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Primary aortoesophageal fistula: a rare cause of acute upper gastrointestinal bleeding

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ABSTRACT

Acute upper gastrointestinal bleeding is a potentially life-threatening emergency, especially in the elderly. This condition accounts for approximately 1% of all emergency room admissions. Among the causes of such bleeding is aortoesophageal fistula, a dreaded but apparently rare condition, first recognized in 1818. The great majority of cases are of primary aortoesophageal fistula, caused by atheromatous aortic aneurysms or, less frequently, by penetrating aortic ulcer. The clinical presentation of aortoesophageal fistula is typically characterized by the so-called Chiari's triad, consisting of thoracic pain followed by herald bleeding, a variable, short symptom-free interval, and fatal exsanguinating hemorrhage. The prognosis is poor, the in-hospital mortality rate being 60%. Conservative treatment does not prolong survival, and the in-hospital mortality rate is 40% for patients submitted to conventional surgical treatment. Here, we report the case of a 93-year-old woman who presented to the emergency room with a history of hematemesis. The patient was first submitted to upper gastrointestinal endoscopy, the findings of which were suggestive of aortoesophageal fistula. The diagnosis was confirmed by multidetector computed tomography of the chest. Surgery was indicated. However, on the way to the operating room, the patient presented with massive bleeding and went into cardiac arrest, which resulted in her death.

Keywords: Aortic aneurysm; Atherosclerosis; Esophageal fistula; Gastrointestinal hemorrhage.

INTRODUCTION

Acute upper gastrointestinal bleeding (UGIB) is a life-threatening emergency situation that requires hospitalization. The incidence of acute UGIB ranges from 37 to 172/100 000 population, and the condition is responsible for approximately 1% of all emergency room admissions.[1,2] In a study conducted in Scotland, Blatchford et al.[3] showed that the incidence of acute UGIB rises sharply with advancing age, being almost 6 times higher among individuals over 75 years of age than among those in the 15-29 year age bracket,

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as has also been reported by other researchers.[4-6] Various studies have shown that the predominant cause of acute UGIB is peptic ulcer (duodenal or gastric), followed by varices, esophagitis, esophageal ulcer, gastritis, duodenitis, Mallory-Weiss syndrome, and malignancy.[1] Population-based studies of all-cause acute UGIB have shown that mortality ranges from 3% to 14% and increases markedly in parallel with increasing age. Blatchford et al.[3] also showed that the mortality rate for acute UGIB was 100 times greater among individuals over 75 years of age than among those in the 15-29 year age bracket. There are at least two situations in which acute UGIB becomes life-threatening: abnormal communication between the aorta and the esophagus; and abnormal communication between the aorta and the gastrointestinal tract. These are represented by aortoesophageal fistula (AEF) and aortoenteric fistula, respectively.[6] These uncommon causes of acute UGIB are typically identified post-mortem after an exsanguinating hemorrhage.[7] The first report of AEF was in 1818 by Dubrueil, who described the case of a soldier dying from massive upper gastrointestinal hemorrhage 5 days after esophageal impaction of a bone fragment, which had penetrated the thoracic aorta.[7,8] Since then, numerous cases of AEF, attributed to various causes, have been reported in literature.

When AEF occurs in patients with no history of thoracic surgery, it is classified as primary AEF, which accounts for 95% of all cases, whereas it is classified as secondary AEF, which accounts for only 5% of all cases, when it occurs after thoracic aortic or esophageal surgery. Chart 1 shows the causes of AEF documented in literature.

Chart 1 – Etiology of AEF[a]

Primary AEF	Secondary AEF
(95%)	(5%)
Ruptured thoracic aneurysm (atherosclerotic, dissecting, mycotic, syphilitic)	Repair of thoracic aortic aneurism (primary, allograft, prosthetic)
Penetrating aortic ulcers	Patent ductus arteriosus ligation
Malignant thoracic neoplasms (esophageal, bronchial)	Repair of coarctation of aorta
Esophageal foreign bodies	Esophageal surgery
Corrosive ingestion	Endovascular aortic stent-grafting
Benign esophageal ulcer	Esophageal instrumentation
Barrett's ulcer	
Infections (tuberculosis, mediastinal abscesses)	
Prolonged nasogastric intubation	
Other (blunt or penetrating thoracic trauma, radiotherapy)	

[a] Compiled from various sources[6,7,9,10].

