

A Rare Cause of Haematemesis; a Primary Aorto-Esophageal Fistula: Case Report and Review of Literature

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Abstract

Aorto-Esophageal fistula is a rare condition, causes excessive bleeding from the upper gastrointestinal tract and is associated with a high mortality. This case report demonstrates the presentation of a 63-year old female with a primary aorto-esophageal fistula, due to a ruptured thoracic aneurysm. She first presented with a sentinel haemorrhage, followed by a new bleeding several hours later. An earlier performed upper gastrointestinal endoscopy elsewhere, because of dysphasia, already mentioned an impression from a non pulsing compressive swelling. No further analysis was performed. At current presentation, she was hemodynamically instable, with mild haematemesis. After stabilising she was admitted to the Intensive Care unit; however, haematemesis worsened. An immediate Computed Tomography (CT) scan showed an Aorto-Esophageal Fistula (AEF), due to a ruptured thoracic aortic aneurysm. Emergent surgical treatment consisted of endovascular stent-graft of the thoracic aorta and a total thoracic esophagectomy combined with immediate esophago-gastrostomy. Unfortunately, the subsequent multi-organ failure was fatal. A critical view to the surgical approach is given, combined with a review of diagnostic and therapeutic options in aorto-esophageal fistulae.

Keywords: Aortic aneurysm thoracic; Aortic rupture; Esophageal Fistula; Endovascular Procedure; Aortoesophageal Fistula; Haematemesis

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Introduction

An Aorto-Esophageal Fistula (AEF) often has a lethal outcome, despite advanced techniques that have improved treatment. It is a rare condition which frequently does not present with classical symptoms, like Chiari's triad [1, 2] (dysphasia or midthoracic pain, a sentinel haemorrhage, and after a period without symptoms, a massive, often lethal, haemorrhage) in most cases. It requires immediate treatment. Therefore, it is important to be aware of the possibility of an AEF in a patient with haematemesis. Upper gastro-intestinal bleeding accounts for approximately 1% of all emergency room admissions [2]. A primary Aorto-Esophageal Fistula (AEF) is a very rare cause of

upper gastrointestinal bleeding (only 3.5%) [3]. Primary AEF account for 95% of all cases. The other 5% (secondary AEF) occur as a complication of thoracic surgery [2].

Most primary AEF are caused by an atherosclerotic aneurysm of the thoracic aorta (51-75%) [4, 5]. Other possible causes of AEF are shown in table 1.

Table1. Etiology of AEF [4, 7-10]

Primary AEF (95%)	Secondary AEF (5%)
Ruptured (atherosclerotic) thoracic aneurysm (51-75%)	Following thoracic/esophageal surgery
Benign esophageal ulcer	Pseudoaneurysm at the vascular anastomosis
Barrett's ulcer	Mechanical erosion of a stent of graft (after several years)
Aortic ulcer (penetrating)	
Corrosive ingestion	
Malignant neoplasms (esophagus, bronchus)	
Esophageal corpus alienum	
Infections (abscesses, tuberculosis*, syphilis*, bacterial, Takayasu's arteritis), Q-fever	
Radiotherapy	
Prolonged nasogastric intubation	
Trauma (penetrating thoracic trauma)	

*most cases before 1970

The classic triad of symptoms of an aorto-esophageal fistula (Chiari's triad) consists of dysphasia or midthoracic pain, in 80% of cases followed by a sentinel haemorrhage, and after a period without symptoms, a massive, often lethal, haemorrhage [2, 6]. Most likely, the sentinel bleeding stops by formation of a blood clot, hypotension, and arterial spasms. This clot is weakened by corrosion (caused by gastrointestinal contents) or bacterial infection, which will eventually lead to further haemorrhage [6]. However, this classical triad is seen in only 45% of patients [7]. Most cases present with gastrointestinal bleeding, with or without haematemesis. 45% of patients have dysphasia, 59% report a central chest pain [3]. This pain is probably due to distention or dissection of the aortic wall, or to mediastinitis by esophageal leakage [3, 8]. We present a case of

a classical presentation of AEF followed by a review of possible presentations, etiology, diagnostic and therapeutic options.

