CASE REPORT

Extragastric Dieulafoy's lesion

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Accepted 26 August 2014

SUMMARY

A 74-year-old man on warfarin for aortic valve replacement presented with recurrent episodes of melaena. An initial oesophagogastrroduodenoscopy (OGD) was normal, as were red cell scanning and colonoscopy. It was a third OGD that revealed the cause of the melaena—a vascular lesion in the duodenum, at the junction between D1 and D2. An extragastric Dieulafoy's lesion was diagnosed, and the lesion was injected with epinephrine and tattooed. Over the following months, episodes of bleeding recurred despite further attempts at injection. Percutaneous radiologically assisted embolisation of the gastroduodenal artery, and eventually duodenotomy and oversuturing of the lesion were performed to no avail. The patient has undergone over 10 endoscopies, and has received over 70 units of packed red cells to date, since his initial presentation 6 years ago. Attempts to stop the bleeding permanently have been difficult, highlighting the complexity of managing such a lesion.

BACKGROUND

Acute upper gastrointestinal (GI) bleeding is a common medical emergency. Most cases will be found to have gastro-oesophageal varices, peptic ulcer disease or mucosal erosions; these are often readily identified at endoscopy.¹ The cause of bleeding is obscure in 5% of patients,² and 1–2% of cases have been attributed to Dieulafoy's lesion.³

This lesion consists of a histologically normal vessel with an abnormally large diameter, which maintains a constant width.⁴ It is most commonly found in the stomach, but has been identified in other parts of the GI tract, and even outside it.

Erosion or ischaemia of the mucosa overlying a previously asymptomatic lesion leads to its usual presentation with melaena and/or haematemesis.⁵ Despite being an uncommon cause of bleeding, Dieulafoy's lesion is potentially life-threatening and must be included in the differential diagnosis in any patient who presents with acute GI haemorrhage.

While GI endoscopy is the standard initial investigation, the first attempt will only reveal the lesion in 70% of patients,⁶ and multiple endoscopies may thus be required. Alternative investigations that may be useful in the diagnosis include wireless capsule endoscopy, red cell scanning techniques and angiography.

Treatment modalities include endoscopic repair, which has the highest success rates, and surgery. The lesion is often refractory to treatment, as reflected in the case report.

CASE PRESENTATION

Our patient is a 74-year-old man with a history of aortic valve replacement for aortic stenosis. He was started on warfarin with a target international normalised ratio (INR) of 2.0–3.0.

He presented with an episode of melaena in 2007, just over a year after starting anticoagulation. He denied any other GI symptoms, anorexia or weight loss.

Blood tests revealed haemoglobin of 11 g/dL. The patient was advised to omit his warfarin for 2 days, after which the melaena resolved spontaneously. An oesophagogastrroduodenoscopy (OGD) showed no abnormalities other than a positive CLO test and the patient was prescribed triple therapy.

He had another episode of melaena 2 months later and a repeat OGD was also normal, save for a positive CLO test. Triple therapy was once again prescribed and he was kept on omeprazole.

Episodes continued over the following months and his haemoglobin was occasionally found to be as low as 5 g/dL. He required transfusion on multiple occasions.

INVESTIGATIONS

In view of two negative OGDs, a diagnosis of obscure GI bleeding was made, and red cell scanning was carried out in 2008. This showed no abnormal foci suggestive of an active GI bleed up to 4 h postlabelling. However, a 24 h delayed image showed activity in the caecum and ascending colon, indicative of active bleeding.

A colonoscopy was thus organised to investigate this possible source of bleeding. A normal vascular pattern was observed and a benign adenomatous polyp was detected.

A mesenteric angiogram showed no abnormalities. A third OGD was performed in 2010—this revealed a vascular lesion in D1 (figure 1).

Episodes of bleeding continued over the following 2 years, and repeat OGDs were performed, each time demonstrating a rebleed.

TREATMENT

When the lesion was first diagnosed, it was injected with epinephrine and tattooed endoscopically. This was repeated during follow-up OGDs.

In 2010, 3 years following presentation, percutaneous radiologically assisted embolisation of the gastroduodenal artery was undertaken via a right femoral approach. The bleeding was still refractory to this treatment and a decision was taken to perform a duodenotomy. No actual bleeding was observed during the operation and oversuturing of the lesion was performed.

OUTCOME AND FOLLOW-UP

Since his initial presentation with bleeding 6 years ago, the patient has undergone over 10 OGDs and received a total of 77 units of packed red cells.

Although the patient still reports the occasional passage of dark stools, his haemoglobin level has

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To cite: Gauci J, Galea S, Galea J, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-204627

Figure 1 Dieulafoy's lesion in the first part of the duodenum (seen above the duodenal lumen).

dropped significantly on only one occasion since oversuturing of the lesion. The surgery has therefore resulted in a dramatic improvement in the patient's requirement for admission and transfusion and hence his quality of life.

Follow-up OGDs have demonstrated the recurrence of bleeding from the same lesion, highlighting its refractory nature. The patient was kept on omeprazole indefinitely. In order to maintain anticoagulation for his aortic valve replacement, but avoid the occurrence of such episodes, tight control of the patient's INR had to be achieved.

