

Two unique cases of primary aortoenteric fistula following a small aneurysm and penetrating ulcer of the abdominal aorta

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ABSTRACT

A primary aortoenteric fistula (PAEF) is a rare condition, and its associated mortality has remained high. We have presented two challenging cases of PAEF related to a small abdominal aortic aneurysm and an abdominal penetrating aortic ulcer. In both cases, a definite diagnosis was established intraoperatively, and the PAEF was repaired with in situ aortic reconstruction. Despite the successful outcome for the first patient, the postoperative complications were fatal for the second patient. The diagnosis of PAEF requires high suspicion and should be a part of the differential diagnosis in every case of gastrointestinal bleeding, especially when a history of, or risk factors for, aortic pathology are present. (*J Vasc Surg Cases Innov Tech* 2022;8:450-4.)

Keywords: Abdominal aorta; Gastrointestinal bleeding; Penetrating aortic ulcer; Primary aortoenteric fistula; Small aneurysm

A primary aortoenteric fistula (PAEF) is an uncommon, albeit lethal, condition.¹ The lack of specific symptoms and the difficulty in depicting the connection between the aortic and enteric lumen has made managing this disease challenging.² In the present report, we have described two cases of this extremely rare condition, which had been induced by a small abdominal aortic aneurysm (AAA) and an abdominal penetrating aortic ulcer (PAU). The first patient provided written informed consent for the report of his case details and imaging studies. The second patient's next of kin provided written informed consent for the report of his case details and imaging studies.

CASE REPORT

Patient 1. A 58-year-old man had presented to the emergency department with a sudden onset of hematochezia and associated coffee-ground emesis. He had a history of hypertension, smoking, and an under-surveillance infrarenal AAA 4.1 cm in diameter according to his last computed tomography angiography (CTA) performed 6 months previously. His systolic blood pressure was 95 mm Hg, and he was tachycardic and

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Author conflict of interest: none.

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The editors and reviewers of this article have no relevant financial relationships to disclose per the Journal policy that requires reviewers to decline review of any manuscript for which they may have a conflict of interest.

2468-4287

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<https://doi.org/10.1016/j.jvscit.2022.06.013>

appeared weak. The physical examination revealed a pulsatile abdominal mass, positive digital rectal examination findings, and coffee-ground content in the nasogastric tube. His new CTA had confirmed an intact AAA, 4.3 cm in diameter. However, the horizontal part of his duodenum had a tangential course to the anterior wall of the AAA, where the enteric and aortic boundaries were indistinguishable. Moreover, at that specific point, an aortic mural thrombus with a distinct concave formation was present (Fig 1). His gastroscopy revealed no pathologic findings. However, before colonoscopy, a new episode of massive hematochezia had led to hypovolemic shock. He was urgently transferred to the operating theater, where the attachment of the horizontal part of the duodenum to the AAA was confirmed (Fig 2, A). Following distal and proximal aortic cross-clamping, further exploration revealed an aortoenteric fistula at the level of the suspected attachment (Fig 2, B). After debridement, aortobifemoral bypass was performed using a silver-impregnated polyethylene terephthalate (PET) graft. After the primary repair of the enteric lesion, omentoplasty was performed to safeguard the graft against a possible enteric leak. On completion of the procedure, the patient was transferred to the intensive care unit. On the third day, diffuse sigmoid colon necrosis had caused deterioration. Therefore, widespread colectomy was successfully performed, and he was discharged after 13 days without any other complications. A 6-week oral antibiotic regimen (ciprofloxacin) was provided. At the 12-month follow-up, the patient remained stable and afebrile with sufficient nutritional status. CTA confirmed a patent aortobifemoral bypass with no signs of infection.

Patient 2. A 70-year-old man had presented with an emergency complaint of nausea and epigastric pain that had been present for the previous 24 hours. He had a history of smoking and misuse of nonsteroidal anti-inflammatory drugs because of persistent lumbar pain for the previous month. The physical

