








Thoracic endovascular aortic repair in management of aorto-oesophageal fistulas: a case series

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Background

Aorto-oesophageal fistula (AOF) is a rare, catastrophic disease with an extremely poor prognosis. A ruptured thoracic aortic aneurysm is a common aetiology for AOF. The clinical presentation is usually massive haematemesis and collapse. Timely diagnosis and appropriate treatment are crucial in managing AOF.

Case summary

We hereby report two cases of AOF, who underwent successful emergency thoracic endovascular aortic repair (TEVAR) to control active bleed and exsanguination. Case 1, an elderly lady with atherosclerotic aneurysm had TEVAR followed by open surgery for oesophageal rent and necrosed left main bronchus. Case 2 had mycotic tubercular aneurysm who later had infected graft-stent following TEVAR.

Discussion

Open surgery is the conventional treatment for AOF; however, TEVAR can be an alternative and less invasive approach in selected high-risk patients. Various management issues related to TEVAR with AOF have been discussed in the article.

Keywords

Case series • Aortic aneurysm • Mycotic aortic aneurysm • Aorto-oesophageal fistula • Infected endograft • Thoracic endovascular aortic repair • Tuberculosis

Learning points

- Aorto-oesophageal fistula is a rare, life-threatening emergency with poor prognosis.
- Clinical presentation includes massive haematemesis, exsanguination and haemodynamic collapse.
- Conventional open surgical repair carries a high morbidity and mortality.
- Thoracic endovascular aortic repair is a less invasive and alternative approach to the open surgical repair in selected high-risk patients.

Introduction

Aorto-oesophageal fistula (AOF) is a rare cause of upper gastrointestinal bleeding. Its aetiologies include thoracic aortic aneurysm,

oesophageal carcinoma, impacted foreign body in oesophagus, aortic or oesophageal interventions, and thoracic endovascular aortic repair (TEVAR).^{1,2} The clinical presentation is usually massive haematemesis and haemodynamic collapse. It is invariably fatal if

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urgent management is not performed. Though surgery is a conventional treatment for ruptured thoracic aortic aneurysm with AOF, TEVAR can be an alternative and less invasive approach in selected high-risk patients.³ We hereby report 2 cases of AOF, who underwent successful emergency TEVAR to control the active bleeding and discussed the various management issues related to AOF.

Timeline

Case 1	
A 60-year-old female presented with dysphagia and	For 1 week
Haematemesis—3 episodes	For 1 day
Emergency endoscopy (on the day of presentation)—Day -1	Pulsatile extraluminal mass causing luminal narrowing of oesophagus
Computed tomography (CT) angiography (on the day of presentation)—Day -1	Descending thoracic aorta (DTA) pseudoaneurysm
Index procedure—Day 0	Emergency thoracic endovascular aortic repair (TEVAR) of aortic pseudoaneurysm
Post-procedure	
Day +1	Left lung collapse
Day +5	Open surgical repair
Day +10	Melena and hypotension. Succumbed to her illness in spite of resuscitation
Case 2	
A 42-year-old female presented with dysphagia (2 months after completion of anti-tubercular therapy) and	For 1 month
Haematemesis—2 episodes	For 1 day
CT angiogram	DTA pseudoaneurysm with contained rupture compressing the oesophagus
Index procedure—Day 0	Emergency TEVAR
Post-procedure	
1 month	Discharged in haemodynamically stable condition
At 7 months of TEVAR	Diagnosed with endograft infection. Started on antibiotics and responded to treatment. Planned for open repair after 6 months of antibiotics
After 12 months of TEVAR	Massive haematemesis, exsanguination, and succumbed to illness

