afebrile with a normal heart rate and blood pressure. He was pale with anicteric sclerae. Rest of the systemic examination was unremarkable.

Laboratory results revealed an initial haemoglobin level of 13 g/dL which dropped to 10 g/dL on the second day of hospitalisation. He had markedly

increased blood urea nitrogen to serum creatinine ratio of 55. Upper endoscopy was performed, which showed an adherent blood clot in the distal oesophagus, with fresh red blood oozing from the surrounding of the clot (figure 1). A small vessel was seen protruding into the lumen without an evidence of ulceration (figure 2).

Case 2

A 68-year-old Caucasian male was admitted to our medical centre after a motor vehicle accident. On day 2 of his admission, he started to have fresh rectal bleeding. The patient denied nausea, haematemesis, abdominal pain or distention. He had no history of any chronic disease or hospitalisation.

On examination, he had pallor and multiple abrasions on the left arm. He was well oriented to time, place and person, and had normal vital signs. Cardiovascular, gastrointestinal and chest examination was also unremarkable.

Laboratory studies revealed normal platelet and white cell counts, but his haemoglobin level dropped from 12 to <8 g/dL during the hospital stay. The coagulation profile was within the normal limits and the hepatitis serologies were all negative. Upper endoscopy revealed a bleeding vessel protruding into the lumen of the distal oesophagus through a small mucosal defect (figure 3).

Case 3

An 80-year-old Caucasian man presented to the outpatient department with haematemesis for the last 2 days. Blood was bright red and was a cup full of an amount. He denied any nausea or previous similar episodes. The vomitus was non-projectile

Figure 1 Upper gastrointestinal endoscopy revealing an adherent clot.

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Figure 2 A blood vessel protruding into the oesophageal lumen without surrounding ulceration consistent with Dieulafoy's lesion.

and was yellowish in colour with bright red streaks of blood. The patient had no history of alcoholism, chronic liver disease or bleeding disorders.

On physical examination, he had a normal heart rate of 80 bpm, blood pressure of 130/80 mm Hg and respiratory rate of 18 breaths/min. He was lying comfortably in bed and had no pallor or scleral icterus. His systemic examination was insignificant.

Laboratory evaluation revealed a normal metabolic and renal profile. His liver function tests and urinalysis were also within normal limits. Hepatitis serologies were negative. He had the haemoglobin level of 14 g/dL, and platelet and white cell counts of 250 000/ μ L and 7000/ μ L, respectively. Upper endoscopy showed a small vessel spurting bright red blood into the lumen of the oesophagus (figure 4).

DIFFERENTIAL DIAGNOSIS

On presentation, the differentials were broad and included portal hypertension, bleeding disorders, angiodysplasia, Mallory-Weiss syndrome and a small ulcer with bleeding vessels. However, negative hepatitis serologies, normal coagulation profile and endoscopy findings ruled out all these possibilities.

TREATMENT

All the three patients were initially resuscitated with an intravenous fluid. Additionally, 2 units of packed red blood cells were transfused to cases 1 and 2. They were kept 'nil per oral' (NPO) in anticipation of an endoscopic management. On endoscopy,

Figure 3 Upper endoscopy demonstrating a protruding vessel in the distal oesophagus.

Figure 4 Oesophagoscopy revealing a vessel protruding into the lumen of the distal oesophagus.

epinephrine was injected around the blood clot in case 1, which was then removed with a snare. In all cases, two to three hemoclips were deployed endoscopically at the base of the vessels (figure 5). No further bleeding was evident after the procedure.

OUTCOME AND FOLLOW-UP

All the patients were discharged home in a stable condition and were advised to have a close follow-up with the gastroenterologist. None of these patients reported any further episode of bleeding on their three subsequent monthly follow-up visits.

DISCUSSION

Dieulafoy's lesion accounts for ~1.5% of acute gastrointestinal bleeding.¹ Previously, most of the cases described in the literature documented the involvement of the stomach and lower gastrointestinal tract. Veldhuyzen van Zanten *et al*⁴ in their review of 101 cases showed that 81% of cases of Dieulafoy's lesion were found in the stomach. Similarly, reports of the lesion involving the duodenum, ileum and colon are also available.⁵⁻⁸ However, a structured literature search of the PubMed database showed that only four cases of the oesophageal Dieulafoy's lesion have been reported to date.⁹⁻¹² We present a series of three more cases of the upper gastrointestinal bleeding secondary to Dieulafoy's lesion of the oesophagus.

Figure 5 Hemoclip deployed at the base of the vessel to stop the bleeding.

A comprehensive review of the previously reported cases showed that almost all patients with Dieulafoy's lesion of the oesophagus were male and above the age of 30. More than 90% of the cases were reported from the USA, and the most common presentation was either haematemesis or melena. Urgent endoscopy and epinephrine injection around the lesion were successful in all the cases, including ours with an exception of one case that was found dead at the time of presentation (table 1).