CASE REPORT

A 93-year-old female patient sought treatment in the emergency room of the University Hospital of the University of São Paulo, presenting with acute upper gastrointestinal bleeding. She had a clinical history of hypertension and cardiomyopathy with chronic atrial fibrillation. She reported dyspnea on mild exertion, together with chronic leg edema. She had been under treatment with carvedilol, captopril, furosemide, and spironolactone for the last 2 years. She complained of epigastric and substernal pain followed by evacuation resembling melena for the last few days before admission. Her first upper gastrointestinal bleeding episode was characterized by the emission of a small amount of blood mixed with saliva from her mouth while sleeping, as noted by her family. Because the bleeding was minimal, she was taken to a primary care facility, where she remained under clinical observation after being started on vitamin K, omeprazole, and intravenous saline infusion. On the next day, she began to bleed again and was referred to our hospital for endoscopic examination. At admission, she was conscious and lucid, although emaciated and pale. There were traces of blood in the oral cavity. Her blood pressure was 95/69 mmHg, and her pulse was 100 bpm. Pulmonary examination revealed decreased breath sounds at the right lung base. The abdomen was slightly painful on gentle palpation of the epigastrium. Rectal examination failed to reveal melena. The laboratory test results are shown in Table 1.

Upper gastrointestinal endoscopy revealed pulsatile extrinsic compression of the esophagus at 25 cm from the superior dental arch (Figure 1). At the most prominent area of the mass, we observed a small ulceration of the mucosa with whitish material, interpreted as probable atheroma, emerging into the esophageal lumen (Figure 2). Because of these findings, the examination was promptly interrupted.

The patient was submitted to multidetector computed tomography (CT) of the chest. The scan showed an elongated, tortuous, atheromatous

Table 1 – Laboratory test results

Variable	Result	Reference value
Hemoglobin (g.dL^{-1})	11.3	12.3-15.3
Hematocrit (%)	35.7	360-45.0
Leukocytes (mm^3)	8900	4.4-11.3 × 10^3
Platelets (mm^3)	281 000	150-400 × 10^3
Prothrombin time (INR)	1.58	1
AST (IU.L^{-1})	45	10-35
ALT (IU.L^{-1})	44	9-43
Amylase (IU.L^{-1})	48	20-104
Total bilirubin (mg.dL^{-1})	0.6	0.3-1.2
K (mEq.L^{-1})	4.2	3.5-5
Creatinine (mg.dL^{-1})	4.6	0.4-1.3
BUN (mg.dL^{-1})	70	10-50
Glucose (mg.dL^{-1})	111	70-99

INR, international normalized ratio; AST, aspartate aminotransferase; ALT, alanine aminotransferase; K, potassium; BUN, blood urea nitrogen.

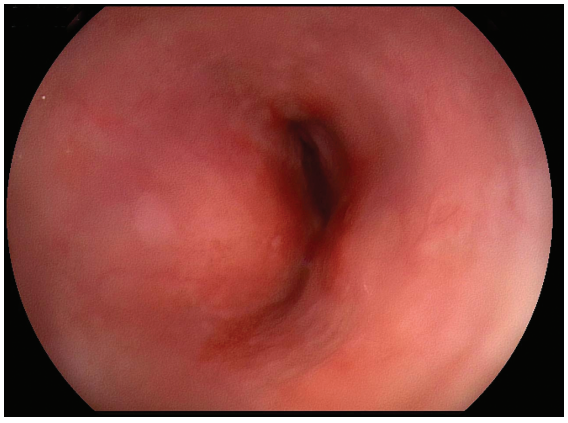

Figure 1 – Upper gastrointestinal endoscopy showing extrinsic compression of the esophagus at 25 cm from the superior dental arch.