DISCUSSION

Obscure GI bleeding is defined as persistent and/or recurrent bleeding from a source that cannot be readily determined by initial standard investigations, including bidirectional endoscopy and barium studies.⁷ It is most commonly due to a vascular cause, such as Dieulafoy's lesion.

Studies have attributed only 1–2% of acute GI bleeding to such lesions.³ However, as they are asymptomatic until presentation, it is likely that lesions are under-recognised, as they are easily overlooked or misidentified at endoscopy.⁸ They were previously believed to be acquired in nature; however, more recent reports have described cases occurring in newborns.⁹ This may suggest that they are actually congenital lesions.

Analysis of the case studies presented to date has enabled researchers to discern a common patient profile. Dieulafoy's lesion is twice as common in males and is more common in the elderly.³ Patients typically have multiple comorbidities, and are often on medications such as non-steroidal anti-inflammatory drugs, aspirin and warfarin.¹⁰ The index case described in this case report has several of the characteristics of the typical patient profile.

Dieulafoy's lesion is commonly on the lesser curve of the stomach, usually within 6 cm of the gastro-oesophageal junction.¹¹ Lesions have also been reported in the distal oesophagus, small intestine, colon, rectum and anal canal.¹¹ In our patient, the lesion was located in the duodenum (figure 1). A recent case report also described a lesion outside the GI tract.¹²

The standard initial investigation is GI endoscopy. Criteria for diagnosis include the detection of active arterial spurting or streaming from a normal-looking mucosa or from a mucosal defect <3 mm in size, a protruding vessel, once again in the absence of an associated ulcer, and/or a fresh clot densely adherent to a normal-looking mucosa.¹³ The first endoscopy will

identify 70% of patients⁶ though other lesions may be more elusive.¹⁴ Studies have suggested the use of a heparin bolus to provoke bleeding to allow for easier identification of the lesion.¹⁵

Other means of investigation can be used if endoscopy has been unsuccessful. These include wireless capsule endoscopy, endoscopic ultrasound, red cell scanning techniques and angiography.^{13 14 16 17} The latter two investigations were negative in our patient.

There are several treatment modalities of repair. Endoscopic repair is the first-line choice in easily accessible lesions,⁸ and includes thermal methods, mechanical methods and regional injection with local epinephrine or sclerotherapy.⁵ Combination therapies give the best results⁶—in hindsight, perhaps such an approach should have been adopted in the index patient.

Angiography and embolisation of the actively bleeding lesion is another non-invasive method of treatment, as was attempted in the index patient.⁸ This treatment should be particularly considered for patients who are unfit for surgery, who have failed to respond to endoscopic therapy or in whom the lesion is beyond the reach of therapeutic endoscopy.¹⁴

Surgery is currently being reserved for the 5–10% of cases that are not amenable or not satisfactorily treated by endoscopy.^{3 16} This takes the form of under-running of the vessels, wide wedge resection or the more radical gastrectomy.³ Recently, the use of laparoscopic wedge resection has been described.¹⁸

Rebleeding is a significant risk and may be as high as 9–40%. Once again, we reiterate the option of using combination endoscopic therapies to control the bleeding, since endoscopic monotherapy has been associated with higher rates of rebleeding.^{6 10}

Patient's perspective

- ▶ The index patient was asked to attend a short interview. He accepted immediately, asking us to consider this as a token of his appreciation for the care he was given over the years.
- ▶ Despite having been diagnosed 3 years ago, he has still not understood the exact nature of his condition, and the reason why he has experienced persistent bleeding. He does not appreciate the importance of achieving such tight control over his international normalised ratio (INR). Interestingly, he does not know the name of his condition, though he claims that this does not trouble him.
- ▶ The patient understands that his condition is rare and accepts the fact that it has taken us so long to reach a diagnosis. He is very grateful for the close monitoring and follow-up provided.
- ▶ When asked about the impact of the condition on his life, he claims to be "tired of coming in and out of hospital". He recognises that the surgery has led to a great improvement in his quality of life, however, we noted a degree of apprehension: "every time I open my bowels, I check the colour of my stools."
- ▶ Listening to our patient's outlook on his condition was definitely an eye-opener. Although he is not bothered by his unfamiliarity with the condition, this undoubtedly reflects a lack of clear explanation on our behalf.
- ▶ As Dieulafoy's lesion is so refractory to treatment, it will inevitably have a bearing on the patient's quality of life. Nevertheless, we feel that providing the patient with more information throughout the years could potentially have alleviated some anxiety and given him a better grasp of his condition.

Learning points

- ▶ A diagnosis of obscure gastrointestinal bleeding is made when the initial two oesophagogastroduodenoscopies are negative. It comprises 5% of all cases of gastrointestinal bleeding.
- ▶ Dieulafoy's lesion is one cause of obscure gastrointestinal bleeding and may be detected in any part of the gastrointestinal tract and also outside it.
- ▶ Despite being uncommon, bleeding following rupture of a Dieulafoy's lesion is a potentially life-threatening situation. Such a lesion must therefore be kept in mind in any patient who presents with acute gastrointestinal haemorrhage.

Contributors JG drafted most parts of the paper. SG went through the patient's clinical records for the case presentation. JG and MS were instrumental in contacting the patient for his consent; they also revised the draft paper.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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