CASE REPORTS

Dieulafoy Disease of Stomach - An Uncommon Cause of Gastrointestinal System Bleeding

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Abstract

Dieulafoy disease is an uncommon cause of gastrointestinal system bleeding. It is characterised by bleeding from abnormal submucosal arteriole. Endoscopic therapy and surgery are the preferred treatment option for this lesion. Paul Georges Dieulafoy (1839–1911), a professor of pathology in Paris, France, was the first to describe a series of 10 patients who presented with Dieulafoy disease. The advent of endoscopy has drastically changed the process of diagnosing and treating Dieulafoy lesions, with techniques such as endoscopic banding, haemoclips, thermocoagulation, and injections with adrenaline as alternatives to replace surgical management as was described to be a best practice management.

Introduction

Dieulafoy disease is an uncommon cause of gastrointestinal system bleeding. Although the exact cause is not known, it is characterised by bleeding from abnormal submucosal arteriole. Endoscopic therapy and surgery are the preferred treatment option for this lesion.

Case Report

A 42 yr old male presented with history of 2 to 3 episodes of haematemesis per day and melaena since 3-4 days. There was no history of alcohol intake or NSAID consumption. The clinical examination was significant for pallor and tachycardia (heart rate 114/min). His haemoglobin was 5.8 g/dl. Coagulation parameters were normal. Patient underwent oesophagogastroduodenoscopy (OGD) at our institute which was suggestive of suspicious lesion with ulceration above it (Figure 1). Patient managed conservatively and was given

Fig.1: Oesophagogastroduodenoscopy suggestive of suspicious lesion with ulceration (arrow)

Fig.2: Oesophagogastroduodenoscopy showing active spurt (arrow) from the Dieulafoy’s lesion

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injection pantoprazole drip but, he still continued
to have hematemesis and melena. On performing
repeat EGD we noticed active spurt from the lesion,
suggestive of Dieulafoy lesion (Figure 2). Endoscopic
argon plasma coagulation attempted but haemostasis
was not achieved, so patient posted for emergency
partial gastrectomy. Intra-operative findings revealed
a patent vessel with surrounding normal mucosa
(Figure 3). Surgery was uneventful and patient
improved post surgery. Histopathology report was
consistent with dieulafoy lesion (Figure 4)

Discussion

Paul Georges Dieulafoy (1839–1911), a professor of
pathology in Paris, France, was the first to describe
a series of 10 patients who presented with massive
haematemesis due to a bleeding gastric vessel,
without any evidence of ulceration in the first three
lessons of the 1897-1898 edition of “Clinique Médicale
de l’Hôtel-Dieu.”. Dieulafoy concluded that this
lesion was not a typical gastric ulcer and named
it an “exulceratio simplex,” which in time became
known as a “Dieulafoy lesion”. Modern literature
has broadened the original definition of a “Dieulafoy
lesion.” Initially only described in the stomach, such
findings have been additionally reported in the rest
of the gastrointestinal tract such as the oesophagus,
duodenum, ileum, jejunum, colon, anal canal, and
rectum. Dieulafoy lesions have also been reported
in non gastrointestinal sites such as the bronchus.

The advent of endoscopy has drastically changed
the process of diagnosing and treating Dieulafoy
lesions, with techniques such as endoscopic banding
(ELB), haemoclips, thermocoagulation, and injections
with adrenaline as alternatives to replace surgical
management as was described to be a best practice
management.

In approximately 4-9% of massive upper
gastrointestinal haemorrhage, no demonstrable cause
can be found. Dieulafoy’s lesion is thought to be the
cause of acute and chronic upper gastrointestinal
bleeding in approximately 1-2% of these cases. The
incidence, however, might vary from 0.5% to 14%. It
is thought to be more common in males (M:F = 2:1)
with a median age of 54 years at presentation. There
is usually no significant NSAIDs or alcohol abuse.

