American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2018; 19: 1025-1029
DOI: 10.12659/AJCR.910283

Received: 2018.03.28
Accepted: 2018.05.16
Published: 2018.08.28

Aortoesophageal Fistula Causing Massive Gastrointestinal Bleeding and Death in a Patient with Dermatomyositis: A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

AEF 1 Teressa Reanne Ju
CDF 2 Chi Chan Lee
ACDF 1 Yu-Chao Lin

1 Department of Pulmonary and Critical Care Medicine, China Medical University Hospital, Taichung, Taiwan
2 Department of Critical Care Medicine, Oregon Health Science University Hospital, Portland, OR, U.S.A.

Corresponding Author: Yu-Chao Lin, e-mail: D10001@mail.cmuh.org.tw
Conflict of interest: None declared

Patient: Female, 46
Final Diagnosis: Aortoesophageal fistula
Symptoms: Gastrointestinal bleeding • hypotension
Medication: —
Clinical Procedure: Angiography • esophagogastroduodenoscopy
Specialty: Pulmonology

Objective: Rare disease

Background: Aortoesophageal fistula is a rare etiology of serious gastrointestinal bleeding. Most aortoesophageal fistulas resulted from thoracic aortic aneurysms, foreign bodies, or esophageal malignancy. To our knowledge, spontaneous aortoesophageal fistula due to dermatomyositis and high dose steroid therapy has not been reported.

Case Report: A 46-year-old Asian female with a history of dermatomyositis and duodenal ulcers presented with black stool for one day. She was initially admitted for dermatomyositis flare-up and received high dose steroid therapy. Four weeks after discharge, she experienced gastrointestinal bleeding from multiple duodenal ulcers. Due to a continuous fall in hemoglobin level, she received angiography and embolization to the gastroduodenal artery. After the procedure, the patient developed another episode of dermatomyositis flare-up and required endotracheal intubation. During ventilator weaning, she developed recurrent gastrointestinal bleeding. Repeated esophagogastroduodenoscopy showed one esophageal ulcer with active bleeding. Epinephrine hemostasis therapy was performed but with poor therapeutic response. Angiography showed no visible extravasation. Chest computed tomography with intravenous contrast revealed contrast extravasation in esophageal lumen with blood clots consistent with an aortoesophageal fistula. Despite our attempt to arrange an emergent endovascular stent, the patient went into cardiac arrest from circulatory collapse.

Conclusions: Dermatomyositis leads to esophageal structural abnormalities through various mechanisms. Aortoesophageal fistula is a life-threatening etiology of gastrointestinal bleeding and should be suspected if bleeding from an esophageal ulcer responds poorly to hemostatic treatment. Abdomen computed tomography with intravenous contrast is the preferred image modality for diagnosing aortoesophageal fistula. Thoracic endovascular aortic repair is a reasonable procedure to stop bleeding in patients with unstable hemodynamic profiles.

MeSH Keywords: Aorta, Thoracic • Dermatomyositis • Esophageal Fistula • Gastrointestinal Diseases

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/910283

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

Background

Aortoesophageal fistula (AEF) is a rare etiology of serious gastrointestinal (GI) bleeding. It is an abnormal passage between the esophagus and the aorta. Most AEFs resulted from thoracic aortic aneurysms, foreign bodies, or esophageal malignancy [1]. To the best of our knowledge, AEF due to dermatomyositis had not been reported. Here we present a case of a patient with an underlying history of dermatomyositis who received high dose corticosteroid therapy. She developed an AEF from a spontaneous esophageal ulcer and experienced massive upper GI bleeding from the lesion. No anatomical vascular abnormalities, foreign body ingestion, or malignancy were noted.