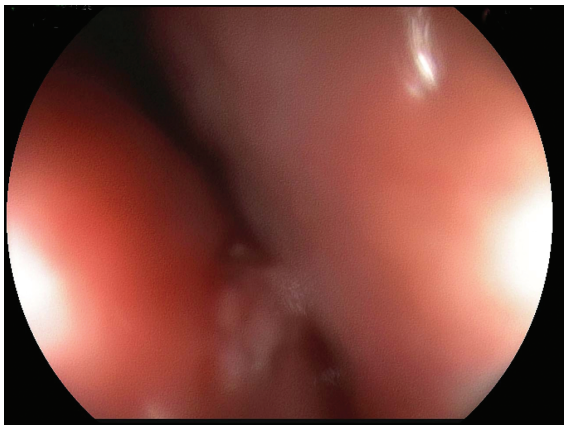

Figure 2 – Upper gastrointestinal endoscopy showing extrinsic esophageal compression with exteriorization of a whitish fibrinoid material through the fistula.

aorta, with wall thickening and calcified plaques. The descending aorta was irregular and fusiform, with an average diameter of 46 mm. Multiple parietal ulcers were seen on the descending aorta. A fistula, measuring 15 mm in diameter, was observed in the proximal descending aorta, with active contrast extravasation generating a hematoma of 60 × 46 mm, which compressed the esophagus (Figures 3A and B).

A massive thrombus was observed on the descending thoracic aorta extending inferiorly to the thoracoabdominal junction (Figures 4, 5 and 6).

Considering the severity of the illness, the advanced age of the patient, the presence of multiple comorbidities, and the relative hemodynamic stability, she was immediately referred to a tertiary hospital for surgical treatment. However, extensive rebleeding and cardiac arrest resulted in her death prior to surgery.

DISCUSSION

Thoracic aortic aneurysms constitute the most common cause of AEF. In 1978, Carter et al.[11] reported 24 cases of AEF,16 (66%) of which were caused by aortic aneurysms, most due to arteriosclerosis. In 1991, Hollander & Quick showed that the cause of AEF is thoracic aneurysm in 54% of cases, esophageal foreign body in 19%, and malignancy in 17%.[12] An additional cause of AEF is penetrating aortic ulcer (PAU).[13] First described in 1934 by Shennan,[14] PAUs are among the least common causes of acute aortic syndrome. Resulting from the ulceration of a previous atherosclerotic plaque, PAUs penetrate the aortic wall from the internal elastic lamina to the arterial media. The hematoma is initially contained by the tunica adventitia leading to a false aneurysm formation.[15] In elderly patients with multiple comorbidities, PAUs are common, high blood pressure being the most frequent risk factor.[15] The patient described in the present case report had fusiform dilatation of the descending aorta with multiple atheromatous plaques. The ulceration and rupture of one of those plaques may have initiated the formation of the AEF. Approximately 10% of thoracic aneurysms break into the esophagus, which is the third leading site of rupture, after the pericardium and left pleura.[12] Another possible explanation for the AEF reported here is the development of a PAU, which was located in the distal aortic arch, as is typical. Some authors have reported that PAUs have a benign clinical course and present a low risk of progression. However, aortic rupture and other serious complications have also

