CASE REPORT

Primary aortojejunal fistula: a rare cause for massive upper gastrointestinal bleeding

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SUMMARY

A 68-year-old man presented to the emergency department with haematemesis and shock. Upper endoscopy and selective angiography could not identify the source of bleeding. He underwent selective embolisation of the gastroduodenal artery. The patient then had a period of about 24 hours with relative haemodynamic stability before having another episode of massive upper gastrointestinal bleed. A second attempt to embolise the common hepatic artery and distal coeliac axis was unsuccessful. Hence, he was urgently taken to the operating room for exploratory laparotomy. The source of bleeding could not be identified in the operating room. The patient went into cardiac arrest and expired. Autopsy revealed a fistula between proximal jejunum and a previously unknown abdominal aortic aneurysm (AAA). We present an entity that has only been described a few times in the literature while highlighting the importance of having a broad differential with upper gastrointestinal bleeding, especially when the source is not clearly evident.

BACKGROUND

Aortoenteric fistula (AEF) is a pathological communication between the aorta and any portion of the gastrointestinal (GI) tract. They can be classified into primary aortoenteric fistula (PAEF) and secondary aortoenteric fistula (SAEF). SAEF is the more common entity (PAEF represents only ~0.4%–1.6% of all types of AEF),¹ and it typically occurs as a complication from aortic reconstructive surgery where a fistulous tract is formed between the aorta and an adjacent portion of the bowel due to chronic, low-grade perigraft infection and the repetitive pressure on the intestine from aortic pulsations.² In contrast, PAEF is rare, and has an incidence of 0.04%–0.07% in the general population and 0.69%–2.36% of patients with an abdominal aortic aneurysm (AAA), as reported in a large autopsy study.³ Aortoduodenal fistulae are the most common type (~60%) due to the close relationship of the duodenum (particularly the D3 and D4, as they are attached to the retroperitoneum) with the underlying abdominal aorta.⁴ Less frequently, PAEF does occur between the abdominal aorta and other portions of the alimentary tract including proximal small bowel, colon and oesophagus.^{5,6}

Patients with PAEF can present with a classical triad of symptoms known as the Cooper's triad which includes upper GI bleeding (64%),

abdominal pain (32%) and pulsatile abdominal mass (25%),² but these three features are concomitantly present in only 11% of patients.⁷ Patients who present with GI bleeding are usually evaluated with a esophagogastroduodenoscopy (EGD). However, the detection rate of PAEF with EGD is only 25%.¹ We describe the case of a 68-year-old man who presented to the hospital with massive upper GI bleed due to a primary aortojejunal fistula likely caused by previously unknown AAA.

CASE PRESENTATION

A 68-year-old man with a history of severe ischaemic cardiomyopathy with cardiac stent and ventricular assist device placement about a year prior on aspirin and clopidogrel, presented to the emergency department with complaints of diaphoresis, dizziness and nausea. At intake, his vitals showed a systolic blood pressure of 70 mmHg. While in the emergency department, he had an episode of emesis with a large amount of bright red blood and a bowel movement with maroon stools. His initial haemoglobin was 8.8 g/dL down from 10.8 g/dL (from a previous admission 5 months prior). He was fluid resuscitated and 2 units of packed red blood cells were administered. While arrangements were being made to transport the patient for an emergent endoscopy, the patient became unresponsive with severe bradycardia and arrested. A dose of epinephrine was administered with return of spontaneous circulation.

INVESTIGATIONS

The patient was intubated in the emergency department and was taken to the endoscopy suite for an upper endoscopy. This procedure revealed large clots in the distal oesophagus, stomach and duodenum, covering almost 80% of the mucosal surface. By this time, he was haemodynamically stable. He was then taken to the interventional radiology suite where angiography was performed which did not show any active bleeding but severe stenosis of the coeliac artery was noted. Subselective angioembolisation of the gastroduodenal artery (GDA) was performed with the presumption that the patient was bleeding from a duodenal ulcer in an attempt to prevent another bleeding episode. Notably, a previously unknown AAA was identified during distal washout of contrast in the angiography films. However, no active extravasation was noted in the anterior/posterior view (see figure 1). He remained haemodynamically stable for nearly

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Figure 1 An approximate 5 cm abdominal aortic aneurysm was identified during selective angiography and attempted angioembolisation. No active extravasation is seen on this anterior posterior view.