Case Report

A 63-year-old woman presented at our emergency room with haematemesis. She mentioned general malaise for several days, with retrosternal pressure and pain. She felt neither dyspnoea nor palpitations. Her stool analysis had shown no abnormalities. A few hours after visiting her GP she collapsed at home, and threw up fresh blood. At presentation she felt nauseous, and had mild haematemesis. She had not used any medication, used to smoke several cigarettes a day and drank a bottle of brandy a week. On examination, we saw a pale, cold, diaphoretic patient, who was less alert, though well oriented. Her blood pressure was 80/46 mmHg, heart rate 146 beats per minute (bpm),

oxygen saturation 96% (with a non-rebreather mask (100% O₂). She was in respiratory distress, with rhonchi over the left lung. The abdomen showed tenderness in epigastrium, normal bowel sounds, without distention or hepatosplenomegaly. Laboratory examination revealed a hematocrit of 0.23 L/L, Haemoglobin (Hb) 5.0 mmol/L (reference range 7.5-10mmol/L; earlier that day at her GP the Hb was 8.9 mmol/L), mean corpuscular volume 104 fL, platelet count 120/nL (150-400/nL). The white blood cell count was 10.1/nL (4-10/nL), glucose 11.0 mmol/L (<7.8mmol/L). Bilirubine was 18.0 µmol/L (<17 µmol/L), ASAT 82 U/L (<30 U/L), ALAT 50 U/L (<35 U/L), alkaline phosphatase 151 U/L (40-120 U/L), gamma GT 530 U/L (<40 U/L), and LDH 190 U/L (<250 U/L). Renal function and electrolyte tests were normal, albumin 25 g/L (35-50 g/L), CRP was 240 mg/L(<6 mg/L). Arterial blood sample (taken while on a non-rebreather mask): pH 7.41 (7.35-7.45), pCO₂ 31 mmHg (32-45mmHg), standardized bicarbonate 20.8 mmol/L (22-28 mmol/L), bicarbonate 19.2 mmol/L, pO₂ 120 mmHg, saturation 98%, lactate 5.7 mmol/L. An electrocardiogram revealed sinus tachycardia at a rate of 145 bpm, but was otherwise normal. Radiographs of the chest showed no abnormalities (Figure 1).



Figure 1 – Chest-X-ray

An earlier performed upper gastrointestinal endoscopy elsewhere 42 days before presentation, because of dysphasia, mentioned a mild gastritis and an impression from a smoothly

lined, non pulsing, compressive swelling, which was pressing on the distal esophagus from the outside. After saline 0.9% infusion her blood pressure rose to 117/81mmHg, with normalisation of pulse rate. We started erythrocyte transfusion and intravenous proton pump inhibition and admitted her to the Intensive Care Unit (ICU). An upper gastrointestinal endoscopy was performed, which demonstrated a sub mucosal tumour with normally overlying mucosa in the mid-esophagus (25-30cm from dental arch) (Figure 2a). Distally, the esophagus appeared ulcerated, necrotic, and thorn, with an adherent blood clot. A perforation could not be ruled out. The stomach showed no active bleeding, although blood was seen. A tri-luminal tube was placed.

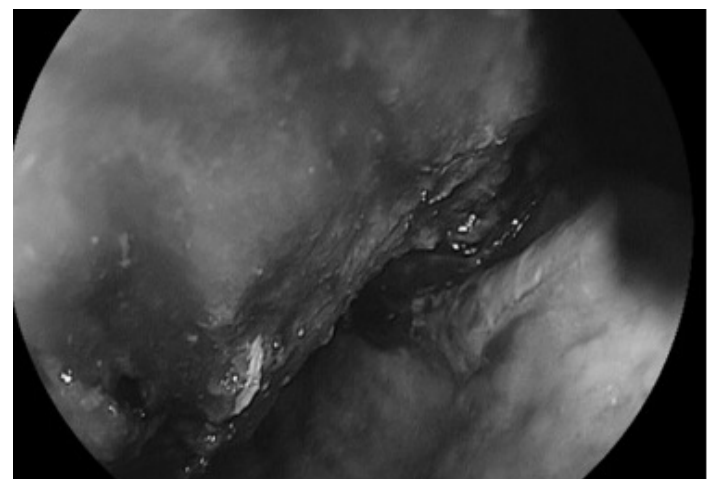


Figure 2 – View at 30 cm in the esophagus during the first gastroscopy: ulceration and necrosis with adherent clot, but no active bleeding.