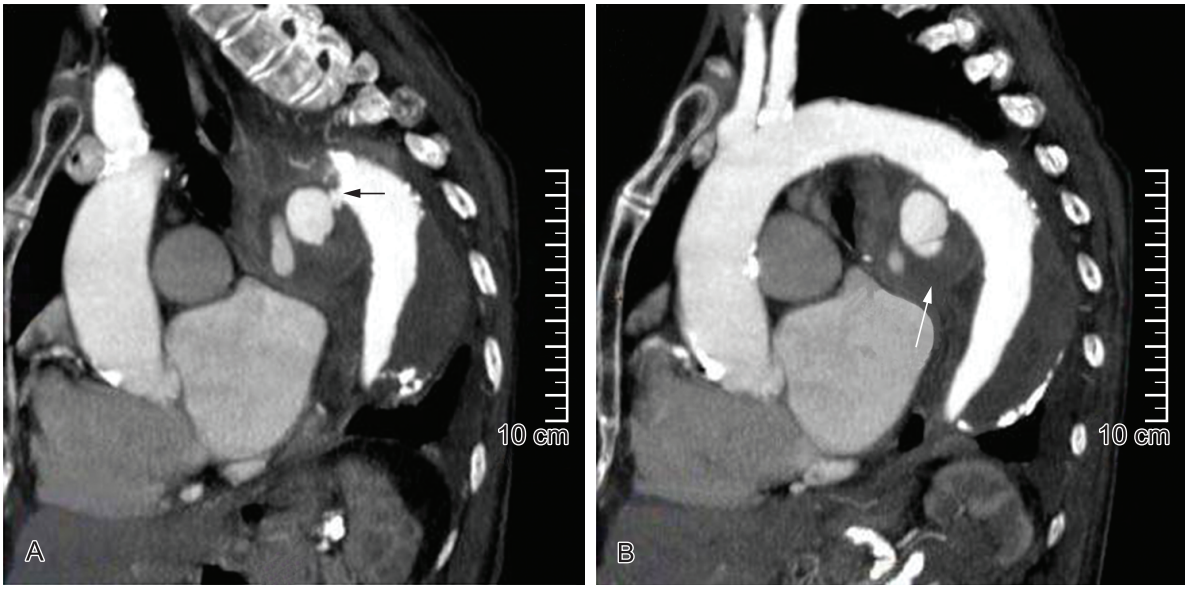

Figure 3 – Multidetector CT angiography. Sagittal reformatted images through the thorax showing active aortic bleeding generating an anterior hematoma (black arrow in **A**), which is compressing the posterior wall of the esophagus (white arrow in **B**).

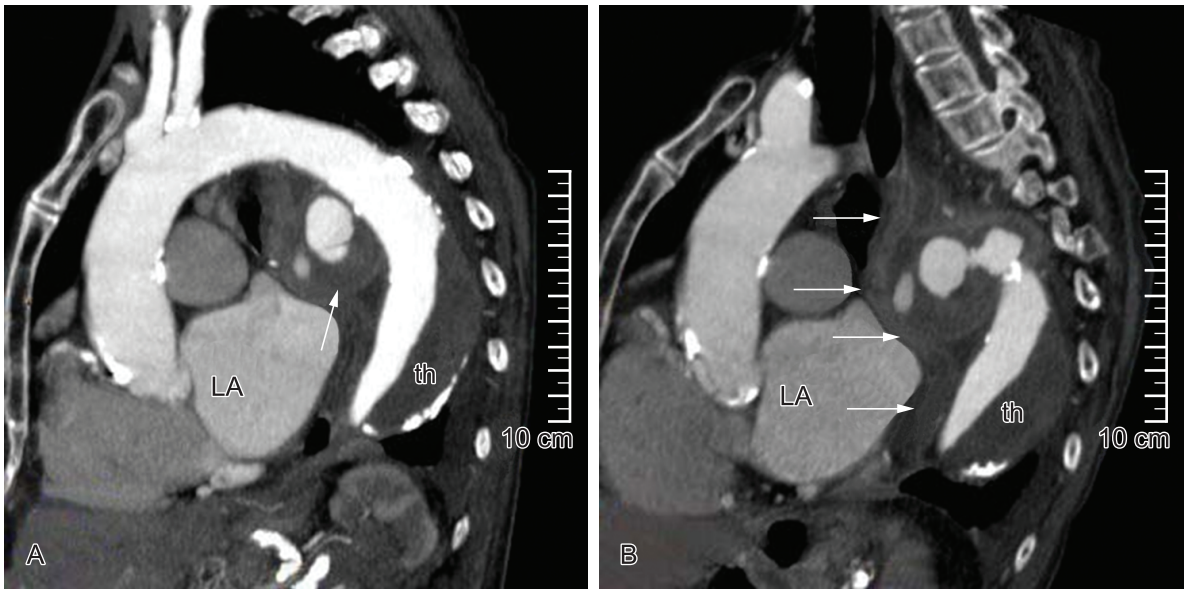

Figure 4 – Multidetector CT angiography. Sagittal reformatted images through the thorax showing an anterior hematoma (in **A**, arrow), which is compressing the posterior wall of the esophagus (better observed in **B**, arrows) and a mural thrombus (th) into the descending aorta. Note the enlarged left atrium (LA).

been reported.[16,17] Formation of an AEF occurs at the descending thoracic aorta, where the aorta and the esophagus are in direct contact. Erosion of the esophagus by mechanical compression and ischemia results in infectious destruction of the aneurysm wall and consequently in the creation of a fistula.[12] In the case reported here, it is possible that the rupture of the ulcer led to the formation of a pseudoaneurysm that created the AEF.

