Gastric Arteriovenous Malformation Presenting With Upper GI Bleeding And Haemoperitoneum

R Sellayah, G Pande

Citation

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Abstract
Gastric AVMs are a rare cause of upper gastrointestinal bleeding and are difficult to diagnose radiologically, as most are small and submucosal, and even when associated with massive bleeding, are difficult to appreciate endoscopically. Our patient is an unusual case of this phenomenon, presenting with haematemesis and melaena, as well as intraperitoneal bleeding. Furthermore, the histopathological diagnosis revealed not only a gastric AVM but also an incidental leiomyoma, which was mistaken as the culprit lesion intraoperatively. Although endoscopic and endovascular approaches are utilised more and more in the management of these lesions, our case highlights the important of surgery as a definitive procedure in emergencies and a treatment option in elective cases.

BACKGROUND
Most gastric lesions bleed into the gastrointestinal tract and present with haematemesis and melaena. It is uncommon to find a patient with bleeding into the gastrointestinal tract presenting with haemoperitoneum. The presence of haemoperitoneum in a patient with upper gastrointestinal bleeding often signifies perforation of the bowel either because of tumour eroding through, or a perforation of a pseudocyst full of blood. We present an unusual cause of upper gastrointestinal bleeding which also resulted in haemoperitoneum

Gastric arteriovenous malformation (AVM), an abnormal shunt between the arterial and venous vascular systems, is a rare and unusual cause of upper gastrointestinal haemorrhage, with a reported incidence of 1-2% in cases of upper gastrointestinal bleeding [1]. These lesions form because of the failure of the embryonic vascular plexus to differentiate and develop a mature capillary bed. Bleeding from these lesions occurs due to the high pressure resulting from their direct connection to the arterial system.

CASE PRESENTATION
A 74-year-old gentleman was brought into the emergency department of a peripheral hospital after being found collapsed by his wife in the early hours of the morning. She reported a history of 24 hours of vague abdominal pain and one episode of haematemesis and melaena the day before. His past medical history included atrial fibrillation, for which he was receiving warfarin. He was also known to have a 6cm infrarenal abdominal aortic aneurysm, type 2 diabetes mellitus, hypercholesterolaemia and extensive cardiac surgery in 2010, which involved a tissue aortic valve replacement, a Bentall’s procedure and two coronary artery grafts. His wife also reported a history of cognitive decline over several months however no formal diagnosis of dementia had been made at the time of presentation.

On arrival he was unstable with a blood pressure of 90/70mmHg, heart rate of 160 beats per minute, and was hypothermic (34.6 degrees Celsius). His pH was 7.2 (normal range 7.32-7.42) and lactate was 5.6 mmol/L (normal range 0.5-2.2). His haemoglobin was 114 g/L, INR was 3.2 and he had mild acute renal impairment (creatinine 111 umol/L). He was resuscitated with normal saline.

CT of the abdomen (Figure 1) demonstrated what appeared to be a large volume of free fluid with a large mass at the greater curvature of the stomach suggestive of a possible malignancy with haemorrhage, or large volume ascites associated with a gastric malignancy. There was also a smaller, contrast-enhancing lesion at the lesser curvature. A peritoneal tap was performed which showed frank blood. At this stage he was referred to our institution for specialist
upper GI surgical management.

Figure 1
Coronal and axial CT images demonstrating a large clot at the greater curvature with free fluid in the abdomen, as well as a smaller contrast-enhancing lesion at the lesser curvature.

On arrival to our institution he was still haemodynamically unstable despite initial resuscitation, with a haemoglobin level of 102 g/L and INR of 1.3. He was transfused 6 units of blood throughout this time, and his INR was reversed with fresh frozen plasma, prothrombinex and Vitamin K. Given the degree of haemodynamic compromise and the inaccessibility of emergency angiography that evening, the decision was made to proceed to exploratory laparotomy.

The primary diagnosis at this stage was a gastric malignancy, possibly gastrointestinal stromal tumour or lymphoma, with bleeding intra-abdominally and into the gastrointestinal tract.

At laparotomy 1.5 litres of blood was suctioned immediately with a further 2 litres apparent intraperitoneally. A large clot was noted at the greater curvature and there was dense tissue over the lesser curvature (which was difficult to appreciate on CT). The bleeding lesion was identified at the incisura with thickening of the stomach wall noted intraoperatively. A distal gastrectomy with Roux-en-Y gastrojejunostomy was performed.

