Progress report

Recurrent massive haematemesis from Dieulafoy vascular malformations – a review of 101 cases

Dieulafoy vascular malformations can be easily overlooked as a cause of massive, often recurrent haematemesis. Six cases are described and a review is given of 101 cases described in the literature. Bleeding is caused by an unusually large artery that runs in the submucosa and lies in close contact with the mucous membrane. In more than 80% of the cases the site of bleeding is found within 6 cm of the gastric-oesophageal junction, particularly at the site of the lesser curve. The preference for this location can be explained by the vascular architecture of the gastric blood supply. Diagnosis can be made during emergency gastroscopy. Sometimes repeated endoscopies are necessary. Surgery is the best treatment and a wide wedge resection is recommended.

A well defined cause for massive upper gastrointestinal haemorrhage cannot be found in approximately 4–9% of the cases. Superficial gastric mucosal lesions have been recognised as a rare cause for abundant gastric bleeding by Dieulafoy, who described three cases in 1898. Because of the high mortality rate if the diagnosis is not made, we report on our experience of six cases during the period 1977–1984 and discuss the pathogenesis of this lesion.

Patients

Case 1
A previously healthy 26 year old man without a history of gastric complaints presented with repetitive haematemesis. This was soon followed by the passage of tarry stools and shock. Two gastroscopies, an angiography and a scan with technetium labelled erythrocytes were all negative. The third gastroscopy, done during the acute phase of renewed bleeding, revealed a spurting blood vessel close to the cardia at the posterior wall of the stomach. The mucosa surrounding the bleeding spot was unremarkable. During acute gastrotomy this bleeding vessel was ligated. After four years of follow up the patient is in excellent health.

Case 2
A previously healthy 41 year old man was admitted to the hospital with repeated haematemesis, melaena and shock. A history of gastric complaints was absent. Gastroscopy was negative. Five days later urgent laparotomy was done for recurrent severe haematemesis accompanied by shock. An ‘oozing’ superficial vessel high in the fundus of the stomach was
coagulated after which a truncal vagotomy was carried out. Two days later he was transferred to our hospital because of recurrent massive haematemesis. Gastroscopy and angiography both were negative. An urgent second laparotomy was done. No bleeding point was found and a technically difficult B-II resection was carried out. The patient died 18 days later because of recurrent haematemesis. Histologic sections of the resected stomach showed an abnormally wide blood vessel in the submucosa running towards the proximal site of resection.

**CASE 3**
A 71 year old previously healthy man was admitted to the hospital because of massive repetitive haematemesis. There was no history of peptic ulcer disease. The third gastroscopy carried out during an acute bleeding episode showed a spurting vessel high in the fundus. Laser coagulation was unsuccessful. During acute gastrotomy the vessel was ligated. One month later he died of sepsis of unknown origin. Necropsy revealed no abnormalities in the stomach except for the signs of the ligation.

**CASE 4**
A 30 year old man with moderate congenital insufficiency of the tricuspid valve (Ebstein's syndrome) was admitted to the hospital because of haematemesis. He had no previous history of stomach problems. When the massive haematemesis recurred a second gastroscopy was carried out. A small, actively bleeding site was seen high in the fundus along the posterior wall of the stomach close to the greater curve. During emergency gastrotomy the bleeding vessel was ligated. After two years of follow up the patient is in excellent health.

**CASE 5**
A 37 year old man with a history of alcoholic pancreatitis presented with massive repetitive haematemesis. During the third gastroscopy a bleeding point was seen high in the fundus along the posterior gastric wall. During emergency gastrotomy the lesion was ligated. On the sixth postoperative day massive gastrointestinal bleeding recurred. A technically difficult total gastrectomy was undertaken. No evidence of liver cirrhosis was present and pressure in the splenic vein was normal. One day later he died of gastrointestinal bleeding in combination with pulmonary aspiration. Necropsy did not reveal the site of bleeding. Histological sections of the resected stomach showed abnormal vessels high in the fundus in the submucosa, protruding into the mucosa with some widened capillaries beneath the surface epithelium. Despite the fact that serial sections of the whole stomach were made no ulceration was found (Fig. 1).

