The Dieulafoy gastric malformation: an under-recognized cause of massive upper gastrointestinal haemorrhage

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Summary: The Dieulafoy gastric malformation is a rare cause of upper gastrointestinal haemorrhage. When no obvious bleeding lesion is seen at laparotomy this diagnosis ought to be considered. Three such cases were identified and treated with simple underrunning of the lesion with no mortality and minimal morbidity. Follow-up endoscopy in each patient showed complete healing of the lesion.

Introduction

We present three cases of massive upper gastrointestinal haemorrhage from the Dieulafoy vascular malformation. Fewer than 150 such cases have been reported in the literature, yet we have seen three cases in a 4-month period in a district general hospital. We feel that this is an under-recognized cause of massive upper gastrointestinal haemorrhage, and one which must be carefully sought in the absence of an obvious bleeding lesion.

Case reports

Case 1

A 59 year old man was admitted with an 8-hour history of periumbilical pain, haematemeses and malaena. He had a history of dyspepsia over the preceding 5 months, and his alcohol intake was 8 pints of beer each day. On examination he was pale and sweaty with a tachycardia of 130 beats/minute and blood pressure 115/65 mmHg. Haemoglobin estimation was 13.3 mmol/l, urea 14.6 mmol/l. He was resuscitated with intravenous fluids, catheterized, and transfused 3 units of blood.

Four hours after admission he had a further massive haematemesis, passed fresh malaena, and became shocked. Urgent gastroscopy was performed: there was no evidence of oesophageal varices, but the stomach was full of fresh blood and clot, preventing visualization of any lesion there. Laparotomy was performed through an upper mid-line incision. An initial duodenotomy revealed no evidence of duodenal ulceration, and therefore a long anterior gastrotomy was performed. Despite a careful search no bleeding lesion was identified, but blood was seen to be coming from the region of the cardia. By palpation a small abnormality 1 cm distal to the cardia on the lesser curve was identified, and with careful positioning of retractors a split in the mucosa with a bleeding vessel projecting through was seen. The lesion was treated by underrunning with several linen sutures.

The patient made an excellent post-operative recovery, having received a 10-unit blood transfusion in all, and was discharged after 8 days. A follow-up gastroscopy after 2 months showed no evidence of the lesion.

Case 2

A 49 year old man presented with a 12-hour history of haematemeses, malaena and dizziness. On examination he was pale but not shocked. Haemoglobin estimation was 13.6 mmol/l, with a urea of 16.4 mmol/l. He was resuscitated with intravenous fluids and 2 units of blood. Urgent gastroscopy was performed, excluding oesophageal varices and duodenal ulceration, but fresh blood and clot prevented adequate views of the gastric mucosa. He had further episodes of malaena, and after 4 more units of blood, haemoglobin estimation was only 8.0 g/dl. Laparotomy was therefore performed, via an upper midline incision. Opening the stomach by an anterior gastrotomy all blood and clot was evacuated, and no obvious bleeding lesion was seen. The region of the cardia was therefore

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palpated, and an abnormality felt 2 cm distal to the
cardia on the lesser curve side. This was seen to be a
split in the mucosa with a large artery in its base, a
branch of which was projecting through the
mucosal defect and bleeding continuously. The
lesion was underrun with several linen sutures.
Twelve units of blood had been transfused in all.
The patient made an excellent recovery and was
discharged on the eighth post-operative day. At
follow up gastroscopy 2 months later, the area was
a little erythematous and one linen stitch was still
present, but the lesion had otherwise healed.

Case 3

A 78 year old woman presented with a 2-day
history of indigestion and sudden collapse. On
examination she was pale and shocked. Haemoglo-
bin estimation was 5.6 mmol/l with a urea of
24.0 mmol/l. She was quickly resuscitated with
intravenous fluids and 4 units of blood. Twelve
hours later she had a massive haematemesis asso-
ciated with further hypotension, and a laparotomy
was therefore performed. The duodenum was
scared but there was no bleeding lesion. A long
anterior gastrotomy was made and the stomach
emptied of fresh blood and clot, but no obvious
lesion was seen. An abnormality near the cardia
was identified by palpation, and when visualized
was a split in the mucosa 1 cm distal to the cardia
on the lesser curve, with a large blood vessel in its
base capped by some thrombus. The lesion was
underrun with several linen sutures.

She made an uneventful post-operative recovery,
having received 9 units of blood in all, and was
discharged after 17 days. Follow-up gastroscopy 2
months later showed complete healing of the
lesion.

Discussion

Although originally described by Gallard in 1884,
the Dieulafoy gastric erosion or vascular malfor-
mation was so named in 1896. The lesion consists
of a small mucosal erosion with a cap of thrombus
overlying a ruptured submucosal vessel, with a
predilection for the lesser curve 1–3 cm from the
cardia. The lesion was originally thought to be
caused by an aneurysm of a submucosal vessel, but
more recent reports believe that the affected vessel
represents part of a localized congenital malforma-
tion, with possibly focal pressure from the enlarged
vessel on the overlying gastric mucosa, precipi-
tating erosion of the mucosa and exposed vascular
wall. However, histological examination of such
lesions has not shown the typical changes of
endarteritis which are usually seen in response to
peptic ulceration. Voth believes that the primary
lesion is the superficial erosion which occurs in a
part of the stomach where an abnormally large
artery happens to run through the submucosa. This
may explain why the majority of the lesions arise
along the proximal part of the lesser curve of the
stomach, where the arrangement of blood vessels in
the submucosa differs from the rest of the stom-
ach.

The patient typically presents with massive
haematemesis and melaena, with circulatory
shock. The reported age range is 20 months to 93
years, with the majority between the ages of 50–70
years. The male:female ratio is 2:1. There may be a
previous history of peptic ulceration or alcohol
abuse, but generally there are no obvious pointers
to the diagnosis from the history and examination.

Urgent gastroscopy is required and may provide
the diagnosis but often one is faced with perform-
ing a laparotomy without knowing the source of the
bleeding. A history of non-steroidal anti-
inflammatory drug ingestion or previous duodenal
ulceration would prompt one to perform an initial
duodenotomy, but if no lesion is found an anterior
gastrotomy should be made, and the gastric
mucosa carefully examined for the source of bleed-
ing. This usually requires the help of at least one
assistant holding two retractors inside the stomach
in order that the region of the fundus and cardia
may be visualized. If no lesion is seen, a finger is
inserted into the distal oesophagus and gently
brought back into the stomach, carefully feeling the
mucosa. In each of the three cases described, a
slight mucosal abnormality was felt on the lesser
curve just distal to the cardio-oesophageal junc-
tion, and with repositioning of the retractors and
the use of suction the mucosal defect and bleeding
lesion was visualized.

The literature recommends wedge excision of the
Dieulafoy vascular malformation; however, the
reported morbidity and mortality is relatively high.

Our experience suggests that simple underrunn-
ing of the bleeding lesion with 3 or 4 linen sutures is
the correct emergency treatment of this bleeding
lesion. Each of our patients made an uneventful
post-operative recovery and endoscopic follow-up
in all of them has shown the lesions to be healed.

Patients may afterwards be treated with H2
antagonists; the small number of patients who
require subsequent elective surgery will be deter-
mined by clinical and endoscopic criteria.

In conclusion, the Dieulafoy gastric malforma-
tion is a cause of massive upper gastrointestinal
haemorrhage which, in the absence of any obvious
bleeding lesion, must be carefully sought. Digital
palpation in the region of the cardia will identify
the lesion which is not immediately visible. Simple
suture ligation of the lesion is the treatment of
choice, with mandatory endoscopic follow-up.
References

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