

Achalasia Revealed by Respiratory Failure and Hemodynamic Instability

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ABSTRACT

Achalasia is a rare condition that most often presents with progressive dysphagia to solids and liquids. We report a case of achalasia presenting with acute respiratory failure and hemodynamic instability requiring both ventilator and vasopressor support because of extrinsic compression of the airway and left atrium by a dilated and fluid-filled esophagus. This is the first case reported of achalasia, causing both left atrial compression and airway compression.

INTRODUCTION

Achalasia is a primary esophageal motility disorder characterized by a lack of peristalsis and impaired relaxation of the lower esophageal sphincter (LES) during swallowing. Achalasia is secondary to neuronal degeneration of myenteric plexus ganglion cells in the esophagus and LES.¹ It is a rare condition, with an annual incidence of approximately 1 in 100,000 worldwide.² Achalasia classically presents with progressive dysphagia to both solids and liquids and is commonly associated with chest pain and regurgitation.³ The following is an unusual case of achalasia initially presenting as respiratory failure with cardiovascular compromise.

CASE REPORT

A 63-year-old woman with a history of ovarian cancer in remission and current tobacco dependence presented to an outside hospital with shortness of breath and was found to be in respiratory distress. Chest x-ray demonstrated a large, poorly defined extrapleural soft tissue density in the right paratracheal and pericardiac regions (Figure 1). Thoracic computed tomography demonstrated a severely dilated fluid-filled esophagus extending from the superior thoracic aperture caudally to the gastroesophageal (GE) junction with associated compression of the right bronchus and left atrium (Figure 2). While in the emergency room, the patient became hypoxic and was intubated. She was then transferred to our hospital for further evaluation with the diagnosis of acute hypoxic respiratory failure.

On arrival, her SpO₂ was in the 80s on FiO₂ of 70%. Her vital signs were notable for a blood pressure of 87/58 mm Hg and a heart rate of 100 beats per minute. Arterial blood gas revealed a pH of 7.31 and a pCO₂ of 82 mm Hg, consistent with respiratory acidosis. An echocardiogram demonstrated a normal left ventricular ejection fraction with pseudodyskinesis of the basal inferolateral wall secondary to extrinsic compression and severe compression of the left atrium (Figure 3).

Esophagogastroduodenoscopy confirmed a massively dilated and tortuous esophagus, with a large amount of retained food and fluid, which was gradually removed endoscopically. Severe esophagitis with multifocal esophageal ulceration secondary to stasis was noted, along with moderate resistance in traversing the GE junction. The cardia was examined and was unremarkable. During the procedure, the patient required both norepinephrine and dobutamine to maintain adequate perfusion pressures. Because the bolus was removed and the esophagus decompressed, the patient was weaned from pressor support. Within a few hours after the



Figure 1. Chest x-ray depicting soft tissue density in the right paratracheal and pericardiac regions, with no gastric air bubble visualized.

procedure, she was extubated to room air. A repeat echocardiogram demonstrated normal left atrial size with reduced extrinsic compression (Figure 4).

Once extubated, further history was obtained from the patient. She reported progressive dysphagia to solids over the past 4 weeks leading up to admission. She denied liquid dysphagia. The dysphagia was associated with throat pain, nausea, and frequent belching. The patient's shortness of breath began 1 week before presentation with gradual worsening throughout the week. She felt as though she "could not fill her lungs." She denied a history of similar symptoms and had no history of cardiopulmonary disease.

Once the patient was extubated and stable from a clinical standpoint, further testing was performed. Medtronic high-resolution esophageal manometry demonstrated a normal upper esophageal sphincter pressure, a LES pressure of 61.9 mm

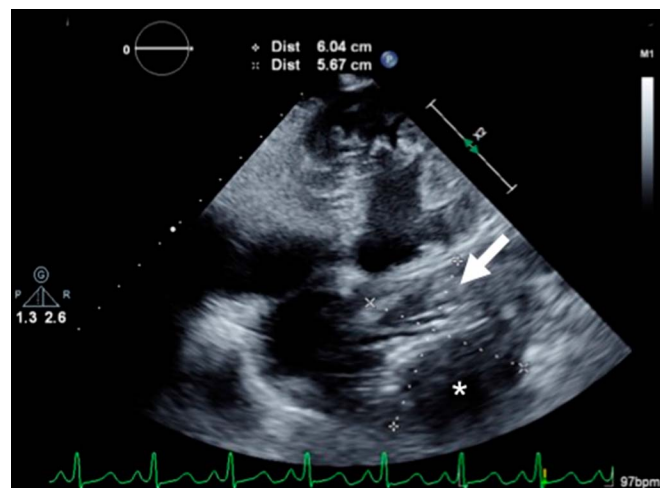


Figure 3. Echocardiogram showing the compressed left atrium (arrow) and dilated esophagus (*).