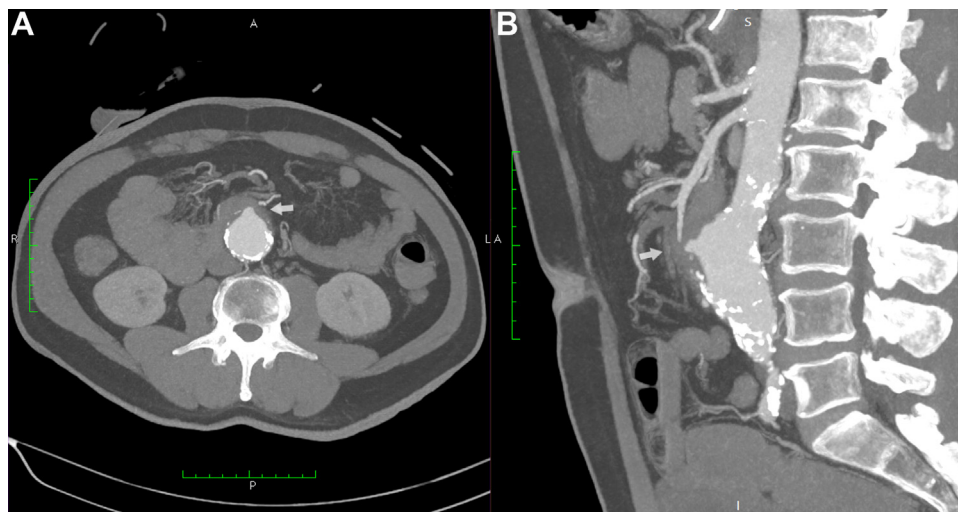


Fig 1. Preoperative computed tomography scan in axial (A) and sagittal (B) planes. *White arrows* indicate the point at which the boundaries between the duodenum and anterior wall of the abdominal aortic aneurysm are indistinguishable. At the same point, the aortic mural thrombus is irregular, with a distinctive concave formation. However, no signs of extravasation were present. Considering that a relatively small aneurysm would be less likely to cause repetitive trauma by virtue of contiguity, an assumption can be made that a focal, penetrating ulceration at the described site infiltrated the fixed retroperitoneal duodenum. Such haziness between the duodenum and anterior wall of the abdominal aortic aneurysm (AAA) observed on a computed tomography angiography (CTA) scan would justify close surveillance even in the case of a small AAA.

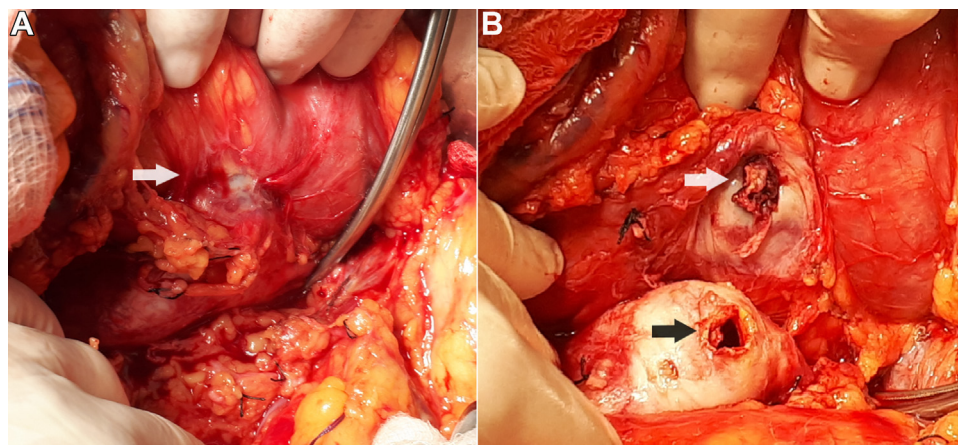


Fig 2. Intraoperative images. **A**, The *white arrow* indicates the strong attachment of the horizontal duodenum to the anterior wall of the aortic aneurysm. **B**, Following proximal and distal aortic control, further exploration revealed the primary aortoenteric fistula. *White and black arrows* indicate the enteric and aortic breach, respectively.

examination revealed diffuse abdominal tenderness and positive digital rectal examination findings. During the assessment, the patient exhibited hematemesis; therefore, intravascular volume restoration and proton pump inhibitor administration were performed. Although hemodynamically stable, he underwent gastroscopy, during which multiple small Forrester IIC ulcers (flat, pigmented hematin on the ulcer base) and a large Forrester III ulcer (a clean-based ulcer without signs of bleeding) were observed in the superior wall of the first part of the duodenum.³ Later, CTA revealed a contained ruptured PAU of the infrarenal abdominal aorta surrounded by multiple air bubbles. The

haziness of the periaortic tissue restricted clear recognition between the aortic wall and ascending duodenum (Fig 3). Given that a PAEF was highly suspected and massive melena had occurred after completion of the CTA, urgent midline laparotomy was performed, which confirmed the intense attachment of the third duodenal part to the infrarenal aorta. The fistula was exposed, and the enteric wall was detached from the aortic wall (Fig 4, A). Following debridement, we performed in situ restoration of the blood flow using a bifurcated silver-impregnated PET graft (Fig 4, B). The graft was fully covered by the retroperitoneal wall, and the enteric breach was primarily repaired in