Case Report

A 46-year-old Asian female with a history of dermatomyositis and recent diagnosis of duodenal ulcer presented to the hospital with tarry stool for one day. Her dermatomyositis was under good control with oral methylprednisone 6 mg per day, mycophenolate 1500 mg per day, and cyclosporine 100 mg per day. One month prior to her current admission, she was hospitalized because of flare-up of dermatomyositis and received 3 days of intravenous therapy (IV) methylprednisolone pulse therapy. Complaining of dysphagia, she also had an upper endoscopy, which showed multiple duodenal ulcers and stigmata of recent hemorrhage. A biopsy of ulcers disclosed chronic duodenitis with a positive result for *Helicobacter pylori*. No malignancy was noted. Esophageal transit study showed mild delay in emptying bolus fragmentation in the mid-lower esophagus possibly related to motility disorder. Her complete blood count revealed hemoglobin of 11.7 milligrams per deciliter. Platelet count was 145 000 per microliter and coagulation profiles were within normal range. Since her muscle weakness improved and no active bleeding was noted, she was discharged with her regular medications and oral proton pump inhibitor (PPI).

Four weeks later, the patient presented to the hospital with black stool for one day. She also experienced nausea and epigastric pain. No hematemesis or dysphagia were reported. Her serum hemoglobin dropped to 8.6 milligrams per deciliter. Her upper endoscopy revealed multiple ulcers in variable sizes with exudates located from bulb to the third portion. No esophageal lesions were seen. Fresh blood was noted but the bleeder could not be found. Due to continuous fall in hemoglobin level, the patient underwent angiography. Contrast extravasation in branch of gastroduodenal artery was noted, and 2 microcoils were delivered for embolization. Follow-up angiography showed no contrast extravasation.

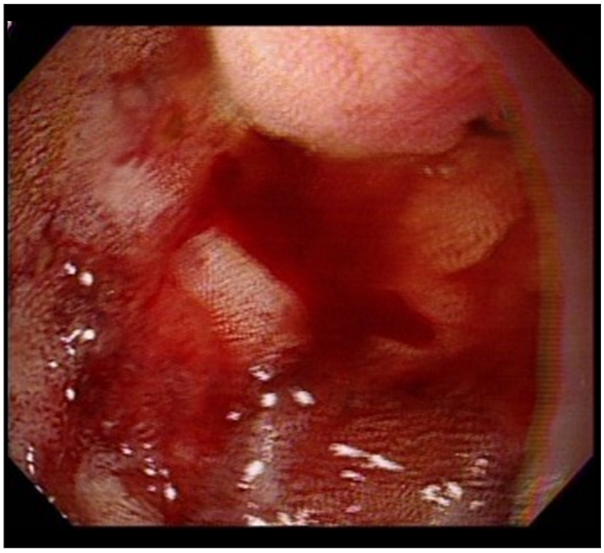

Figure 1. Upper endoscopy: fresh blood noted in esophageal lumen and visualization is limited.

After the procedure, patient's hemoglobin and hemodynamic profiles were stable. Elevated serum gastrin level of 921 picograms per milliliter was noted in the setting of PPI infusion. Abdomen computed tomography (CT) with IV contrast showed no evidence of gastrinoma. Although the bleeding was mitigated, the patient developed respiratory muscle weakness and required mechanical ventilator support. A nasogastric (NG) tube was placed. Under the impression of a recurrent dermatomyositis flare-up, rituximab infusion and high dose methylprednisone were started, but both failed to improve her muscle weakness. She then received 5 days of IV immunoglobulin therapy. Due to weaning failure from mechanical ventilation, she underwent tracheotomy for airway access.

While patient underwent ventilator weaning in the respiratory care unit, coffee ground material from the NG tube was noted. Repeated upper endoscopy showed one esophageal ulcer, located at 30 cm from the incisor with massive bleeding. Fresh blood limited the visualization and no satisfactory endoscopic image could be made (Figure 1). Epinephrine hemostasis therapy was performed but with poor therapeutic response. Sengstaken-Blakemore tube (SBT) was inserted in an attempt to achieve temporary hemostasis. The patient underwent repeated angiography, but no visible extravasation was seen. Chest CT with intravenous contrast disclosed contrast extravasation in esophageal lumen with blood clots (Figures 2, 3). The bleeding point seemed to be from the thoracic aorta, and an aorta-esophageal fistula was highly suspected. She experienced massive blood loss and developed progressive hypovolemic shock despite blood transfusion and fluid resuscitation. Despite our attempt to arrange emergent endovascular stent, the patient went into cardiac arrest due to circulatory collapse.