Hg, an integrated relaxation pressure of 50.3 mm Hg, with no motility or panesophageal pressurization (Figure 5). Using the Chicago 3 Criteria, these findings were most consistent with type 1 achalasia.⁴ A double-contrast esophagram was also performed, with findings consistent with a sigmoid-shaped megaesophagus (Figure 6). The patient was treated with pneumatic balloon dilation of the distal esophagus and GE junction to 30 mm. Pneumatic dilation was the treatment of choice as the patient declined surgery, and the presence of severe esophagitis and her sigmoid esophagus were relative contraindications to peroral endoscopic myotomy. The patient tolerated the procedure well with improvement in her dysphagia, with results sustained at a 3-month follow-up visit.

DISCUSSION

Achalasia is characterized by impaired relaxation of the lower esophageal in the setting of absent peristalsis, classically presenting with dysphagia to solids and liquid. We report dyspnea and hemodynamic instability as a unique presentation of achalasia. Respiratory symptoms are relatively common in achalasia, with up to 40% of patients reporting daily respiratory symptoms, including cough, aspiration, hoarseness, wheezing, and less commonly, shortness of breath.⁵ Most respiratory

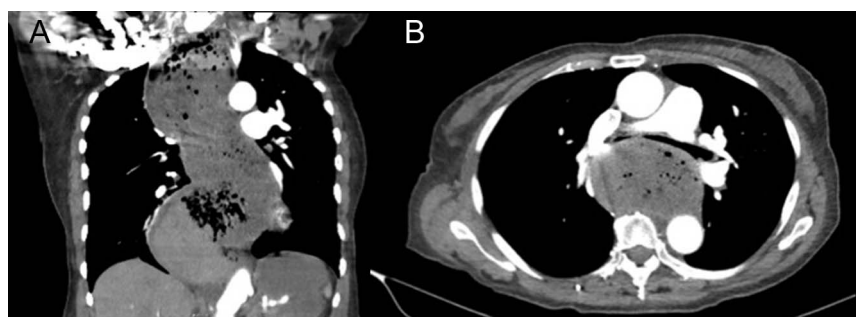


Figure 2. Computed tomography showing (A) the dilated and fluid-filled esophagus and (B) compression of the right bronchus.

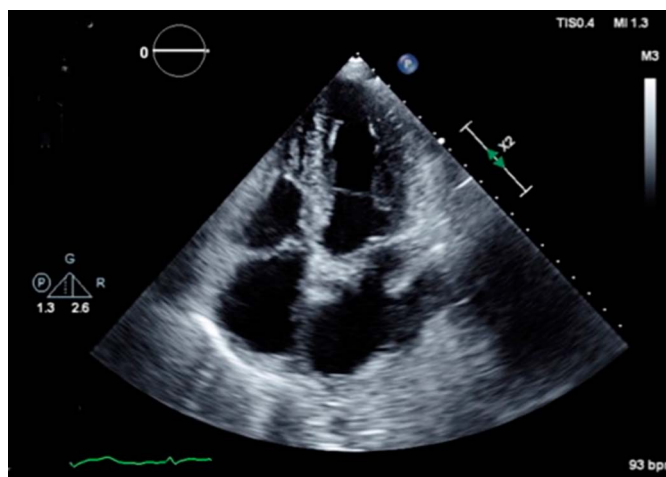


Figure 4. Echocardiogram postesophagogastroduodenoscopy showing resolution of left atrial compression.

symptoms are believed to be secondary to regurgitation and aspiration in the setting of dysphagia. However, respiratory failure is an uncommon complication, with about 50 reported cases since 1950, with a few cases severe enough to require ventilator support. Several of these cases were because of extrinsic compression of the trachea or mainstem bronchi, as seen in our patient.^{6–8} In our patient, the degree of extrinsic compression on

the trachea and mainstem bronchi resulted in both obstruction and decreased aeration, resulting in hypercapnia and hypoxia. Moreover, left atrial compression by a dilated esophagus is rare, with 7 reported cases. Left atrial compression causes reduced left ventricle volume leading to decreased cardiac output and hemodynamic instability. It also has been reported to cause pulmonary edema mimicking heart failure.^{9,10}

As demonstrated in our patient, end-stage achalasia with an enlarged thoracic esophagus can present as an acute illness because of extrinsic compression of nearby structures—namely, the trachea, bronchial tree, and the left atrium—resulting in respiratory failure and severe hemodynamic compromise. Although a nasogastric tube was not placed in our patient, an aspiration nasogastric tube can be used for esophageal decompression as an emergency measure. Endoscopic decompression of the esophagus alleviates the extrinsic compression on the lungs and heart, restoring respiratory and circulatory function.