Case presentations

Case 1

A 60-year-old female, a known case of chronic obstructive airway disease on steroids and long-acting beta-agonist inhaler with no other

comorbidities, presented in the emergency room with rapidly progressive dysphagia to both solids and liquids for the last 7 days and three episodes of haematemesis in last 1 day. On physical examination, she was drowsy, had a blood pressure of 80/50 mmHg on vasopressor support, respiratory rate of 34/min, oxygen saturation of 90% at FiO₂ of 0.4 (PaO₂ 50 with PaO₂/FiO₂ ratio of 125) with reduced air entry on the left side of the lung and with normal heart sounds. Her plasma haemoglobin was 7.6 g% (12–18 g%), while routine serum biochemistry was normal. A 2-day-old chest X-ray before the admission was normal (Figure 1A). She was managed with multiple units of blood transfusion, broad-spectrum intravenous antibiotics (Meropenem and Teicoplanin), mechanical ventilation, and supportive therapy. An emergency oesophagoscopy revealed a pulsatile extraluminal mass with overlying mucosal ulceration and luminal narrowing of the oesophagus. A computed tomography (CT) angiogram showed a large 43 mm × 33 mm × 37 mm pseudoaneurysm of descending thoracic aorta (DTA) at the level of D5–D6 thoracic vertebrae (Figure 1B), with surrounding haematoma and active contrast leak into the oesophagus suggestive of AOF. After our institutional multidisciplinary team meeting, and informed patient consent, an emergency TEVAR was performed on an emergent basis. A 32 mm × 140 mm thoracic stent-graft (Zenith TX2 TAA Endovascular Graft with Pro-Form, Cook Medical Inc., Bloomington, IN, USA) was deployed in DTA to exclude the pseudoaneurysm (Figure 1C). Post-procedure Day 1, she developed high-grade fever with worsening ventilation parameters secondary to left lung collapse (Figure 2A). A repeat CT scan chest revealed a compressed left main bronchus and collapsed left lung (Figure 2B). A bedside bronchoscopy revealed extensive compression of left main bronchus and its upper and lower lobe bronchi, along with the collapse of the left lung. Even after mechanical ventilation support, she had worsening lung performance and therefore subjected to open surgical exploration on Day 5. A left posterolateral thoracotomy was performed. Intraoperative findings include ~5 cm × 5 cm size haematoma with abscess formation around aortic isthmus, compressing and strangling the left main bronchus, along with a large 1 cm × 2 cm bronchopleural fistula. The mid-thoracic part of the oesophagus was denuded—necrosed with a rent of 2 cm × 2 cm size (Figure 2C) and had pus discharge. The left pneumonectomy was performed as the left main bronchus was necrosed and irreparable. The oesophagus was repaired with an intercostal muscle flap. A cervical oesophagostomy, tube gastrostomy, and feeding jejunostomy were performed along with debridement of infected tissue. Postoperatively, the patient had a gradual and steady recovery and could be extubated on the fourth postoperative day of open surgical exploration. On the next day, she had an episode of massive melena and hypotension, which was resuscitated with blood transfusion, intravenous fluids, and inotropic support. However, she had persisted hypotension and died of cardio-respiratory arrest. An autopsy could not be done.

Case 2

A 42-year-old female with no comorbidities was evaluated for backache in May 2015 and found to have disseminated tuberculosis involving bilateral lungs, spine (D5–D6 thoracic vertebrae) with a paravertebral abscess at the same level. A CT scan detected a small DTA pseudoaneurysm of size 3.1 cm × 2.6 cm × 4.1 cm with a narrow neck at the level of D10–D12 thoracic vertebrae (Figure 3A). She was started on anti-tubercular therapy (ATT). A repeat CT after