24 hours. An EGD was performed the following morning, which redemonstrated a large amount of blood clots in the stomach and duodenum with no signs of active bleeding. A few hours later, he became bradycardic and hypotensive with bright red blood seen in the nasogastric tube. His haemoglobin level dropped from 8.1 g/dL to 6.1 g/dL. He was urgently taken to the interventional radiology suite where he underwent embolisation of common hepatic artery and distal coeliac axis near its branching points into left gastric, common hepatic and splenic arteries for presumed gastric source of haemorrhage with confirmed reduced flow. Despite the angioembolisation, his bleeding persisted with blood coming out of his mouth and around the nasogastric tube. He was taken directly to the operating room for an emergent laparotomy.

DIFFERENTIAL DIAGNOSIS

Patients presenting with haematemesis, as in our case, are almost always found to have an upper GI source of bleeding. The most common cause of upper GI bleed (UGIB) remains peptic ulcer disease and accounts for up to 67% of UGIB cases.⁸ Hence, peptic ulcer disease was at the top of the differential in this case as well. Other less common aetiologies of UGIB are listed in [box 1](#). The initial work-up for any suspected UGIB is upper endoscopy which not only confirms the source of bleeding but also allows potential control of the bleeding.⁹ In our case, an oesophageal source was ruled out, but given the large clot burden in the stomach and duodenum, the exact source of bleeding could not be identified on endoscopy.

When an obscure GI bleed (OGIB) is suspected, details pertaining to the nature of bleeding (eg, haematemesis), bleeding diathesis (eg, von Willebrand disease), comorbidities and systemic symptoms (eg, vasculitis, amyloidosis), drug use (eg, non-steroidal anti-inflammatory drugs), prior procedures (eg, AAA repair) and prior radiation and family history (eg, malignancies) may provide important clues regarding the underlying aetiology.¹⁰

Box 1 Differential diagnosis for obscure upper gastrointestinal (GI) bleed

- ▶ Differential diagnosis for upper GI bleed
- ▶ Oesophageal varices
- ▶ Oesophageal ulcers
- ▶ Gastritis
- ▶ Gastric varices
- ▶ Cameron erosions
- ▶ Non-steroidal inflammatory drug-induced gastropathy
- ▶ Vascular lesion (angioectasias)
- ▶ Malignancy (adenocarcinoma, GI stromal tumour)
- ▶ Mallory-Weiss tear
- ▶ Portal hypertensive gastropathy
- ▶ Haemobilia
- ▶ Haemosuccus pancreaticus
- ▶ Aortoenteric fistula
- ▶ von Willebrand disease
- ▶ Amyloidosis

TREATMENT

In the operating room, a midline laparotomy incision was made. The peritoneal cavity contained serous fluid without any free blood. His stomach was tensely distended with no significant distension of the small bowel. A longitudinal gastrotomy along the anterior portion of the stomach was made. Copious amount of fresh blood and old clots were encountered. The clots were removed and the stomach was packed. The gastrotomy was extended proximally and there was no evidence of ulceration or areas of active bleeding in the fundus and proximal stomach. The gastrotomy was then extended across the pylorus and onto the proximal duodenum. Copious amount of bright red blood was noted in the duodenum. The haemorrhage was difficult to control. In the mean time, the patient had multiple episodes of hypotension with brief periods of cardiac arrest. Advanced cardiac life support (ACLS) protocols were followed with return of spontaneous circulation. The second and third portions of the duodenum were mobilised and area of GDA was compressed with a little success in controlling bleeding. Blunt aortic compression in the supraceliac region was attempted with minimal success. By this time, the patient had lost spontaneous circulation again. Since the source of bleeding could not be identified with ongoing exsanguination, resuscitation efforts were stopped and the patient expired.

OUTCOME AND FOLLOW-UP

After the patient expired, the abdominal packs were removed and the area of the ligament of Treitz was palpated. A palpable mass-like structure was encountered in the proximal jejunum. An enterotomy was made directly over this area. There was an inflammatory mass with a circular hole in it. Subsequently, an autopsy was performed. It showed an ulcer in proximal jejunum communicating with a previously unknown, 6 cm saccular atherosclerotic AAA (see [figure 2](#)).

DISCUSSION

The most common cause for primary AEF is previously unknown atherosclerotic AAA.⁵ Less common causes include infectious causes like syphilis, tuberculosis, mycotic infection and collagen vascular disease where the chronic inflammation leads to aortitis, erosion and formation of the fistula. The mechanism of fistula

Figure 2 Photograph at time of autopsy showing the posterior proximal jejunal defect which was openly communicating with the aorta.

formation likely starts with ischaemia and subsequent necrosis of the intestinal wall due to repetitive traumatic pulsations of the adjacent aortic aneurysm. Consequently, ischaemia of the bowel wall could result in GI contamination of the aneurysmal wall leading to aortic perforation and fistula formation.²