He was admitted to the intensive care unit post-operatively for monitoring. His admission was complicated by aspiration pneumonia while in ICU requiring broad-spectrum antibiotics and respiratory support. He had a slow recovery in hospital and was discharged back to the referring facility 5 weeks after initial presentation with no further complications.

Formal histology demonstrated ectatic abnormal vessels suggestive of an AVM, with an incidental leiomyoma of the stomach in the same region.

DISCUSSION
An AVM is a congenital lesion that involves the persistence of an abnormal connection between arteries and veins. This is thought to be the result of failure of the embryonic vascular plexus to differentiate fully into a mature capillary bed in a specific area, the structure of which is affected by changes due to aging [2].

Gastrointestinal AVMs account for 1-2% of upper GI haemorrhage [1, 3]. The stomach is a particularly unusual location, accounting for 1.4% of these cases, the caecum and ascending colon being more common (77.5%) [4]. Most cases present with chronic anaemia, recurrent haemorrhage or acute massive haemorrhage, however there are cases discovered incidentally. There is an equal distribution between the genders, with most patients aged between 60 and 80 years of age [5]. Our patient presented with both upper gastrointestinal haemorrhage as well as intraperitoneal haemorrhage. This is an unusual combination rarely reported in the literature.

The pathogenesis is still unclear, however it is postulated to be a combination of congenital (failure in vascular development) and mechanical factors (distension and contraction of the bowel wall intermittently occluding submucosal veins leading to the development of dilated, tortuous vessels in the submucosa and mucosa) [6]. This may explain the distribution in the right colon and caecum, as this area has the greatest wall tension.

Morphologically, AVMs appear as nests of veins, venules and capillaries, the vascular channels of which are sometimes only separated from the gastrointestinal lumen by a layer of epithelial cells [6]. There may be large, dilated vessels, sometimes extending into the muscle, and individual vessels are often difficult to characterize as either arterial or venous. Although the diagnosis of gastric AVM was made post-operatively in our case based on microscopic
examination, the clinical picture intraoperatively was clouded by the presence of a leiomyoma very close to the site of bleeding. Furthermore, the CT finding of a large gastric mass originating from the greater curvature was found intraoperatively to be a large blood clot, with the lesion located at the incisura.

The aetiology of gastrointestinal AVM has been suggested to be an acquired degenerative cause resulting from changes due to aging [7]. Most AVMs occur in the 60–80 year old age group [8], however some have been reported in the paediatric age group [5,9], raising the possibility of a congenital aetiology in these cases. There are also reports of gastrointestinal bleeding from AVMs associated with left ventricular assist devices. There are a few postulated causes of this. One is that the chronic decrease in pulse pressure caused by aortic stenosis leads to decreased perfusion, causing ischaemia and stimulating the development of new fragile vessels likely to bleed [10]. Another is related to the development of increased sympathetic tone resulting in smooth muscle relaxation and angiodyplasia. A further theory is the development of an acquired type of von Willebrand disease due to the creation of a high shear stress environment and impaired platelet aggregation [11].

A system for classification of AVMs was proposed by Moore et al [4] in 1976 which differentiated them into three categories: Type 1 (single, localised, microscopic lesions that typically present later in life), Type 2 (congenital lesions that present earlier in life), and Type 3 (gastrointestinal lesions that are associated with hereditary haemorrhagic telangiectasia). Our case presented here would be classified as Type 1 given the age of the patient and the solitary lesion found under microscopic examination.

Despite the suggested aetiology of an acquired degenerative cause of gastric AVMs, the current cases reported in the English-speaking literature suggest that the disease can occur at any age, ranging from 14 to 80 years of age, with no predilection towards gender (Table 1). There are a variety of presentations ranging from acute haemorrhage [12–14] to chronic abdominal pain and/or gastrointestinal bleeding [2, 4, 5, 15–17] to iron deficiency anaemia [18–21]. A small number were discovered incidentally [7, 22]. Matsuda et al reported in the Japanese literature 38 cases of gastrointestinal AVM [23]. Of these, 5 were gastric and presented with acute bleeding. Although lesions have been found in most parts of the stomach except the pylorus, a tendency to towards lesions occurring in the antrum and proximal third of the stomach has been described [24].