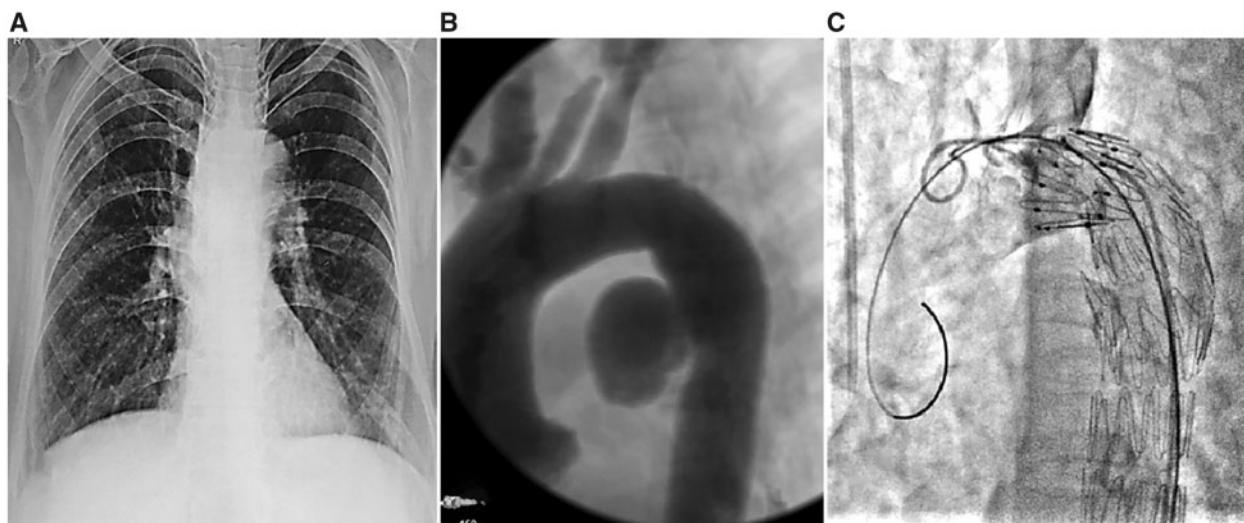


Figure 1 The chest X-ray was unremarkable (A). A computed tomogram angiogram showed a large pseudoaneurysm of descending thoracic aorta (B), which was successfully excluded by a stent-graft (C) following transthoracic endovascular aortic repair.

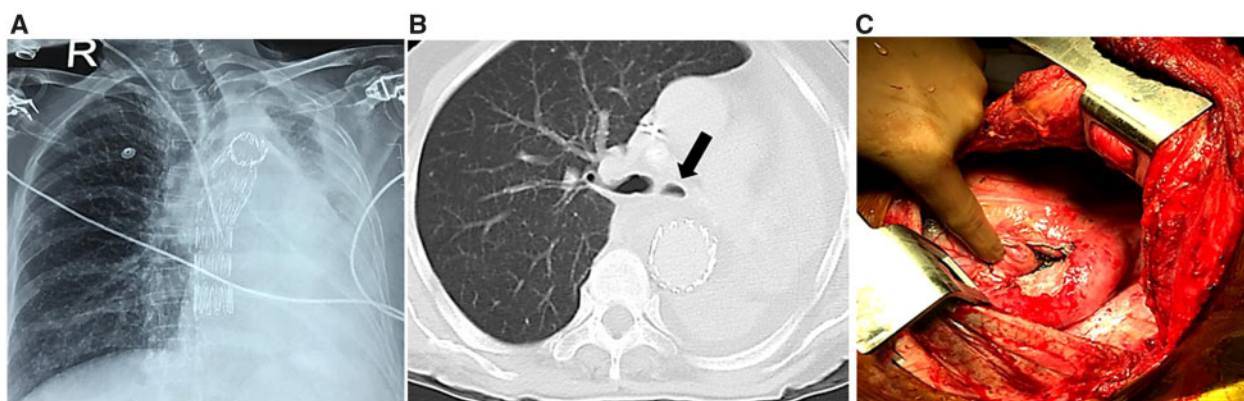


Figure 2 A chest X-ray showed the collapsed left lung with aortic stent-graft *in situ* (A). Axial computed tomography chest showed compressed left main bronchus (black arrow), collapsed left lung, and aortic stent-graft *in situ* (B). An operative image showed a rent in the oesophagus with surrounding unhealthy wall (C).

5 months showed similar findings of the pseudoaneurysm (Figure 3B). She completed an extended 14-month course of ATT. At 16 months of follow-up (September 2016), she presented to the emergency room with progressive dysphagia initially for solids and subsequently for liquids from the last 1 month and two recent episodes of haematemesis. Physical examination revealed tachycardia (pulse rate of 120/min), blood pressure of 90/60 mm of Hg, and tachypnoea with a systemic oxygen saturation of 94% on room air. Clinical examination revealed marked cachexia, weight loss (body mass index 16 kg/m²), and dehydration, for which she was optimized with intravenous fluids and two units of blood transfusion. The systemic examination of the respiratory and cardiac system was unremarkable. An oesophagoscopy showed near-total occlusion of the lower oesophagus by a large pulsatile extraluminal mass and overlying mucosal ulceration