This rare disease entity could present with upper GI haemorrhage, abdominal pain and a palpable abdominal mass, described as the Classic or Cooper's triad. But the triad is only seen in approximately 11% of cases.⁷ Other less common findings include back pain, melaena, fever, syncope or a 'herald bleed'. 'Herald' or 'premonitory' haemorrhage is a very important aspect of the clinical picture associated with primary AEFs. A 'Herald bleed' is an initial bleeding episode that precedes the catastrophic exsanguination where the bleeding is small enough that it is temporarily sealed off through thrombus formation.⁴ This then progresses to expansion of the fistula and dislodgement of the thrombus due to the high blood pressure within the aorta leading to the massive haemorrhage. According to a review performed by Steffes and O'Leary, in 29% of the cases, the time between initial bleed and death is greater than 1 week,¹¹ emphasising the importance of early diagnosis and its influence on the prognosis of the patients. In our case, the patient had a 'Herald' bleed approximately 24 hours prior to the catastrophic haemorrhage, which was initially not recognised as such.

The diagnosis is not evident until the time of emergency surgery in as much as 50% of cases.¹² Since majority of these cases present with upper GI bleed, endoscopy has traditionally been the first diagnostic tool used in these patients. However,

the overall sensitivity of upper endoscopy for detecting an AEF is only about 50%.¹³ If the source of bleeding is not clearly identified by upper endoscopy, other modalities like CT angiography, percutaneous aortic angiography, nuclear scan, colonoscopy and video capsule study are warranted based on clinical suspicion. The modality of choice for detecting an AEF is CT scan of the abdomen with intravenous contrast. It is readily available, less invasive and superior to endoscopy. It may show abnormal communication between the aorta and the bowel and loss of continuity of the aneurysmal wall.² The presence of periaortic gas in the presence of GI bleeding was 100% specific for an AEF.¹⁴ Performing a CT angiography earlier in the diagnostic process could not only lead one to earlier diagnosis but it will also guide the clinician in planning an intervention whether it is embolisation or operative intervention.

Our patient did not have any prior aortic work-up or repair. However, he did have aortic cannulation for his cardiac catheterisation and ventricular assist device placement less than a year prior to his UGIB episode. Notably, there was no reported complications from his cardiac catheterisation procedures. Although primary AEF due to recurrent trauma caused by his previously unknown AAA is the most likely aetiology of the AEF as numerous reports postulate in literature, whether or not a wire injury to the aortic wall during his catheterisation was the leading event toward the formation of his AEF is up to debate.

PAEF, when left untreated is fatal in 100% of cases.⁴ The mortality rate is 74% if surgical intervention is delayed until a catastrophic bleed.¹⁵ During a life-threatening haemorrhage,

use of intra-aortic devices such as Resuscitative Endovascular Balloon Occlusion of the Aorta (REBOA) could provide early aortic occlusion and transiently stabilise patients.¹⁶ A systemic review of REBOA use showed that it can increase the systolic blood pressure of patients in haemorrhagic shock by 53 mmHg, thereby allowing additional time to perform definitive treatment.¹⁷ Notably, successful use of intra-aortic balloon devices in massive UGI bleeds have been reported.^{18–20}

The definitive treatment most commonly involves open surgery, culture and debridement of the aneurysmal aorta, repair with *in situ* prosthetic graft and primary repair of the GI tract when there is no gross contamination of the peritoneal cavity. If the patient has favourable aortic anatomy, endovascular aneurysm repair (EVAR) is an attractive option.²¹ However, a systematic review by Antonious *et al*²² showed that over 50% of the patients with AEF who underwent EVAR developed bleeding and infectious complications postulated to be due to placing synthetic material in a potentially infected field. Extra-anatomic bypass is a described treatment option in grossly contaminated cases, but several studies and literature reviews showed a much higher mortality rates in patients with that approach.^{23 24} Post-operatively, the patient should be treated with intravenous broad spectrum antibiotics for up to 6 weeks according to the culture results.⁵

In conclusion, PAEF is a rare cause for upper GI bleeding. One needs a high index of suspicion especially when the endoscopy is inconclusive for an obvious source of bleeding, and there is no known history of AAA as in this case. Performing a CT scan of the abdomen might lead to an earlier diagnosis and improved outcomes with surgical management in these patients.

Learning points

- ▶ Primary aortoenteric fistula (PAEF) is a pathological communication between the aorta and any portion of the gastrointestinal (GI) tract on someone with no prior history of aortic procedures.
- ▶ If the source of upper GI bleeding is not clearly identified by endoscopy, further radiographic investigation is warranted.
- ▶ The most common cause of PAEF is an unknown abdominal aortic aneurysm.
- ▶ A 'Herald bleed' could precede a catastrophic haemorrhage in this condition.
- ▶ Treatment for PAEF is emergency surgery involving aortic aneurysm repair with graft and primary repair of the GI tract.