A Computed Tomography (CT) of the thorax was scheduled for the next morning, after further stabilisation of the patient at the Intensive Care Unit (ICU). However, seven hours later, fresh blood was aspirated from the tube and she had haematemesis again. An immediate gastroscopy demonstrated the same image of a sub mucosal tumour, without an active bleeding site. CT scan of the thorax showed a thoracic aortic aneurysm, perforating the distal esophagus, with a maximal diameter of 99 mm (Figure 3a and 3b).

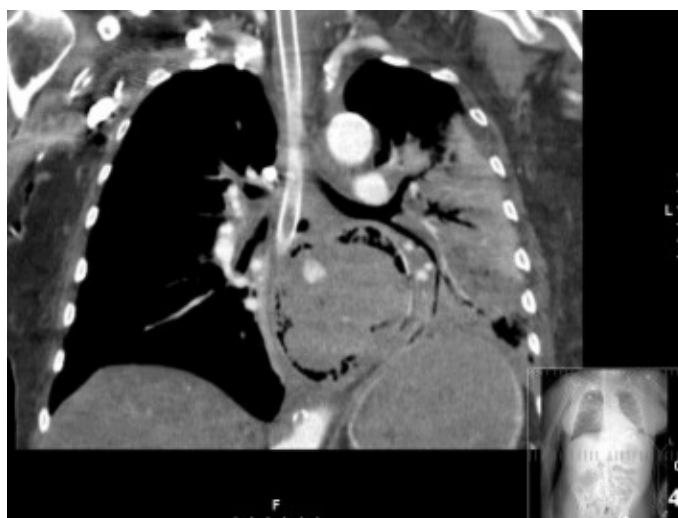


Figure 3a – CT-thorax, demonstrating air in the ruptured thoracic aorta, a pathognomonic sign of an AEF.



Figure 3b – Coronal reconstruction of thoracic CT-scan, demonstrating the ruptured thoracic aneurysm in proximity of the esophagus, containing air and pleural effusion.

The patient was diagnosed with an Aorto-Esophageal Fistula (AEF), due to a ruptured thoracic aortic aneurysm. Emergency surgery was scheduled, after intubation and vasoactive medication. An open procedure to repair the thoracic aorta was considered too hazardous, therefore an endovascular stent-graft procedure was chosen. Unfortunately, an abdominal approach with a PTFE (polytetrafluoroethylene)-conduit was necessary,

because of a highly calcified infrarenal aorta and severe stenosis of the iliac-femoral tract. To seal the ruptured thoracic aneurysm, two C-TEG stents (diameter of 31mm, length of 150mm and 100mm) were deployed, with 50mm overlap. The celiac trunk remained patent by this procedure. This procedure went well, and the patient's condition seemed good enough to perform immediate esophageal reconstruction. A total thoracic esophagectomy and esophago-gastrostomy with a cervical anastomosis after gastric tube reconstruction were performed. The patient was readmitted to the ICU. Unless all efforts and supportive care she deteriorated, without signs of rebleeding. The next day she died of multi-organ failure.

The histologic examination of the removed esophagus and upper part of the stomach demonstrated an oval mucosal defect of 4x3.5 cm in the esophagus. Microscopic examination demonstrated a mild chronic inflammation. No clues were found suggesting an underlying infectious disease. Autopsy showed generalised atherosclerosis and concentric hypertrophy of the left ventricle, signs of a leakage of the aorta thoracalis, with a residual haematoma of 200 cc, the stent-grafts were in situ without signs of leakage. The cervical remnant of the esophagus showed transmural necrosis without anastomotic leakage. No signs of an acute myocardial infarction, pulmonary embolism or signs of septic shock were found.

Discussion

In our patient a one-step approach for surgery was chosen. The main reason for immediate repair was that a laparotomy was already performed, as well as to prevent the esophagus to become a source of postoperative infections. It should be discussed whether this was the right decision, or if a two-step approach would have led to a better outcome. Also, our patient's diagnose might have been delayed.

Possible features to diagnose an AEF are chest X-ray, endoscopy and Computed Tomography. On a postero-anterior and oblique chest x-ray, widening of the mediastinum, a tortuous aorta, and calcification could be seen. In our patient

chest X-ray was not helpful and may even have delayed diagnosis.