Fig 3. Preoperative computed tomography scan in the axial plane (A), coronal plane (B), and coronal plane (C) with augmented maximum intensity projection. *White arrows* indicate a contained ruptured penetrating aortic ulcer (PAU) of the infrarenal abdominal aorta and the presence of multiple air bubbles. The haziness of the periaortic tissue restricted clear recognition between the aortic wall and ascending part of the duodenum.

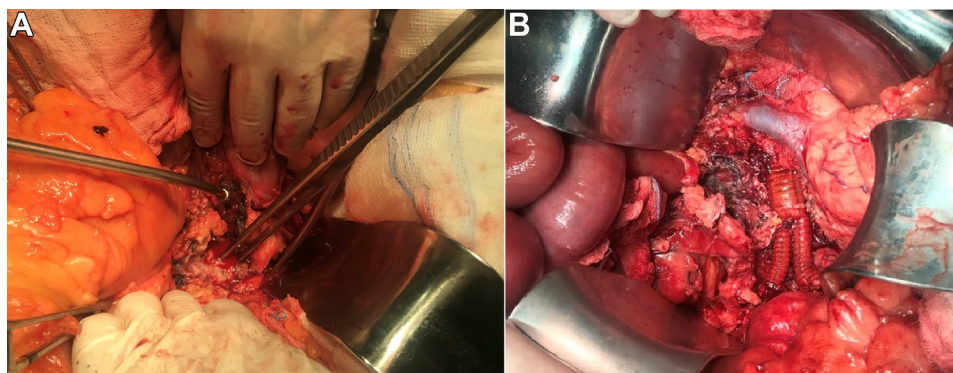


Fig 4. Intraoperative images. **A**, The aortoenteric fistula. Although proximal and distal control had been secured, further exploration revealed the aortic and duodenal breach, indicated by the forceps and irrigation instrument, respectively. **B**, In situ aortic reconstruction. A silver-impregnated polyethylene terephthalate (PET) graft was used for aortobifemoral bypass.

two layers. Subsequently, he remained intubated in the intensive care unit. However, he experienced acute anuric renal failure, which necessitated continuous renal replacement therapy. On the third postoperative day, widespread necrosis of the descending and sigmoid colon was observed. Despite a prompt extended right colectomy, the patient's status deteriorated. However, after 8 days of hospitalization, he had died of sepsis and multiorgan failure.

DISCUSSION

A PAEF is a rare clinical condition. The incidence has ranged from 0.04% to 0.07%.⁴ It is more common among men, and 80% of the cases have been associated with an AAA and with involvement of the ascending part of the duodenum.⁵ Mycotic aneurysms, radiation therapy, cancer, and the ingestion of a foreign body are exceptional causes of PAEF.^{6,7} When a PAEF is associated with an underlying AAA, the mean diameter of the aneurysm has been 6.2 to 7 cm, which correlates with the theory that the pathogenesis of PAEF is repetitive trauma to

the posterior duodenal wall induced by the aortic pulses of the large diameter aorta.^{1,8} However, in the first case, even the presence of a 4.3-cm AAA was capable of causing a PAEF, making our case unique, to the best of our knowledge, across the English literature. In a review of the CTA images from the first patient, one could assume that the etiology of the PAEF was not the AAA but a focal, penetrating ulceration at the site of the irregular mural thrombus, which had infiltrated the fixed retroperitoneal duodenum.⁹ Although the first patient had had a history of hematochezia and a pulsatile mass was present, the rarity of the PAEF, the inconclusive CTA findings, and the small size of the associated aneurysm, moderated our suspicion of a fistula. For the second patient, a history of nonsteroidal anti-inflammatory drug misuse and the positive findings of an ulcer during endoscopy would have moved the diagnosis away from PAEF if CTA had not been performed. Although various studies have reported on thoracic PAUs associated with a primary aorto-esophageal fistula, our second case was