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

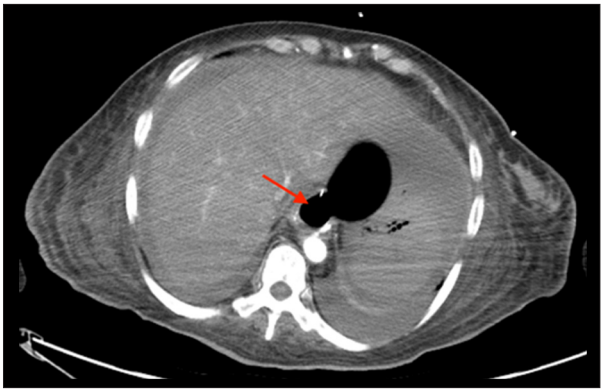

Figure 2. Axial view of abdomen computed tomography scan: extravasation of intravenous therapy contrast into esophageal lumen (esophageal lumen shown in red arrow).

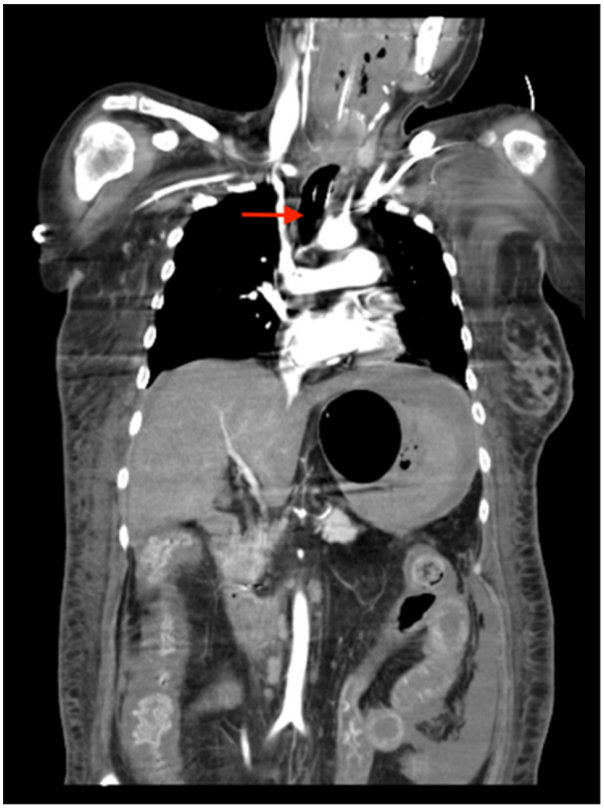

Figure 3. Coronal view of abdomen computed tomography scan: extravasation of intravenous therapy contrast into esophageal lumen (esophageal lumen shown in red arrow).

Discussion

Since the first AEF was reported in 1967 [2], cases have been identified to address this life-threatening structural anomaly. It is estimated that 78% of AEFs result from thoracic aortic aneurysms, foreign bodies, or esophageal malignancy [1]. Other common etiologies included post-surgical complications [3], GERD [4], and tuberculosis [5].

In our case, while no common cause of AEF was noted, the patient did have severe dermatomyositis and received 2 rounds of corticosteroid pulse therapy. A literature review conducted by Marie et al. found the prevalence of esophageal involvement in dermatomyositis and polymyositis was 40% [6]. Dermatomyositis mostly impaired esophageal motility, while it could also result in structural abnormalities, including stricture, fistula, or ulcer formation. In a case series assessing esophageal abnormalities in patients with severe dermatomyositis and polymyositis, 8 out of 16 cases had ulcerative lesions involving esophageal mucosa [7]. Severe dermatomyositis also delayed gastric and esophageal emptying and resulted in drug retention, which predisposed patients to drug-induced esophagitis [8]. Suzuki et al. reported that a patient with severe Parkinsonism died from GI bleeding due to AEF [9]. In that case, AEF was assumed to be a product of local esophageal injury due to drug retention. It is worth mentioning that several reported cases suggested an association between dermatomyositis and occult esophageal malignancy [10,11]. We postulate that the esophageal ulcer in our case was not a result of an underlying malignancy since it was not visualized during the previous endoscopy. Rather, in our patient, we suspect that esophageal ulcer was formed either due to dermatomyositis itself or drug-induced esophagitis. The healing process of the ulcer was further compromised by high-dose systematic corticosteroid therapy, constant NG tube irritation. and the patient's critical illness. As a result, the esophageal ulcer may have progressively deepened and penetrated the esophageal wall and into the aorta.