and large clot (Figure 4A). A contrast oesophagogram showed hold up of contrast at the lower oesophagus (Figure 4B). Suspecting a large DTA pseudoaneurysm externally compressing the oesophagus with AOF, a repeat CT angiogram was performed. It showed the pre-existing pseudoaneurysm enlarged to the size of 11 cm × 8 cm × 6.6 cm (Figure 3C) and compressing the oesophagus along with a contained rupture. After our institutional multidisciplinary team meeting, and informed patient consent, an emergency TEVAR was performed. A 28 mm × 28 mm × 100 mm Valiant Thoracic stent-graft (Medtronic Vascular, Santa Rosa, CA, USA) was deployed in DTA to exclude the pseudoaneurysm. Her dysphagia to liquid continued even after a week of TEVAR. A barium oesophagogram showed persistent stasis of contrast at the lower oesophagus (Figure 4D). A nasojejunal feeding tube was inserted under

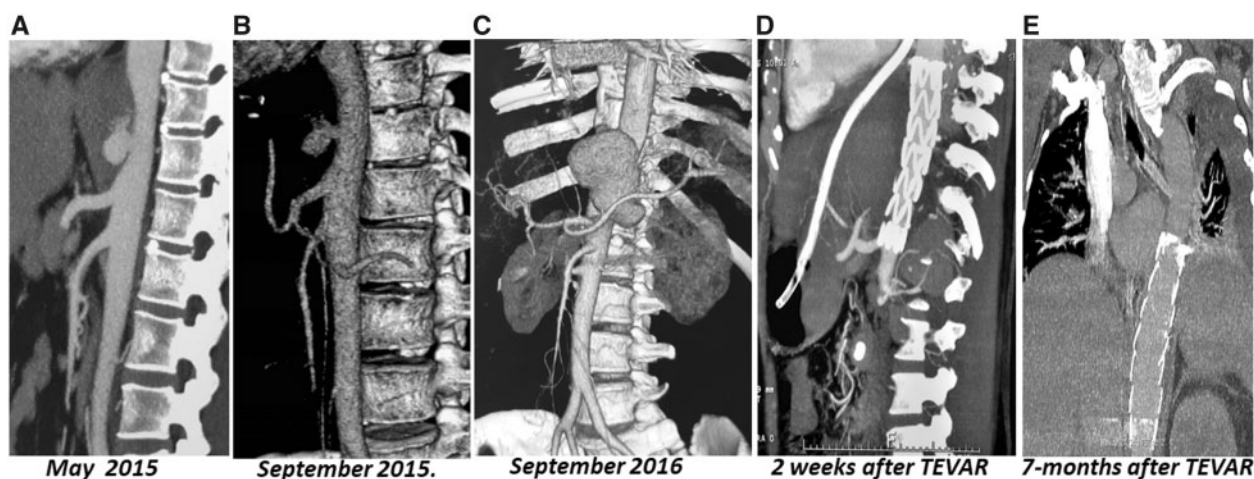


Figure 3 The computed tomography angiogram showed a small descending thoracic aorta pseudoaneurysm with a narrow neck at the level of D10–D12 thoracic vertebrae (A). There is no appreciable increase in the size of pseudoaneurysm after 5 months (B). A coronal reconstructed volume-rendered computed tomography angiographic image showed a large size of pseudoaneurysm above the coeliac trunk (C). A repeat computed tomography after 2 weeks of thoracic endovascular aortic repair showed patent stent-graft and no endoleak (D). A computed tomography at 7 months of follow-up showed patent stent-graft, no endoleak, and no evidence of infection (E). TEVAR, thoracic endovascular aortic repair.