Patients with Dieulafoy's lesion are usually asymptomatic or may present with painless gastrointestinal bleed without associated prodrome of gastric symptoms, seen in ulcer disease. On average, 44% of symptomatic Dieulafoy's lesions present with melena, 30% with haematemesis, 18% with both melena and haematemesis, 6% with haematochezia and 1% with iron-deficiency anaemia.¹³ The amount of bleeding ranges from mild self-limiting episodes to the life-threatening recurrent and profuse bleeding. Patients with the comorbidities such as hypertension, chronic kidney disease, alcoholism, cardiovascular disease and diabetes are relatively at a higher risk to bleed.¹⁴ Interestingly, a significant portion of bleeding occurs in patients hospitalised for other causes pointing towards physiological stress as an inciting factor for arterial perforation.⁹ In our Case 2 prior history of motor vehicle accident and hospitalisation probably triggered the Dieulafoy's lesion to bleed. Moreover, it is hypothesised that the pulsatility of the large submucosal artery inflicts local microtrauma and ischaemia to the mucosa, causing it to thin over time. Once the artery is exposed to the contents of the stomach or bowel, the arterial wall erodes and eventually rupture, resulting in massive gastrointestinal bleeding.^{1 15} Some correlation with the non-steroidal anti-inflammatory drug and aspirin use has also been suggested due to the fact that these medications may cause mucosal atrophy and an ischaemic injury.¹⁶

Dieulafoy's lesion poses a formidable diagnostic challenge as the lesion is usually quite small, bleed only intermittently, and can often be confused with other vascular abnormalities such as aneurysms, arteriovenous malformations or even Mallory-Weiss tears.^{16–18} In the absence of an active bleeding, the lesion can occasionally be seen as a raised nipple or a visible vessel without an associated ulcer. However, aberrant blood vessels can only be visualised during the times of active bleeding.¹⁹ Sometimes, the

lesion may be covered with a clot or obscured by active bleeding making it difficult to identify. According to Reilly *et al*,¹⁶ only 49% of Dieulafoy's lesions are identified during the initial endoscopic examination, 33% of patients require a second examination and 18% require exploratory laparotomy for the accurate diagnosis. It may also be easily missed on endoscopy due to the lack of mucosal inflammation and ulceration. Therefore, it is important to suspect Dieulafoy's lesions in all patients with no clear source of gastrointestinal bleeding and endoscopic ultrasonography should be considered to confirm the diagnosis in the equivocal cases.¹⁹

Radiological techniques such as contrast-enhanced CT and angiography have also been described as helpful, particularly in cases where initial endoscopy fails to diagnose a Dieulafoy's lesion.²⁰ Although diagnosis with a CT scan has frequently been rarer than angiography, near-definitive diagnosis of a Dieulafoy's lesion can be obtained with an enhanced CT of the abdomen.^{15 21} Therefore, it is important to keep CT scan or angiography in mind while investigating the less common sites like oesophagus as a possible site of the lesion, especially when no other source of bleeding is found on gastrointestinal endoscopies.

Surgical procedures such as proximal gastric resection and wedge resection have traditionally been considered the treatment of choice for Dieulafoy's lesion. However, endoscopic therapeutic intervention has recently replaced surgery, with a reported success rate exceeding 90%.^{1 9 15} Endoscopic haemostasis can be achieved with thermal or argon plasma coagulation, regional injection of epinephrine or cyanoacrylate and mechanical therapy like banding and hemoclippping.^{9 15 22} Mechanical methods are often more effective compared to epinephrine injection and coagulation methods, and mechanical therapy in combination with epinephrine injection has lower rebleeding rates compared to monotherapy.^{9 15} However, band ligation is associated with a high risk of perforation in cases of small bowel and colon lesions.²³ Apart from the endoscopy, arterial embolisation by angiography has also been reported successful in selected cases.¹⁸ Following treatment, a Doppler ultrasound can be used to confirm the successful ablation of a lesion by documenting the absence of blood flow.²⁴ Hence, laparoscopic or open surgical procedures should be reserved only for difficult-to-control bleeding.

Table 1 Characteristics of the previously reported cases of Dieulafoy's lesions of the oesophagus

Author	Rikke Kjærgaard <i>et al</i>	Jayaram Thimmapuram <i>et al</i> ¹⁰	Philip Abraham <i>et al</i> ¹¹	Schneider DM <i>et al</i> ¹²	Faisal Inayat <i>et al</i>
Country	USA	USA	India	USA	USA
Publication Journal/Year	The American Journal of Forensic Medicine and Pathology/2012	Gastrointestinal Endoscopy/ 2011	Indian Journal of Gastroenterology/2004	The American Journal of Gastroenterology/1994	The present case series
Age/gender	48/M	38/M	54/M	NA	53,68,80/M
Main symptom	Haematemesis, melena, stomach pain	Aspiration pneumonia, melena, haematemesis	Painless haematemesis	NA	Haematemesis and melena
Comorbidity	Benign prostatic hyperplasia	Mental retardation	Diabetes and hypertension	NA	None
Endoscopy findings	Mucosal defect with a ruptured artery in the distal oesophagus	Mucosal defect with a ruptured artery in the mid-oesophagus	Oesophageal varices with active spurting from vessels	NA	A vessel protruding from a small mucosal defect in the distal oesophagus
Treatment	None	Epinephrine injection followed by hemoclip application	Epinephrine injection and argon plasma coagulation	NA	Epinephrine injection followed by hemoclip application
Outcome	Death	Recovered	Recovered	NA	Recovered
Reference	9	10	11	12	The present case series

Delay in the diagnosis and inadequate management due to the lack of clinical familiarity make the Dieulafoy's lesion a stealth killer.^{5 25} Goldman review of 24 cases reported 19 deaths which give mortality as high as 80%, highlighting the urgent diagnosis and management of the lesion.²⁶

Learning points

- ▶ Dieulafoy's lesion, a dilated aberrant submucosal vessel which erodes the overlying epithelium, is a relatively rare, but potentially fatal, cause of gastrointestinal bleeding.
- ▶ Physicians should maintain a high index of clinical suspicion for this uncommon condition in an unusual location such as oesophagus while dealing with the cases of upper gastrointestinal bleeding.
- ▶ Knowledge of the Dieulafoy's lesion and its manifestations can help guide clinicians to make the correct diagnosis, which can then help improvise endoscopic and endovascular localisation and treatment, ultimately improving the patient outcomes.

Contributors WU and FI did the literature review and QH and AH worked on the discussion and managed the patients.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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