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REFERENCES

- 1 Kane JM, Meyer KA, Kozoll DD. An anatomical approach to the problem of massive gastrointestinal hemorrhage. *AMA Arch Surg* 1955;70:570–82.
- 2 Singh M, Koyfman A, Martinez JP. Abdominal vascular catastrophes. *Emerg Med Clin North Am* 2016;34:327–39.
- 3 Olcott C, Holcroft JW, Stoney RJ, *et al*. Unusual problems of abdominal aortic aneurysms. *Am J Surg* 1978;135:426–31.
- 4 Gordon AC, Agarwal M. Primary aorto-enteric fistula. *Int J Surg Case Rep* 2016;19:60–2.
- 5 Philippakis GE, Moustardas M. Surgical treatment of primary aortojejunal fistula. *Int J Surg Case Rep* 2013;4:477–9.
- 6 Karthaus EG, Post IC, Akkersdijk GJ. Spontaneous aortoenteric fistula involving the sigmoid: a case report and review of literature. *Int J Surg Case Rep* 2016;19:97–9.
- 7 Saers SJ, Scheltinga MR. Primary aortoenteric fistula. *Br J Surg* 2005;92:143–52.
- 8 Rotondano G. Epidemiology and diagnosis of acute nonvariceal upper gastrointestinal bleeding. *Gastroenterol Clin North Am* 2014;43:643–63.
- 9 Nable JV, Graham AC. Gastrointestinal bleeding. *Emerg Med Clin North Am* 2016;34:309–25.
- 10 Pasha SF, Hara AK, Leighton JA. Diagnostic evaluation and management of obscure gastrointestinal bleeding: a changing paradigm. *Gastroenterol Hepatol* 2009;5:839–50.
- 11 Steffes BC, O'Leary JP. Primary aortoduodenal fistula: a case report and review of the literature. *Am Surg* 1980;46:121–9.
- 12 Brown PW, Sailors DM, Headrick JR, *et al*. Primary aortojejunal fistula: a case report. *Am Surg* 1999;65:139–41.
- 13 Korkut AK, Arpinar E, Yasar T, *et al*. Primary aorto duodenal fistula complicated by abdominal aortic aneurysm. *J Cardiovasc Surg* 2000;41:113–5.
- 14 Hughes FM, Kavanagh D, Barry M, Owens B, *et al*. Aortoenteric fistula: a diagnostic dilemma. *Abdom Imaging* 2007;32:398–402.
- 15 Flye MW, Thompson WM. Aortic graft-enteric and paraprothentic-enteric fistulas. *Am J Surg* 1983;146:183–7.
- 16 Napolitano LM. Resuscitative endovascular balloon occlusion of the Aorta: indications, outcomes, and training. *Crit Care Clin* 2017;33:55–70.
- 17 Morrison JJ, Morrison JJ, Galgon RE, *et al*. A systematic review of the use of resuscitative endovascular balloon occlusion of the aorta in the management of hemorrhagic shock. *J Trauma Acute Care Surg* 2016;80:324–34.
- 18 Karkos CD, Bruce IA, Lambert ME. Use of the intra-aortic balloon pump to stop gastrointestinal bleeding. *Ann Emerg Med* 2001;38:328–31.
- 19 Hill SJ, Zarroug AE, Ricketts RR, *et al*. Bedside placement of an aortic occlusion balloon to control a ruptured aorto-esophageal fistula in a small child. *Ann Vasc Surg* 2010;24:822.e7–822.e9.
- 20 Shigesato S, Shimizu T, Kittaka T, *et al*. Intra-aortic balloon occlusion catheter for treating hemorrhagic shock after massive duodenal ulcer bleeding. *Am J Emerg Med* 2015;33:473.e1–473.e2.
- 21 Lonn L, Dias N, Veith Schroeder T, *et al*. Is EVAR the treatment of choice for aortoenteric fistula? *J Cardiovasc Surg* 2010;51:319–27.
- 22 Antoniou GA, Koutsias S, Antoniou SA, *et al*. Outcome after endovascular stent graft repair of aortoenteric fistula: a systematic review. *J Vasc Surg* 2009;49:782–9.
- 23 Goshdas P, Henriksen D, Lynd C, *et al*. Recurrent aortoenteric fistula: case report and review. *Curr Surg* 2005;62:638–43.
- 24 Lozano FS, Muñoz-Bellvis L, San Norberto E, *et al*. Primary aortoduodenal fistula: new case reports and a review of the literature. *J Gastrointest Surg* 2008;12:1561–5.

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