oesophagoscopy guidance. The repeat oesophagoscopy revealed a large defect in the wall of lower oesophagus with necrotic tissue overlying the aortic metal stent (Figure 4C). Two weeks later, she had one episode each of mild haematemesis and melena. A repeat oesophagoscopy at 4 weeks revealed partial healing of oesophageal defect with granulation tissue overlying the graft-stent (Figure 4E). A repeat CT showed no endoleak through stent-graft (Figure 3D). The oesophageal perforation was managed conservatively with continued nasojunal tube feeding and a broad-spectrum antibiotic (Piperacillin, Tazobactam, and Vancomycin) for 4 weeks duration. The patient had a gradual recovery with no further episodes of haematemesis. She could be discharged after a month of hospital stay on nasojunal tube feeding and oral antibiotics (Moxifloxacin and Linezolid) for another 1 month. Following the gradual improvement in dysphagia, the feeding tube could be removed after 2 months of TEVAR. She remained asymptomatic for the next 7 months, with gradual improvement in weight, and no complaints of dysphagia, haematemesis, or fever.

After 7 months of follow-up following TEVAR (April 2017), she presented with high-grade fever, recurrent vomiting, and pain abdomen. There was no history of haematemesis or melena. Investigation revealed 6.9 g% (12–18 g%) haemoglobin, 46 200/mm³ (4000–11 000/mm³) total leucocyte count, and 2.1 mg% (0.5–1.2 mg%) serum creatinine. Ultrasound abdomen revealed hepatomegaly and multiple splenic abscesses. Blood culture and splenic aspirate were sterile for bacteria and fungus. A CT scan confirmed patent stent-graft with no endoleak (Figure 3E), multiple splenic hypodense areas, and left pleural effusion. A repeat oesophagoscopy revealed healed and scared lower oesophagus (Figure 4F). A positron emission tomography CT showed high radio-tracer uptake at the mid-part of infected stent-graft (Figure 5). A diagnosis of septic shock secondary to stent-graft infection was made. She was treated with 12 weeks of

intravenous antibiotics (Meropenem and Vancomycin) and other supportive treatment. A multidisciplinary team meeting was held to decide about the further line of management. The decision was to continue the conservative management with an extended antibiotic coverage for at least 6 months, before open surgery. Subsequently, she was discharged on oral antibiotics (Moxifloxacin and Linezolid) for the next 12 weeks. However, after 8 weeks of domiciliary, oral antibiotic course, she had one bout of massive haematemesis, presented in the emergency room in an exsanguinated state with shock and succumbed to the illness. An autopsy could not be done.

Discussion

The most common cause of AOF is a ruptured thoracic aortic aneurysm. The aetiology for an aneurysm is mostly atherosclerosis in elderly patients (Case 1),^{1,3} but rarely it can be a sequelae of a tubercular mycotic aneurysm (Case 2).^{1,4,5} The usual clinical presentation is backache and dysphagia of variable duration, before massive haematemesis.^{1,3} A high clinical suspicion is required for its diagnosis. A sentinel fresh red blood containing haematemesis, followed by rapidly fatal, massive exsanguinating haematemesis can provide the clinical clue for AOF.^{1,3} A oesophagoscopy revealing a pulsatile extrinsic mass with mucosal ulceration and luminal compression of the oesophagus is suggestive of adjacent DTA aneurysm as an aetiology for AOF.^{3,6,7} The clinical presentation of massive exsanguinating haematemesis and visibility of pulsatile extrinsic mass on oesophagoscopy in our two cases made us suspicious about DTA aneurysm, which was confirmed by CT angiography. As the course of the disease is so rapid and fatal, the vast majority of patients die before a definitive intervention could be done for AOF.¹

Conventional treatment of AOF is the open surgical repair.^{2,8,9} It includes aortic aneurysm repair with prosthetic graft or homograft,