DISCLOSURES

Author contributions: H. Blaney wrote the manuscript. N. Agarwal, S. Ashfaq, and C. Naumann edited and reviewed the manuscript. C. Naumann is the article guarantor.

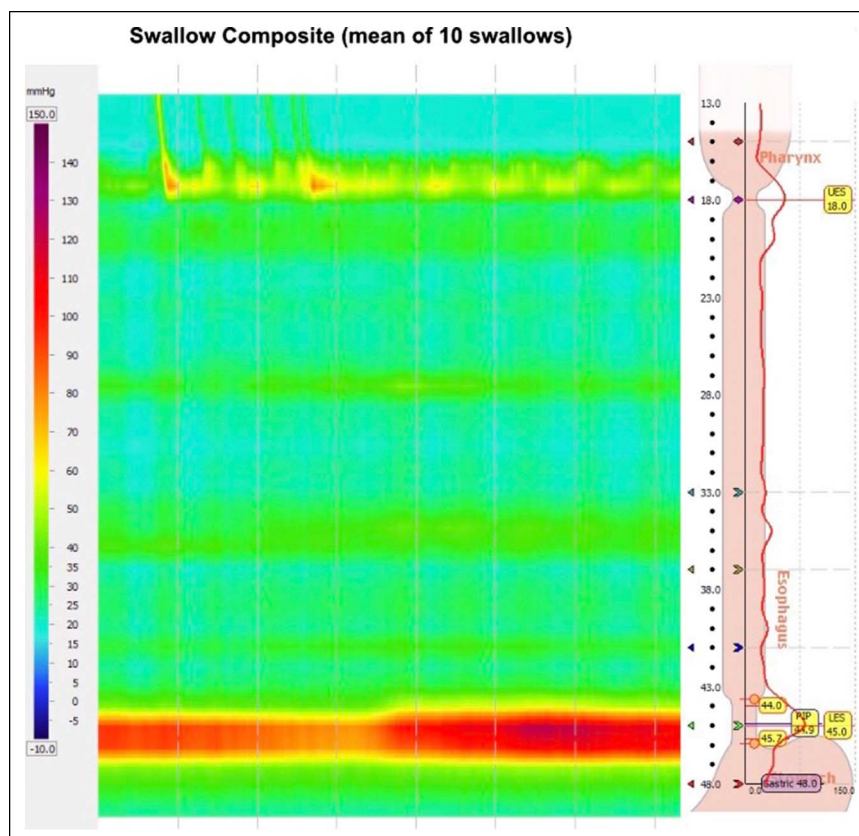


Figure 5. Medtronic high-resolution manometry consistent with type 1 achalasia. LES, lower esophageal sphincter.

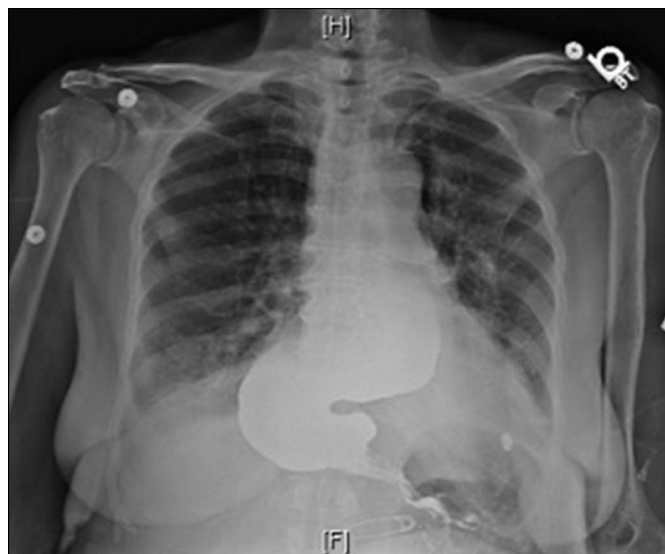


Figure 6. Double-contrast esophagram with achalasia demonstrating a sigmoid-shaped esophagus with a severely dilated lumen and a classic bird-beak appearance at the lower esophageal sphincter.

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Informed consent was obtained for this case report.

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