Dieulafoy’s lesion is caused by an abnormally large-
calibre persistent tortuous submucosal artery. This
has been demonstrated by histological examination
of resected specimens. The artery protrudes through
a solitary, tiny mucosal defect (2-5 mm), commonly
in the upper part of the stomach. It may rupture
spontaneously and lead to massive bleeding. It
has been suggested that the thin mucosa overlying
a pulsating artery is eroded progressively by the
mechanical pressure from the abnormal vessel.

Histologically, the eroded artery appears normal.
There is no evidence of any mucosal inflammatory
process, signs of deep ulcerations, penetration of the
muscularis propria, vasculitis, aneurysm formation,
or arteriosclerosis.

The most common presenting symptom is recurrent,
often massive, haematemesis associated with melaena
(51%). Our patient presented in same way. The lesion
may present with haematemesis alone (28%), or
melaena alone (18%). Characteristically, there are
no symptoms of dyspepsia, anorexia or abdominal
pain. Initial examination may reveal haemodynamic
instability, postural hypotension and anaemia. The
mean haemoglobin level on admission has been
reported to be between 8.4- 9.2 g/dl in various studies.

The endoscopic presentations of Dieulafoy’s lesion
are as follows: (1) a superficial notch in the gastric
mucosa, blood vessels in the mucosa, and coagulum
on its surface; (2) a focal defect of lesser curvature
of stomach mucosa within 6 centimetres of GO junction
complicated with active bleeding; (3) small arteries
can protrude on the mucosa and active bleeding can
occasionally be detected; and (4) occasionally, blood permeation can be detected from the mucosa, and is often detected when bleeding.8

An OGD can successfully identify the lesions in approximately 82% of patients. Approximately 49% of the lesions are identified during the initial endoscopic examination, while 33% require more than one OGD for confident identification. The remainder of the patients with Dieulafoy’s lesions are identified intraoperatively or angiographically. In some recent series, however, the identification of the lesions was more accurate at the initial endoscopic examination, as Dieulafoy lesions were identified in up to 92.3% and 96.4% of cases.8

Traditionally, the treatment of DL was surgical. However, with the development of endoscopic haemostasis techniques, the need for surgery has been reduced, and the mortality rates have decreased from 80% to 8.6%. Therefore, the current treatment of choice in accessible lesions is endoscopy, with a success rate of > 90% and low rates of recurrence and complications. The endoscopic haemostasis procedures are classified into three groups: (1) thermal: electrocoagulation, heater probe and argon plasma coagulation; (2) local injection of substances, such as adrenaline or sclerosing solutions; and (3) mechanical: haemostatic clips and bands. All these procedures achieve high rates of primary haemostasis, with low rates of recurrence (generally <10%). However, in the clinical field, the choice of a procedure depends on the experience, decision of the endoscopist and the field of vision.9

Criteria for determining successful endoscopic haemostasis

The criteria for determining successful endoscopic haemostasis were as follows.

(1) Endoscopic demonstration: blood spurting or capillary haemorrhage stopped, and the endoscopic field of view became clear; (2) Clinical manifestations: there was no haematemesis or dark stool after treatment, blood pressure rose to a normal range and was stable, and pulse rate decreased to normal range.

Criteria for determining unsuccessful endoscopic haemostasis

Criteria for determining unsuccessful endoscopic haemostasis were as follows: (1) haematemesis and/or dark stool occurred 48 h after endoscopic treatment; (2) haemoglobin was decreased by more than 2.0 g/L; (3) there was evidence of hypovolaemic shock; and (4) there was manifestation of bleeding and blood transfusion was necessary. Re-bleeding was confirmed at the original site as demonstrated by endoscopic examination.

Surgical procedures currently employed include under-running of the lesion or a wedge resection of the affected section of gut. Our patient underwent surgery too.

Angiography may also be used therapeutically by gelfoam embolisation. This type of treatment is usually reserved for patients who are not amenable to endoscopic therapy and are poor surgical candidates.

Following endoscopic management, there was no recurrence of bleeding from Dieulafoy lesions over a mean long-term follow-up of 28 months in one series, and 36 months in another.10 This indicates that endoscopic treatment of the Diuelafoy lesion is safe and effective, with very good long-term results in the accessible lesions.

References