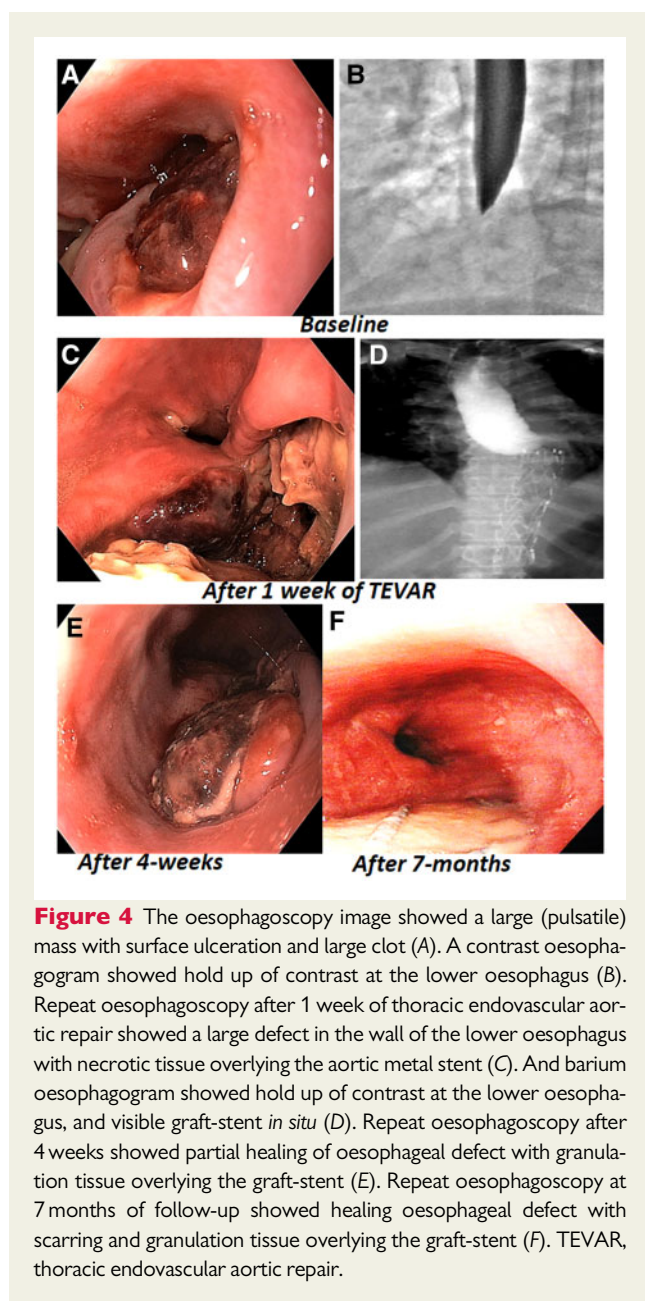


Figure 4 The oesophagoscopy image showed a large (pulsatile) mass with surface ulceration and large clot (A). A contrast oesophagogram showed hold up of contrast at the lower oesophagus (B). Repeat oesophagoscopy after 1 week of thoracic endovascular aortic repair showed a large defect in the wall of the lower oesophagus with necrotic tissue overlying the aortic metal stent (C). And barium oesophagogram showed hold up of contrast at the lower oesophagus, and visible graft-stent *in situ* (D). Repeat oesophagoscopy after 4 weeks showed partial healing of oesophageal defect with granulation tissue overlying the graft-stent (E). Repeat oesophagoscopy at 7 months of follow-up showed healing oesophageal defect with scarring and granulation tissue overlying the graft-stent (F). TEVAR, thoracic endovascular aortic repair.

judicious debridement of infected area, and the oesophageal repair (oesophagectomy with subsequent reconstruction/repair or oesophagus repair with intercostal muscle flap reinforcement). However, open surgery has a high risk of morbidity and mortality. Few of the case series demonstrated a 40–45% mortality with open surgical repair.^{8,9} An emergency TEVAR can be an alternative and less invasive approach in certain situations such as active life-threatening bleeding from the aorta, as a bridge to definite surgery and in patients with high surgical risk.^{10–12} Though TEVAR has shown favourable short-term outcomes (30-day mortality of 19%),^{10,11} there is a risk of stent-graft infection and recurrence of AOF as oesophageal defect and mediastinal infection are not adequately managed with TEVAR alone.^{3,12} Both of our cases were at high risk, hence underwent TEVAR instead of open surgical repair.