Endoscopy is usually the initial test to be performed in patients presenting with GI bleeding. However, in the cohort study performed by Saers et al. [12], AEF detection rate with endoscopy was only 38%. In our case, we did notice an esophageal ulcer with active bleeding through endoscopy, and it responded poorly to homeostatic therapy.

At that time, AEF was not our tentative diagnosis, so we performed angiography first to identify the source of bleeding and to administer embolic agents. Later, we learned that angiography has a limited role for diagnosing AEF [1,12,13], since temporary occlusion of fistula by thrombus did occur, thus preventing contrast extravasation. In the case series written by Saer et al., the AEF detection rate through angiography was only 26%. On the other hand, in this article, 49 out of 81 patients who received abdominal CT with IV contrast had image findings suggesting AEF, including air within aortic wall, focal bowel wall thickening, or disruption of aortic fat cover. Abdomen CT appears to be the favorable diagnostic modality when AEF is highly suspected.

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

AEF can be managed either through surgery or thoracic endovascular aortic repair (TEAVR). Managing AEF with TEAVR alone carries a higher infection and long-term mortality rate [14]. Infection is a valid concern in TEAVR, since the fistula is left unrepaired, and bacteria can spread into the bloodstream from the esophagus lumen. In a literature review, stent graft infection occurred in 15.2% of the patients [15]. In recent years, the role of TEAVR in AEF has been considered as a bridging therapy to control the bleeding while patients are in shock [15,16]. Canaud et al. suggested TEVAR and concomitant or staged adjunctive procedures (resection, repair of the esophagus, or a planned stent graft explantation) were associated with a significantly lower incidence of aortic-related mortality (P=0.0121) [15]. We arranged TEAVR for our patient yet unfortunately she experienced circulatory collapse before the procedure.

It is worth mentioning that in this case we used an SBT for temporary hemostasis while waiting for further interventions. SBT is a double-balloon device inserted first into gastric lumen through the esophagus. Both balloons are then inflated so that one balloon is fixed in the gastric lumen while the other compresses the esophageal wall. This leads to direct compression of esophageal vasculature to arrest bleeding [17]. While conventionally used in patients with variceal bleeding, SBT has been applied to control esophageal bleeding due to other etiologies such as Mallory-Weiss tears [18], esophagitis [19], and Dieulafoy's lesion [20]. We found 3 case reports utilizing SBT in managing massive bleeding from AEF [21–23]. Complications following SBT, such as mucosal ulcerations, esophageal perforation, and aspiration pneumonitis, have been reported previously in the literature [22]. However, in our case, the esophageal ulcer was formed prior to SBT insertion and was not due to procedural complications. Physicians should be cautious of selecting cases which are indicated for SBT placement and vigilant of potential complications.

Conclusions

In a summary, dermatomyositis could lead to structural abnormalities in esophagus through different mechanisms. AEF is a life-threatening etiology of GI bleeding and should be suspected if bleeding from esophageal ulcer responds poorly to hemostatic treatment. Abdomen CT scan with IV contrast is the preferred image modality for diagnosing AEF. TEAVR appears to be a reasonable procedure to stop bleeding in patients with unstable hemodynamic profiles.

Department and Institution where work was done

Department of Pulmonary and Critical Care Medicine, China Medical University Hospital, Taichung, Taiwan.

Conflicts of interest

None.

