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CASE REPORT | ESOPHAGUS

Secondary Aortoesophageal Fistula Associated With Aneurysmal Graft Infection by *Coxiella burnetii*

Chinemerem John Okwara, MD[1], Jan Petrasek, MD, PhD[2], Maeghan Gibson, MD[1], and Ezra Burstein, MD[2]

[1]*Department of Internal Medicine, University of Texas Southwestern Medical Center, Dallas, TX*
[2]*Division of Digestive and Liver Disease, University of Texas Southwestern Medical Center, Dallas, TX*

Abstract

Aortoesophageal fistula is a rare and serious condition that carries a high mortality rate. We present a case of overt gastrointestinal bleeding from an aortoesophageal fistula in a patient with chronic infection of an endovascular prosthesis with *Coxiella burnetii*.

Introduction

Aortoesophageal fistula is a rare and serious condition that carries a high mortality rate. It can occur in patients with thoracic aortic aneurysm who have undergone endovascular repair, especially if vascular graft prosthesis infection is present.[1] *Coxiella burnetii*, the organism responsible for Q fever, most commonly manifests as culture-negative endocarditis, but can infect vascular graft prostheses, leading to a number of potential complications.

Case Report

A 69-year-old white woman with a history of hypertension and thoracic aortic aneurysm underwent thoracic endovascular aortic repair (TEVAR) 8 months prior to admission. She had an uneventful postoperative course until 3 months prior to admission, when she developed abdominal pain, weight loss, and fatigue. Computed tomography (CT) angiography showed aneurysm of the aortic graft. Serologic tests revealed chronic Q fever. Treatment with doxycycline and hydroxychloroquine was initiated with plans for subsequent removal of the endograft.

On admission, the patient presented with excruciating abdominal and back pain, nausea with vomiting, and reported further weight loss over the preceding weeks. She was slightly tachycardiac with otherwise normal vital signs, but was cachectic, uncomfortable, and restless. A pulsatile mass was palpable in the epigastric region. Laboratory evaluation showed elevated C-reactive protein of 185 mg/dL, and a hemoglobin of 9.1 g/dL. The day after admission, the patient had an episode of hematemesis, and hemoglobin dropped to a nadir of 6.8 g/dL. A CT angiogram of the aorta demonstrated air bubbles surrounding the aneurysmal sac. Delayed arterial images showed continuous communication between the aneurysm sac and the esophageal lumen below the level of the carina, consistent with aortoesophageal fistula (Figure 1 and Figure 2). The size of the aneurysm had increased from 7.8 x 5.7 cm to 8.6 x 5.4 cm in its greatest dimensions.

The hematemesis was thought to represent a herald bleed from an aortoesophageal fistula. Emergent surgery was performed with replacement of the thoracoabdominal aorta with a 20-mm HEMASHIELD™ (Atrium USA, Hudson, NH) graft soaked in rifampin. She also underwent an esophagectomy, and a gastrostomy tube was placed for nutrition. A 2-cm communication between the aorta and the mid-esophagus was noted during the operation.

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Correspondence: Chinemerem Johan Okwara, 2140 Medical District Dr., #4043, Dallas, TX, 75235 (Chinemerem.okwara@phhs.org).

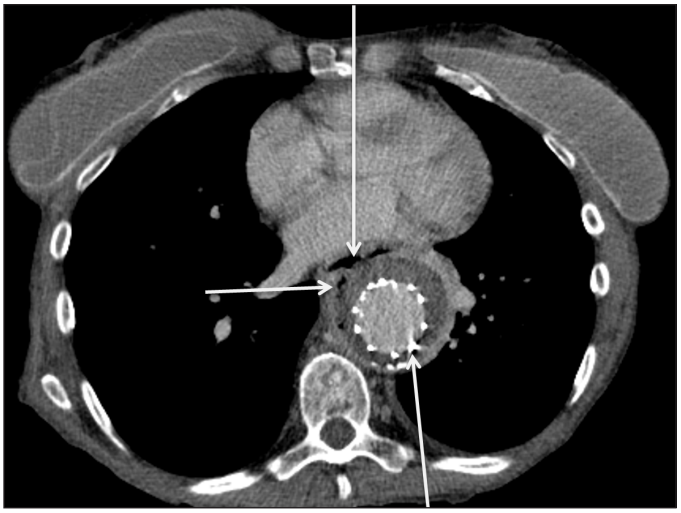

Figure 1. Communication between the aorta and esophagus with infected thrombus. The top arrow shows the esophagus, the left arrow shows air in the aneurysm sac, and the bottom arrow shows the descending aortic aneurysm with an endograft.