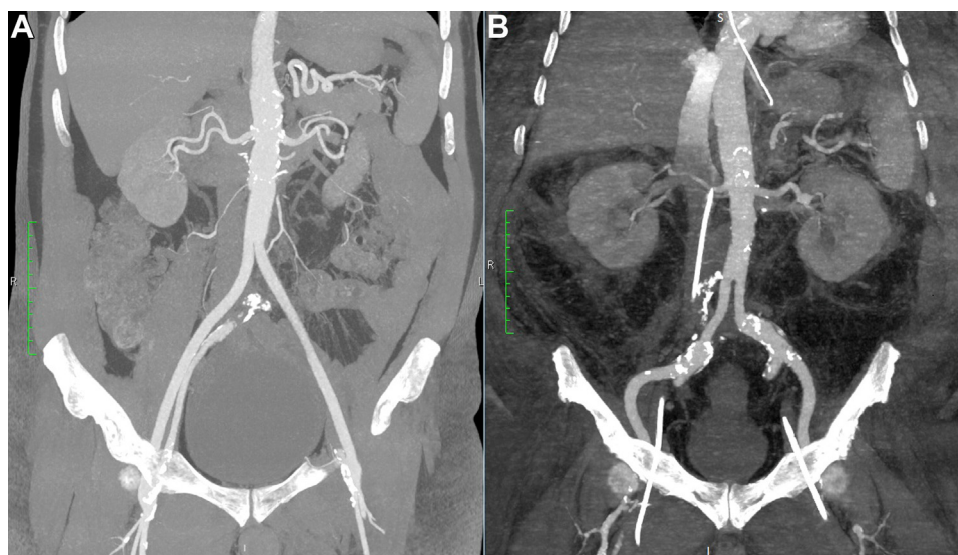


Fig 5. Postoperative computed tomography scans. **A**, Patient 1. At 12 months of follow-up, the aortobifemoral bypass was patent with no signs of infection. Also, the right common iliac artery had retrograde flow but the left common and external iliac arteries have become thrombosed. **B**, Patient 2. The aortobi-iliac bypass is patent; however, the flow to the distal part of the left internal iliac artery was abruptly interrupted.

remarkable because, to the best of our knowledge, it is the first report of an abdominal PAU causing a PAEF in the English literature.¹⁰⁻¹³ Vujcic et al¹⁴ recently reported a case of an aorto-appendicular fistula that might have been related to an abdominal PAU. However, the investigators suggested that undetected chronic appendicitis was the actual cause of the degradation of the aortic wall.¹⁴ In our patient, a focal penetrating ulcerative lesion of the infrarenal abdominal aorta had evolved from an atherosclerotic plaque to a contained rupture, inducing the development of a PAEF.

Aortic rupture into the alimentary tract is frequently self-limiting, causing herald bleeding. Thrombus configuration and associated vasoconstriction will contribute to temporary patient stabilization; however, such stabilization can make it challenging to identify the fistula.¹⁵ CTA techniques such as digital subtraction angiography will usually fail to depict active extravasation because of the provisional thrombus formation.¹⁶ Occasionally, such as in both of our patients, a new incidence of massive bleeding will cause rapid deterioration, and, eventually, open surgery will allow for the definite diagnosis.¹⁷ The decision to proceed with a major procedure without a certain diagnosis is challenging.

Owing to the rarity of PAEF, no consensus has been reached regarding the best treatment. In addition, the available data were derived from case reports. The treatment of PAEF requires hemorrhage control, followed by distal reperfusion and repair of the enteric lesion.¹⁸ Potentially contaminated areas, owing to the PAEF, will jeopardize the integrity of the revascularization procedure and repair of the fragile enteric tract. In both of our

patients, following attentive exposure, minor purulence had occurred, which justified the in situ repair. Otherwise, aortic ligation and extra-anatomic ligation have been recommended in the case of periaortic contamination.⁹ The use of a bifurcated silver-impregnated PET graft prevented infection recurrence without significantly prolonging the procedure.¹⁹ However, the great burden of the aortoiliac occlusive disease contributed to the postoperative thrombosis of the left interior iliac artery in both of our patients and had partly induced the ischemic colitis (Fig 5). We selected primary repair of the enteric lesion, although no consensus has been reached regarding the optimal type of enteric repair for cases of PAEF.²⁰ In addition, treatment of PAEF has been associated with a high mortality rate, reaching $\leq 30\%$ for in situ repair and 40% for extra-anatomic bypass.⁸

CONCLUSIONS

When a history of, or risk factors for, aortic pathology are present, PAEF should be a part of the differential diagnosis of gastrointestinal bleeding. High suspicion, continuous reevaluation, and multidisciplinary collaboration are crucial for achieving optimal outcomes.

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Submitted Sep 18, 2021; accepted Jun 19, 2022.