Case 1 had open surgical exploration after 5 days of TEVAR, because of associated lung complications. According to the surgical protocol, she underwent oesophageal repair and reconstruction along with mediastinal debridement and haematoma/abscess evacuation.^{8,9} In addition, left pneumonectomy was also performed for non-reparable left main bronchus necrosis.¹³ An aortic stent-graft replacement was not performed, as there was no endoleak, though some authors had routinely replaced the aortic stent-graft during open surgery to prevent graft infection and secondary AOF.^{10,12}

Tubercular mycotic aneurysm (Case 2) presenting as AOF is very rare.^{1,4,5} Initially, as the pseudoaneurysm was small along with active tuberculosis, we continued ATT for 14 months without intervening for mycotic pseudoaneurysm. A 5-month follow-up CT showing non-progression of pseudoaneurysm was also assuring about the conservative approach. However, at 16 months of follow-up, she presented with a rapid increase in the size of pseudoaneurysm along with AOF, which was treated with an emergency TEVAR. Following TEVAR, as the oesophagus had spontaneous healing/scarring of its fistulous site, and the bacterial infection of AOF was adequately treated with 2 months of antibiotics therapy, we discontinued it later on. The duration of antibiotic therapy beyond 4 weeks should be tailored depending upon the clinical and laboratory parameters of infection.^{10,12} An initial strategy of TEVAR alone, followed by a routine open surgical repair of the oesophagus, with or without aortic reconstruction and stent-graft explantation could be an alternative in the index case.^{10,12}

However, she later presented with gross stent-graft infection with septic shock after 7 months of TEVAR. The constitutional symptoms of malaise, weight loss, and weakness following TEVAR suggestive of ongoing low-grade infection were possibly ignored by us. In a systematic review by Canaud *et al.*,¹⁰ the incidence of endograft infection following aortic repair of AOF was ~15%. The infection was appropriately treated with a prolonged antibiotic course in the index case. Open surgery is indicated to remove an infected endograft in all the suitable patients, otherwise, the mortality is very high.^{14,15} It was a crucial decision that when to remove the infected endograft along with aortic reconstruction in the index case. These decisions depend upon the patient's general condition and comorbidities, infectious complications, active haemorrhage, and type and extent of surgery.¹⁴ We decided to give at least 6 months of antibiotic course before the open surgery which would have constituted extensive debridement of marked mediastinal adhesions, *in situ* or extra-anatomical aortic reconstruction, additional mesenteric arterial revascularization, and possible oesophageal repair. However, she died off before completing the 6 months of the antibiotic course. The terminal event of massive gastrointestinal bleeding in both the cases was possibly because of the recurrence of AOF secondary to continued ongoing infection in the stent-graft and adjacent aorta.

Conclusion

Aorto-oesophageal fistula is a rare, catastrophic disease with an extremely poor prognosis. As there are no large series and consensus management guidelines, any treatment strategy in these patients is highly individualized. As a multidisciplinary team approach is required to manage such patients, the experience and expertise of the team along with patient-related risk factors play an important role in strategic planning and management. A large prospective registry could