Chiari described the typical syndrome of AEF as a triad comprising central chest pain or dysphagia, a short symptom-free interval, followed by a sentinel hemorrhage, another symptom-free interval of hours or days, and finally fatal exsanguinating hemorrhage.[18] In rare cases, there are recurrent, large, self-limiting hemorrhages. In the study conducted by Carter et al.[11], 80% of the patients evaluated had a history of sentinel bleeding, as did 75% of the patients evaluated by Pipinos & Reddy.[19] In a review of 500 cases of AEF, it was found that 59% of all patients had central chest pain and 45% had dysphagia.[12] A 30-year review comprising all reported cases of AEF revealed that only 45% of patients meet all 3 criteria of Chiari's

triad,[20] as did the case reported here. Central chest pain can be caused by distention, erosion, or localized dissection of the aortic wall, as well as by esophageal perforation with mediastinitis or tumor invasion of the aorta or pleura. The bleeding might halt temporarily because of hypotension, spasm of the aortic wall, or pressure from a periaortic hematoma. After the dissolution of the hematoma or once the blood pressure returns to normal, the patient rebleeds.[7,9]

The suspicion of AEF should arise when there is a fresh, red, upper gastrointestinal hemorrhage in a patient diagnosed with thoracic aorta aneurysm or thoracic aorta dissection, as well as in those with a history of esophageal surgery, aortic surgery, or even foreign body ingestion. Chiari's triad is usually diagnostic.[7] Another situation in which AEF should be suspected is that in which there is a massive hematemesis but only minimal blood in the stomach and no obvious source of bleeding on subsequent endoscopy. Endoscopy can also reveal a pulsatile submucosal mass, with or without clots; bluish grey mucosa due to submucosal dissection by blood or intramural hematoma; foreign bodies; or, in rare cases, an opening fistula. Chest X-ray findings of mediastinal widening, tortuous aorta, or calcifications can also raise the suspicion of AEF.[11,21,22]

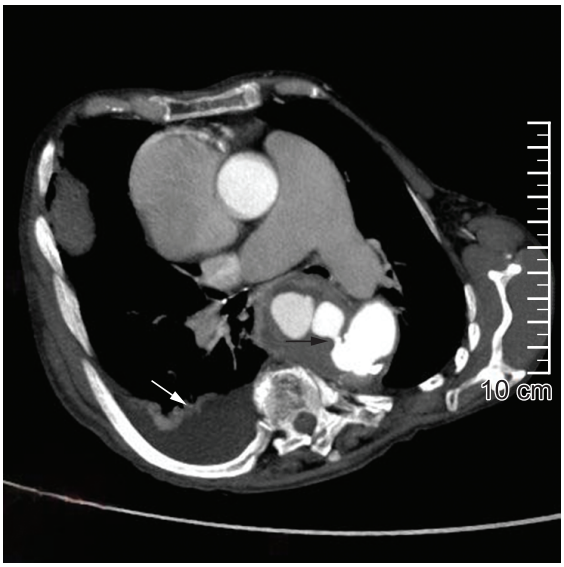

Figure 5 – Multidetector CT angiography. Axial image through the thorax showing active bleeding generating an anterior hematoma (black arrow) and mild right pleural effusion (white arrow).