Endoscopy and angiography was performed in most cases. Some were managed only with angiographic embolization [2, 25, 26], endoscopic coagulation or clipping [17] or both [14], but most proceeded to open surgery, either after endoscopy and/or angiography [5, 7, 12, 27] or immediately if the clinical situation warranted urgent laparotomy or angiographic intervention was not available [13, 15, 18, 19, 28], as in our case.

AVMs are difficult to diagnose, however they are the most common cause of both chronic bleeding and acute massive haemorrhage when a diagnosis cannot be made with endoscopy, EUS or angiography [8]. Gastric AVMs may be diagnosed with endoscopy, however the gold standard for diagnosis is selective mesenteric angiography. Nevertheless, an actively bleeding lesion may be difficult to diagnose with endoscopy, and a slow intermittent bleed may prove difficult to differentiate with angiography [5], thus it remains a difficult diagnosis to make radiologically. Endoscopic ultrasound may be useful to diagnose submucosal AVMs, and also permits visualisation of gastric wall structures. It has been particularly useful in the diagnosis of the submucosal type of AVM as it can demonstrate the site, depth and shape of the lesion and also allows differentiation between the lesion and other structures in the gastric wall and surrounding tissues [7].

There are several factors that make endoscopic diagnosis difficult. Small AVMs can be hidden between folds or be mistaken for endoscopic trauma. Some small lesions may even fade due to shock causing blood to be shunted away from the gastroduodenal circulation [25]. After reviewing 47 cases of vascular malformations of the stomach and duodenum, Moreto et al [24] proposed an endoscopic classification system: Pattern 1 (flattened or slightly protruded bright red lesions with a “frond-like” margin), Pattern 2 (the “telangiectatic form”) and Pattern 3 (a submucosal nodular type, the most difficult to diagnose).

Selective mesenteric angiography has been used in the investigation of gastrointestinal bleeding since the 1960s. It is now indicated in cases where endoscopic control of bleeding is unsuccessful or if endoscopy is unable to localize the site of bleeding [29]. Moore et al [4] determined that the diagnostic accuracy of angiography rises from 50–75% to almost 90% if the patient is actively bleeding. Building on the angiographic criteria for diagnosis outlined by Boley et al in 1977 [30], Charbonnet and colleagues [31] have more recently defined direct (contrast extravasation) and indirect signs (vascular tuft, arteriovenous fistula, an early filling
vein or a hypervascular mass) to diagnose active bleeding or to define the site of bleeding.

Management of a bleeding gastric AVM may be endoscopic, angiographic, operative or a combination of these. While endoscopic coagulation or angiography and embolization may be successful in treating smaller gastric AVMs, complete resection is recommended for larger lesions or those that continue to bleed [7], and is the most common method employed. Post-operative bleeding due to incomplete initial removal of the lesion or the presence or development of another AVM in the gastrointestinal tract, has also been described [30] [7, 32]. Therefore these patients require careful monitoring in the acute post-operative phase, as well as regular follow up studies.

Although endoscopic and endovascular approaches are utilised more and more in the management of these lesions, our case highlights the important of surgery as a definitive procedure in emergencies and a treatment option in elective cases.

Table 1
Summary of gastric AVM cases reported in the English-speaking literature

<table>
<thead>
<tr>
<th>Case</th>
<th>Description</th>
<th>Location</th>
<th>Treatment</th>
<th>Outcomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>57-year-old</td>
<td>Gastric</td>
<td>Endoscopic</td>
<td>Bleeding</td>
</tr>
<tr>
<td>2</td>
<td>65-year-old</td>
<td>Duodenal</td>
<td>Embolisation</td>
<td>No bleeding</td>
</tr>
<tr>
<td>3</td>
<td>42-year-old</td>
<td>Jejunum</td>
<td>Surgery</td>
<td>Complete</td>
</tr>
</tbody>
</table>

References
22. Araki, T. and T. Wakabayashi, Gastric Arteriovenous Malformation Presenting With Upper GI Bleeding And Haemoperitoneum
Author Information

Renishka Sellayah, General Surgical Registrar
Department of Surgery at Launceston General Hospital
Tasmania, Australia

Girish Pande, General Surgeon
Department of Surgery at Launceston General Hospital
Tasmania, Australia