At endoscopy, a sub mucosal mass (as in our case) with or without adherent blood clots, a grey mucosa (due to dissection by blood or haematoma), a foreign body, ulcer, and rarely a fistula can be seen [9].

Computed tomography is valuable to demonstrate a thoracic aneurysm, including its relation to other structures. Haematomas and/or abscesses can be visualized as well. Air within the aortic wall or lumen is pathognomic for AEF; other findings may be focal thickening of the bowel wall or disruption of the aortic fat cover, and extravasation of contrast into the bowel [10, 11]. Angiography has the same sensitivity as CT scan, but is more invasive. Laboratory tests are not informative about the underlying cause. Diagnosis is difficult to establish, because bleeding may be intermittent; diagnostic tests hardly ever reveal the fistulous communication [2, 9]. Failure to diagnose the condition often leads to death.

Retrospectively, the first doctor's delay had occurred several weeks earlier, when a gastroscopy performed for non-specific thoracic showed a sub mucosal non-pulsing tumor, which was not further examined. When a CT scan was performed at that time, maybe this lethal bleeding could have been prevented with elective surgery or stenting of the aorta. The second doctors delay occurred at admission of the patient. Considered diagnoses were ruptured leiomyoma, ulcer or malignant process perforating the esophagus, or a spontaneous mucosal or transluminal esophagus rupture. AEF was not thought of immediately. An immediate CTA might have altered the outcome.

Therapy

With the introduction of endovascular techniques open surgical approach of an AEF is less frequently applied. Two different surgical techniques may be applied for the treatment of an AEF. In our patient, we used endovascular repair, combined with immediate esophago-gastrostomy (one step approach). Another

possibility is a two-step approach. First the bleeding is controlled with endovascular repair, as a bridge to the second procedure. Later, if the patient is hemodynamically stable, debridement of the thoracic and mediastinal tissues (with removal of diseased aorta and esophagus) is performed, followed by esophageal repair [12]. This two-step approach may theoretically lead to an increased risk of mediastinitis [13]. It remains unclear which approach is favourable. The staged approach is in some studies associated with better outcomes [12, 14], although the number of cases is very small. A difficult problem to prevent is mediastinitis, a life threatening condition, with an extremely high mortality rate of 23% [15, 16]. Treatment of mediastinitis may consist of debridement, delayed closure and antibiotic therapy, broad spectrum. Despite this, most patients die, due to sepsis and multi organ failure [16].

Prognosis

The prognosis of AEF is, despite improving techniques, incredibly poor. In general, the mortality of patients with a gastrointestinal bleeding is high (12%) [17], and even higher in patients older than 70 years, with co-morbidity, or shock at presentation. In-hospital mortality ranges when bleeding is caused by an AEF differ between 40% (after conventional surgical treatment) and 60% (conservative or no treatment) [21].

The best treatment of AEF still remains unclear. In literature, different results with both procedures are reported. Immediate repair (one step approach) still is standard treatment [2]. Lately however, most authors prefer a two-step strategy for patients at risk (hemodynamic instability, shock, sepsis), as for these patients an immediate open repair might be too hazardous [12, 14, 18, 19]. With aortic stent-grafting, the esophagus is not repaired, which leads to a major risk of mediastinitis in the time between the two procedures [4]. Our case demonstrates the difficulty of deciding which strategy is best; both have a lot of risks, and outcomes are poor in both [12, 14, 18, 19, 20].

Conclusions

This case demonstrates that primary aorto-esophageal fistula may present as any other upper gastro-intestinal bleeding (such

as a Mallory-Weiss lesion or bleeding ulcer), but with much higher mortality. Only 11% of patients present with the classic Chiari triad [6]. Doctors should be aware of the possibility of AEF and try to select the patients at risk of this cause of bleeding. Most patients with a high suspicion of an AEF have a primary fistula. Only 5% of patients had previous (endo) vascular repair of the thoracic aorta in history. In all patients with an upper gastro-intestinal bleeding a chest X-ray in two directions should be performed in case of suspicion. In some cases the aneurysm is not shown on X-ray, as in our case; a Computer assisted Tomography of the chest should be performed then in case of clinical suspicion. The best treatment strategy remains unclear; the prognosis of an AEF still is very unfavorable.

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