In cases of AEF, CT angiography of the aorta will show the thoracic aneurysm, as well as its relationship with the esophagus and surrounding structures, as in the case reported here. The fistulous communication is rarely identifiable.[23] Because many patients have abdominal aneurysms, an abdominal aortogram should also be obtained.[9] A CT scan can also show periaortic hematoma, foreign body, neoplasia, mediastinal abscess, adherence of the esophagus to the aorta, or the presence of gas in the aneurysmal sac or around an aortic prosthesis.[24,25] Barium esophagogram is contraindicated in the presence of bleeding; when not so contraindicated, it can show extrinsic compression and deviation, usually anterior and to the right, because most AEFs arise from an aneurysm of the descending aorta.[7] Naschitz et al.[26] described a clinical and radiological triad that is suggestive of AEF: upper gastrointestinal bleeding; aneurysmal dilatation of the aorta; and a filling defect on esophagogram.

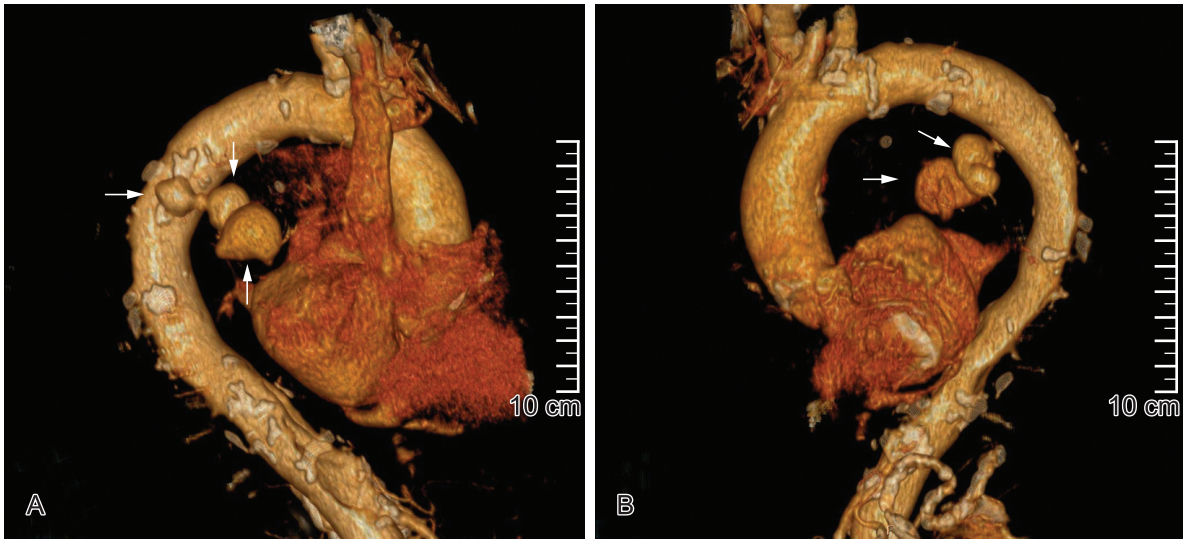

Figure 6 – Three-dimensional volumetric reconstruction showing a descending thoracic aortic pseudoaneurysm (arrows) on the anterior aortic wall, and multiple atheromatous plaques. (**A**, right oblique anterior view; **B**, left oblique posterior view).

The prognosis of AEF is extremely poor. As of 1997, only six survivors had been reported.[9] However, survival rates have increased due to improvements in intensive care facilities and greater awareness of the condition. As of 2005, 55 cases of AEF survival had been reported.[27-29] Two different surgical techniques have been employed in the treatment of AEF: management of aortic rupture, together with immediate esophageal repair, which consists of direct suture or esophageal resection with esophagogastroplasty or coloplasty; and delayed esophageal repair. Although surgery continues to be the standard treatment for AEF, survival after aortic stent-graft implantation has been reported at various centers.[30] However, aortic stent-grafting does not repair the esophagus, which means that there is still a substantial risk of mediastinitis, as well as of stent-graft infection. Marone et al.[13] reported two cases that were successfully treated with aortic endovascular grafting followed by surgical repair of the esophageal lesion.

We conclude that AEF is a rare and usually fatal condition. With the increase in life expectancy and therefore in atherosclerotic complications, as well as the increasing numbers of thoracic aortic procedures being carried out worldwide, the incidence of AEF is likely to increase. Early recognition, a high index of suspicion, improved clinical care, and better emergency services could increase the number of cases recognized prior to fatal hemorrhage and consequently increase the number of survivors.

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