**CASE 6**
A 26 year old man, who had experienced mild gastric complaints during the last year, was admitted because of massive upper gastrointestinal haemorrhage. After three days of conservative management haematemesis recurred. An upper gastrointestinal series revealed a small contrast depot in the upper part of the stomach at the site of the lesser curve. Four days later he again suffered from haematemesis and was transferred to our hospital. During gastroscopy a superficial gastric erosion was seen in the
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lower part of the fundus on the greater curve. A diagnosis of Dieulafoy's lesion was made. During laparotomy this diagnosis was confirmed and a wide wedge resection of the lesion was performed. The patient made an uneventful recovery and two months later is still in excellent health. Histology showed a small well defined erosion of the mucous membrane invading the wall of an artery lying in the submucosa. The size of the artery was abnormally wide for its location and was lying in direct contact with the mucous membrane. The lumen of the artery was partially obliterated by a recent trombus. The internal layers of the vessel wall showed a normal constitution (Fig. 2).

Pathogenesis

Even though Gallard was probably the first to describe the lesion in 1884, Dieulafoy’s name has been associated with it since his publication in 1898.56 He believed the lesion to be the initial stage of a gastric ulcer, whose further development was interrupted by the bleeding episode. Therefore he called the superficial gastric mucosal lesion ‘Exulceratio simplex’. Originally it was thought that the lesion was caused by an aneurysm in one of the vessels within the gastric wall, sometimes in combination with arteriosclerosis.5-10 Congenital or acquired vascular malformation has also been considered as a cause.11 12 It is now generally agreed that the bleeding is caused by an unusually large artery that runs immediately under the gastric mucosa.4 13 The vessel always has an abnormally wide calibre, up to 1.0-3.0 mm in diameter and runs through the submucosa with a tortuous course for variable distances.9-11 14 15 As

Fig. 1 Mucous membrane of the stomach without erosion. Dilated and tortuous vessels in the submucosa are seen, locally penetrating the mucosa (MSB-staining ×21 original magnification).
such it can approach the mucous membrane and bleeding can be caused by erosion of the artery if only a mucosal erosion is present. The size of the mucosal defect is usually small, varying from 2–5 mm.\textsuperscript{10 13 16}

Histologically there are no signs of deep ulceration with penetration of the muscularis propria.\textsuperscript{11 13 14 17} Aneurysms, arteriosclerosis or signs of vasculitis are generally absent.\textsuperscript{11 13 14} The point of bleeding into the gastric lumen can be seen as a necrosis of the involved artery wall where intimal fibrosis is sometimes present.\textsuperscript{13} The occurrence of bleeding of unusually large arteries from only superficial erosion has been explained by Voth through the architecture of the blood supply to the stomach.\textsuperscript{18 19} There is a pattern of regular reduction in arterial branch size when the arteries penetrate the gastric wall, but sometimes so-called calibre-persistent arteries are seen in the submucosa. Voth does not consider this vessel pattern as a congenital abnormality, but more the upper limit of normal variation. He believes that the superficial erosion is the primary lesion which occurs in a ‘locus minoris resistentiae’ of the stomach, namely the place where the abnormally large artery runs through the submucosa. And therefore only a superficial erosion can give rise to massive bleeding. In addition, meticulous study of the gastric blood supply has shown that in the region of the lesser curve the submucosal arteries do not arise from an extensive plexus of large vessels in the submucosa as is the case in the rest of the stomach.\textsuperscript{20} Instead they arise directly from the arterial chain along the lesser curve outside the stomach. Even though it was found that the arrangement of the rich ‘spongeliike’ blood supply in the mucosa was the same in all parts of the stomach the absence of a submucosal plexus does support the idea of calibre-persistency as it has been described by Voth.
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Together with the fact that the submucosal plexus in the antrum consists of smaller vessels, these findings might be an explanation for the fact that bleeding from Dieulafoy's lesions occurs much more frequently in the upper part of the stomach. We believe that the abnormally large artery runs in the submucosa and is lying in close contact with the mucous membrane over a variable distance. Massive bleeding can occur if erosion of the mucous membrane and arterial wall is present (Fig. 2).

Clinical pattern

All our patients showed the typical clinical pattern as it has been described in the literature. Massive, often repeated haematemesis is usually the presenting symptom in combination with passage of tarry stools and circulatory shock.1 5 12 The youngest patient described was 20 months old, the eldest 93 years.12 21 In the 101 cases that we collected from the literature, including our own six cases the mean age was 52 years and the median 54 years.1 4-17 19 21-41 For the age-distribution see Fig. 3. Dieulafoy's lesions are twice as frequent in men than in women (67:34).1 4-17 19 21-41 Occasionally patients have suffered from repeated haematemesis in the years before admission.9 31 Usually a history of gastric complaints or peptic ulcer disease is absent. In the 101 collected cases these were mentioned only 18 times. Alcohol consumption has been implicated as a contributing factor for bleeding.4 10 13 Heavy alcohol intake was reported in only 15 of the 101 cases, signs of liver cirrhosis were present in four cases.