References:

1. Hollander JE, Quick G: Aortoesophageal fistula: A comprehensive review of the literature. Am J Med, 1991; 91(3): 279–87
2. Sloop RD, Thompson JC: Aorto-esophageal fistula: Report of a case and review of literature. Gastroenterology, 1967; 53(5): 768–77
3. Augoustides JG, Gutsche JT, Fairman RM, Bavaria JE: A rationale for managing rare complications: evidence from the European Registry of Endovascular Aortic Repair Complications about aorto-oesophageal fistula. Eur J Cardiothorac Surg, 2014; 45(3): 458–59
4. Cronen P, Snow N, Nightingale D: Aortoesophageal fistula secondary to reflux esophagitis. Ann Thorac Surg, 1982; 33(1): 78–80
5. Na JY, Kim YS, Choi YD et al: Death by aortoesophageal fistula due to disseminated tuberculosis: A case study. Int J Clin Exp Pathol, 2015; 8(4): 4253–57
6. Marie I, Menard JF, Hatron PY et al: Intravenous immunoglobulins for steroid-refractory esophageal involvement related to polymyositis and dermatomyositis: A series of 73 patients. Arthritis Care Res (Hoboken), 2010; 62(12): 1748–55
7. de Merieux P, Verity MA, Clements PJ, Paulus HE: Esophageal abnormalities and dysphagia in polymyositis and dermatomyositis. Arthritis Rheum, 1983; 26(8): 961–68
8. Horowitz M, McNeil JD, Maddern GJ et al: Abnormalities of gastric and esophageal emptying in polymyositis and dermatomyositis. Gastroenterology, 1986; 90(2): 434–49
9. Suzuki H, Kawai S, Okada R et al: [Autopsy results of an elderly case of Parkinson's disease and aorto-esophageal fistula who died of a massive hematemesis during prolonged bed rest]. Nihon Ronen Igakkai Zasshi, 1993; 30(5): 387–92 [in Japanese]
10. Iftikhar I, Abdelmannan D, Daw HA: Dermatomyositis and esophageal cancer. South Med J, 2006; 99(7): 777–79
11. Zhang W, Jiang SP, Huang L: Dermatomyositis and malignancy: A retrospective study of 115 cases. Eur Rev Med Pharmacol Sci, 2009; 13(2): 77–80
12. Saers SJ, Scheltinga MR: Primary aortoenteric fistula. Br J Surg, 2005; 92(2): 143–52
13. Khawaja FI, Varindani MK: Aortoesophageal fistula. Review of clinical, radiographic, and endoscopic features. J Clin Gastroenterol, 1987; 9(3): 342–44
14. Jonker FH, Heijmen R, Trimarchi S et al: Acute management of aortobronchial and aortoesophageal fistulas using thoracic endovascular aortic repair. J Vasc Surg, 2009; 50(5): 999–1004
15. Canaud L, Ozdemir BA, Bee WW et al: Thoracic endovascular aortic repair in management of aortoesophageal fistulas. J Vasc Surg, 2014; 59(1): 248–54
16. Kubota S, Shiiya N, Shingu Y et al: Surgical strategy for aortoesophageal fistula in the endovascular era. Gen Thorac Cardiovasc Surg, 2013; 61(10): 560–64
17. Avgerinos A, Armonis A: Balloon tamponade technique and efficacy in variceal haemorrhage. Scand J Gastroenterol Suppl, 1994; 207: 11–16
18. Welch GH, McArdle CS, Anderson JR: Balloon tamponade for the control of Mallory-Weiss haemorrhage in patients with coagulation defects. Br J Surg, 1987; 74(7): 610–11
19. Chen YI, Dorreen AP, Warshawsky PJ, Wyse JM: Sengstaken-Blakemore tube for non-variceal distal esophageal bleeding refractory to endoscopic treatment: A case report & review of the literature. Gastroenterol Rep (Oxf), 2014; 2(4): 313–15

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

20. Ho KM: Use of Sengstaken-Blakemore tube to stop massive upper gastrointestinal bleeding from Dieulafoy's lesion in the lower oesophagus. Anaesth Intensive Care, 2004; 32(5): 711–14
21. Assink J, Vierhout BP, Snellen JP et al: Emergency endovascular repair of an aortoesophageal fistula caused by a foreign body. J Endovasc Ther, 2005; 12(1): 129–33
22. Seet E, Beevee S, Cheng A, Lim E: The Sengstaken-Blakemore tube: Uses and abuses. Singapore Med J, 2008; 49(8): e195–97
23. Kelly SL, Peters P, Ogg MJ et al: Successful management of an aortoesophageal fistula caused by a fish bone – case report and review of literature. J Cardiothorac Surg, 2009; 4: 21

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]