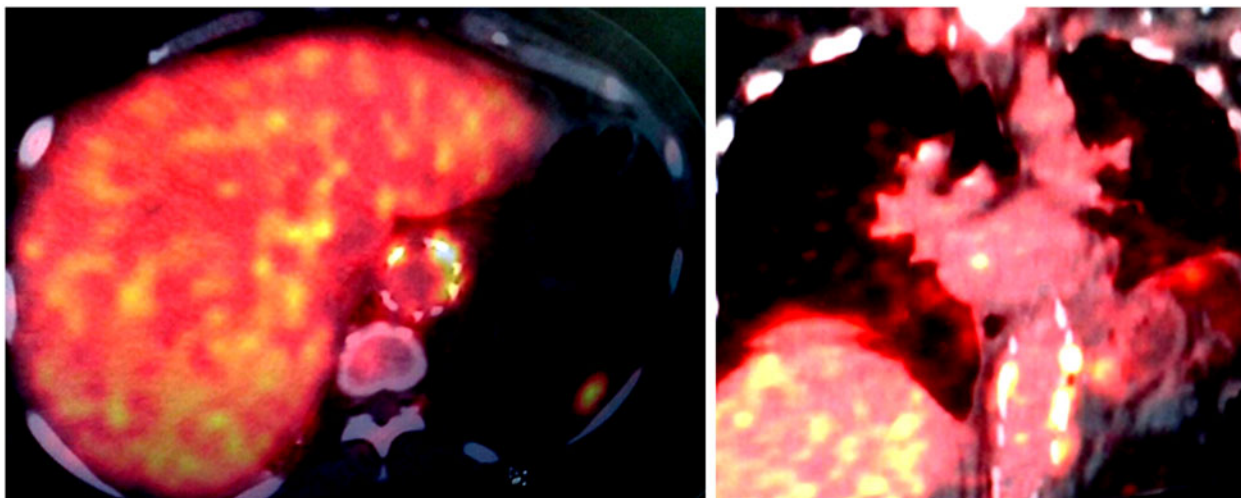


Figure 5 A positron emission tomography computed tomography angiogram in the axial and coronal section showed high standardized uptake value of radio-tracer at the mid-part of infected graft-stent.

help to formulate the guidelines for the management of this rare and catastrophic disease in the future.

Lead author biography



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Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patients in line with COPE guidance.

Conflict of interest: none declared.

References

- Hollander JE, Quick G. Aortoesophageal fistula: a comprehensive review of the literature. *Am J Med* 1991;**91**:279–287.
- Lawrie GM, Earle N, Bakey ME. Evolution of surgical techniques for aneurysms of the descending thoracic aorta: twenty-nine years experience with 659 patients. *J Cardiac Surgery* 1994;**9**:648–661.
- Chiesa R, Melissano G, Marone EM, Kahlberg A, Marrocco-Trischitta MM, Tshomba Y. Endovascular treatment of aortoesophageal and aortobronchial fistulae. *J Vasc Surg* 2010;**51**:1195–1202.
- Yeow Y, Fong SS, Rao J, Sim R. Aorto-oesophageal fistula from miliary tuberculosis: a rare cause of massive haematemesis. *Ann Acad Med Singap* 2014;**43**:559–560.
- Byard RW. Lethal aorto-oesophageal fistula—characteristic features and aetiology. *J Forensic Leg Med* 2013;**20**:164–168.
- Heckstall RL, Hollander JE. Aortoesophageal fistula: recognition and diagnosis in the emergency department. *Ann Emerg Med* 1998;**32**:502–505.
- Lau H. Dysphagia aortica: harbinger of aortoesophageal fistula? *Surgery* 2001;**129**:763–764.
- Kieffer E, Chiche L, Gomes D. Aortoesophageal fistula: value of in situ aortic allograft replacement. *Ann Surg* 2003;**238**:283–290.
- Saito A, Motomura N, Hattori O, Kinoshita O, Shimada S, Saiki Y et al. Outcome of surgical repair of aorto-esophageal fistulas with cryopreserved aortic allografts. *Interact Cardiovasc Thorac Surg* 2012;**14**:532–537.
- Canaud L, Ozdemir BA, Bee WW, Bahia S, Holt P, Thompson M. Thoracic endovascular aortic repair in management of aortoesophageal fistulas. *J Vasc Surg* 2014;**59**:248–254.
- Jonker FH, Schlösser FJ, Moll FL, van Herwaarden JA, Indes JE, Verhagen HJ et al. Outcomes of thoracic endovascular aortic repair for aortobronchial and aortoesophageal fistulas. *J Endovasc Ther* 2009;**16**:428–440.
- Topell, Stehr A, Steinbauer MG, Piso P, Schlitt HJ, Kasprzak PM. Surgical strategy in aortoesophageal fistulae: endovascular stentgrafts and in situ repair of the aorta with cryopreserved homografts. *Ann Surg* 2007;**246**:853–859.
- Canaud L, D'Annunzio T, Ozdemir BA, Marty-Ané C, Alric P. Combined endovascular and surgical approach for aortobronchial fistula. *J Thorac Cardiovasc Surg* 2014;**148**:2108–2111.
- Setacci C, Chisci E, Setacci F, Ercolini L, de Donato G, Troisi N et al. How to diagnose and manage infected endografts after endovascular aneurysm repair. *Aorta* 2014;**2**:255–264.
- Lyons OT, Patel AS, Saha P, Clough RE, Price N, Taylor PR. A 14-year experience with aortic endograft infection: management and results. *Eur J Vasc Endovasc Surg* 2013;**46**:306–313.