Diagnosis

The diagnosis is usually made by gastroscopy or during urgent laparotomy.

Fig. 3 Age distribution of 101 cases.
As shown in our cases, however, repeated endoscopies can be necessary because of the intermittent character of bleeding. The endoscopic appearance of a Dieulafoy lesion is variable. Sometimes no abnormality is seen not even upon meticulous inspection. Only with repetitive endoscopies one may suddenly see spurting bleeding from a pinpoint mucosal defect. In other patients there is a small erosive defect usually with a tiny visible vessel and some adherent clot. Occasionally one may see a small arterial vessel sticking out from the normal looking surface, usually accentuated in length because of adherent clot. On rare occasions diagnosis has been made by angiography. The location of bleeding is strikingly more frequent in the upper third of the stomach especially in and around the cardia. In 95 of the 101 collected cases the bleeding site was mentioned. In 78 of these (82%) the lesion was found within 6 cm of the gastric-oesophageal junction. Of the 58 cases where the location was given in relation to the gastric curves 47 cases were close to the lesser curve, that is 81%. The fact that bleeding originates with such high frequency in the upper stomach and close to the lesser curve can be explained by the vascular architecture as has been described above.

**Therapy**

Surgery is the treatment of choice. During gastrotomy a careful inspection of the gastric mucosa is necessary in order to find the Dieulafoy lesion. The region of the cardia should receive special attention because bleeding has its origin there in the majority of cases. When no bleeding site is found and no peptic ulcer is present the surgeon should refrain from carrying out a ‘blind’ Bilroth-II resection. Rebleeding from the upper third of the stomach frequently occurs in the postoperative phase and has a poor prognosis. This is shown by our case 2.

The operative procedures which have been done are coagulation or ligation of the bleeding vessel, proximal gastric resection and wedge resection. In two of our patients rebleeding occurred after coagulation and ligation. We therefore now advocate a wide wedge resection of the area bearing the Dieulafoy vascular malformation as histology has shown that the abnormal artery can run with its tortuous course over a longer distance. One should also keep in mind that occasionally more than one superficial lesion has been seen. Wedge resection has the advantage that the final diagnosis can be confirmed by pathological anatomic examination. Apart from surgery, incidental successes have been reported by arterial embolisation during angiography.

Before the endoscopic era the prognosis of Dieulafoy’s lesion was poor. Goldman in his review of 24 cases reported 19 deaths which gives a mortality rate of 79%. Of all 35 cases, including our own that we collected since 1970, eight died, that is 23%. The most important factor leading to death was that the diagnosis had not been made before death and/or a ‘blind’ Biroth-II resection was done.

**Conclusion**

The incidence of Dieulafoy vascular malformation is not known. Possibly it occurs more frequently than reported in the literature because this lesion...
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can easily remain unrecognised as a cause of massive upper gastrointestinal haemorrhage. When during early gastroscopy for massive haematemesis no bleeding site is found, it is extremely important to repeat endoscopy as soon as any sign of recurrent bleeding is present. In over 80% of all cases the bleeding has its origin within 6 cm of the gastric-oesophagal junction and in particular at the site of the lesser curve. Therefore this region should receive special attention from the endoscopist. The preference for this location can be explained by the vascular architecture of the gastric blood supply. Surgery is the treatment of choice. As the abnormal artery that causes the bleeding can run over variable distances wide wedge resection is recommended. When no bleeding site is found during gastrotomy a ‘blind’ Bilroth-II resection should be avoided.

Addendum

A previously healthy 82 year old woman was admitted to the hospital with recurrent massive haematemesis. She used Sulindac, a non-steroid anti-inflammatory drug for unspecified joint pains. During endoscopy no bleeding site was found and ranitidine was started. Two weeks later massive haematemesis recurred for which she needed a transfusion with 22 units of blood. She was transferred to our hospital. Four days later urgent gastroscopy was done because of haematemesis. Just below the gastric-oesophagal junction at the site of the lesser curve a visible vessel was seen with some adherent clot (Fig. 4). A diagnosis of Dieulafoy vascular malformation was made. A wedge resection was advised but instead a total gastrectomy was done because the surgeon thought the risk of postoperative rebleeding too high. The patient had a stormy postoperative course with pleural empyema and renal insufficiency for which she needed

Fig. 4 An artery with some adherent clot is seen in a small mucosal defect close to the gastric-oesophagal junction.
continuous assisted ventilation. She developed neurological symptoms and died of multiple organ failure. Pathological examination of the stomach showed a large artery penetrating the submucosa and mucosa just below the gastric-oesophageal junction at the side of the lesser curve. There were no signs of inflammation (Fig. 5).

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