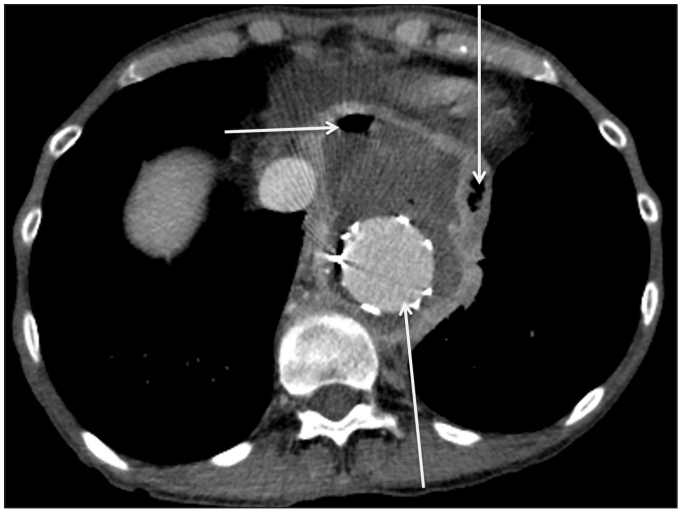

Figure 2. Descending aorta surrounded by infected thrombus with an air pocket within the aneurysm sac. The right arrow shows the esophagus, the left arrow shows air in the aneurysm sac, and the bottom arrow shows the descending aortic aneurysm with an endograft.

Post-operative complications included bilateral pleural effusions (greater on left side) with fluid analysis suggestive of chylothorax (pleural fluid triglyceride 1,233 mg/dL) and paraplegia that was thought to be due to perioperative spinal artery thrombosis, which improved gradually. The patient suffered neurogenic bladder, which required Foley catheter placement and bladder training. She remained in the hospital for 4 weeks and was subsequently discharged in good condition.

Discussion

Aortoenteric fistulas are a rare and deadly cause of gastrointestinal bleeding. Among aortoenteric fistulas, aortoesophageal fistulas account for only 10% of aortoenteric fistulas.[2] Aortoesophageal fistulas most commonly occur in the setting of thoracic aortic aneurysm after thoracic endovascular aortic repair (TEVAR). One study reported a 1.9% incidence of aortoesophageal fistulas in a cohort of patients that underwent TEVAR. Hematemesis and elevated inflammatory markers were associated with aortoesophageal fistulas. CT is the test of choice for diagnosis,although some reports have favored endoscopy.[2,3] We did not perform endoscopy, as we were concerned about the risk of dislodging a clot with the endoscope, which could precipitate massive bleeding from the aortoesophageal fistula.

Infection is not uncommon in the context of aortoesophageal fistulas in patients with TEVAR, and it is suggested to be the main mechanism of fistula formation.[4] The risk of endograft infection is rare (0.2–0.7%), but commonly associated organisms include methicillin-resistant *Staphylococcus aureus*, *Staphylococcus epidermidis*, and *Pseudomonas spp*.[5-7] *Coxiella burnetii*, the agent responsible for Q fever, has been reported as a cause of vascular infection in several cases, but to our knowledge has not been reported in association with aortoesophageal fistula.[1,7-9]

The most common treatment regimen for chronic Q fever infection is doxycycline 100 mg twice daily, and hydroxychloroquine 200 mg 3 times daily.[10,11] A treatment duration of 18 months has been used for *C. burnetii* endocarditis, supported by Centers for Disease Control recommendations for vascular infections.[1,11] Surgical excision of infected graft in addition to antibiotics is an integral part of treatment believed to confer a survival benefit compared to antibiotic treatment alone.[1] Serology studies should be performed monthly during the treatment period. Because of potential retinal toxicity from long-term use of hydroxychloroquine, a baseline ophthalmic examination should be performed before treatment and every 6 months thereafter.[11]

Aortoesophageal fistula is a rare condition with high mortality rate. Diagnosis requires high index of suspicion and early surgical intervention should be pursued. Presentation with hematemesis should raise concerns for subsequent bleeding, which may be fatal. Endovascular infection with *C. burnetii* can be difficult to diagnose, and a high level of suspicion is required, particularly in the context of negative blood cultures. Serologic studies should be considered in patients with history of vascular surgery presenting with a febrile illness, especially when a source is not apparent. Successful management of aortoesophageal fistula requires early diagnosis and a multidisciplinary approach with involvement of a surgical team, critical care physicians, and infectious disease physicians.

Disclosures

Author contributions: CJ Okwara and J. Petrasek wrote the manuscript. M. Gibson and E. Burstein revised the manuscript. CJ Okwara is the article guarantor.

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