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Acute Respiratory Failure Secondary to Achalasia

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In Brief

A patient with untreated achalasia experienced massive dilatation of the esophagus causing airway compression at multiple sites. Acute ventilatory failure ensued, requiring endotracheal intubation. Emergent bronchoscopic intervention was used to overcome distal tracheal compression and thereby enable safe evacuation of esophageal contents by esophagoscopy.

Figure 1. Fiberoptic bronchoscopy demonstrated extrinsic compression of the trachea causing almost complete tracheal occlusion (A) and compression of the right and left mainstem bronchi (B).

The Clinical Challenge

A 47-year-old woman with presumed achalasia was admitted to an outside hospital with shortness of breath, weight loss, and inability to tolerate oral intake. Chest computed tomography (CT) demonstrated a massively dilated esophagus with anterior displacement and compression of the trachea and mainstem bronchi. Two days after admission, respiratory distress developed while the patient was eating. She was endotracheally intubated, mechanically ventilated, and transferred to our hospital.

The woman was extubated uneventfully shortly after arrival. Esophagoscopy showed that the esophagus was densely packed with food. A liquid diet was recommended; however, she continued to eat solid food. Several days later, the patient again developed respiratory distress. Physical examination demonstrated audible breathing and diffuse neck enlargement. On direct laryngoscopy, only the

Figure 2. Chest radiograph demonstrated widened mediastinum.

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epiglottis could be visualized. An endotracheal tube was placed; however, ventilation was nearly impossible despite manipulation of the endotracheal tube and suctioning. Analysis of an arterial blood sample obtained while the patient breathed 100% oxygen demonstrated a pH of 6.86, a PCO_2 of 193 mm Hg, and a PO_2 of 87 mm Hg.

Emergent bronchoscopy revealed severe extrinsic compression of the trachea and mainstem bronchi (Figure 1), causing occlusion of the trachea below the level of the endotracheal tube. The endotracheal tube was advanced beyond the zone of tracheal compression, resulting in normalization of airway pressures and gas exchange. Several hours later, oxygen desaturation occurred and was attributed to right mainstem intubation. Bronchoscopy was again performed, and the tip of endotracheal tube was placed immediately above the carina while the neck was in full flexion. A chest radiograph and CT imaging of the chest showed mediastinal widening and massive esophageal dilatation with compression of both mainstem bronchi (Figures 2 and 3).

A nasogastric tube was placed into the esophagus, but the esophageal contents could not be removed by suctioning. Repeat endoscopy was performed with removal of 1 L of partially liquid contents from the esophagus. After this procedure, the neck enlargement resolved, and the patient was successfully extubated. Achalasia was confirmed by barium swallow (Figure 4) and esophageal manometry. After botulinum toxin injection into the lower esophageal sphincter, the esophagus remained relatively decompressed (Figure 5). She was able to safely tolerate an oral liquid diet and was discharged from the hospital.

Discussion

Achalasia occasionally causes airway compression due to massive dilatation of the esophagus, termed mega-esophagus. Symptoms of central airway compression typically worsen after eating. Neck swelling may be present as well as anterior displacement of the larynx, which can complicate direct

laryngoscopy. Despite these characteristic symptoms and signs, the respiratory complications of mega-esophagus are sometimes misdiagnosed initially as asthma.

When mega-esophagus causes respiratory compromise, a nasogastric tube may be inserted into the proximal esophagus to reduce the volume of

retained food material and thereby reduce airway compression. Other temporizing measures have included the inhalation of a helium-oxygen gas mixture (Heliox) and the administration of sublingual nitroglycerin with the intention of reducing lower esophageal sphincter tension.

Figure 3. Chest computed tomography demonstrated massively dilated esophagus causing anterior displacement and compression of the larynx (A), trachea (B), and mainstem bronchi, especially on the right side (C).

Figure 4. Barium esophagram demonstrated long-standing achalasia with a massively dilated “sigmoidized” esophagus tapering into a classic “bird’s beak.”

Figure 5. Chest radiograph after treatment demonstrates improvement in mediastinal widening.

Interventional treatment for achalasia is aimed at opening the lower esophageal sphincter. Options include mechanical dilatation or injection of botulinum toxin into the lower esophageal sphincter or surgical cleavage of the sphincter muscle (Heller myotomy). In some cases, respiratory compromise occurs even after standard treatment and may be due to failure of the upper esophageal sphincter to relax in response to esophageal distension. This functional defect is sometimes called a “defective belch reflex.” In such cases, a cricopharyngeal myotomy may be performed to open the upper sphincter.

By one of these approaches or another, most reported cases of respiratory compromise caused by mega-esophagus have been managed without endotracheal intubation. In our case, ventilation was severely compromised by distal tracheal compression. Emergent bronchoscopy was required to precisely position the endotracheal tube.

As illustrated by our patient, achalasia can cause respiratory failure due to airway compression at any of several sites from the glottis to the main bronchi. Emergency management depends on rapidly identifying the site (s) of obstruction by CT imaging or other means. Nasogastric tube insertion may be sufficient to overcome upper airway obstruction. Distal airway obstruction may require fiberoptic intubation and bronchoscopy before suction decompression of the esophagus and definitive interventional management.

In cases of immediately life-threatening respiratory failure, early bronchoscopy and continued adjustment of endotracheal tube placement may be needed to optimize ventilation pending definitive treatment. ■

Author disclosures are available with the text of this article at www.atsjournals.org.

Recommended Reading

Ali GN, Hunt DR, Jorgensen JO, deCarle DJ, Cook IJ. Esophageal achalasia and coexistent upper esophageal sphincter relaxation disorder

presenting with airway obstruction. *Gastroenterology* 1995;109:1328–1332.

Aslam MI, Sutton CD, Hunter DC. Acute airway compression with achalasia. *J Am Coll Surg* 2007;205:813.

Becker DJ, Castell DO. Acute airway obstruction in achalasia: possible role of defective belch reflex. *Gastroenterology* 1989;97:1323–1326.

Bello CT, Lewin JR, Norris CM, Farrar GE Jr. Achalasia (cardiospasm): report of a case with extreme and unusual manifestations. *Ann Intern Med* 1950;32:1184–1190.

Dunlop SP, Travis SPL. Achalasia presenting as acute stridor. *Eur J Gastroenterol Hepatol* 1997;9:1125–1128.

Miyamoto S, Konda Y, Matsui M, Sawada K, Ikeda K, Watanabe N, Kawanami C, Chiba T. Acute airway obstruction in a patient with achalasia. *Intern Med* 2011;50:2333–2336.

Wagh MS, Matloff DS, Carr-Locke DL. Life-threatening acute airway obstruction in achalasia. *MedGenMed* 2004;6:12.

Requena R, Hart AK, Kuhn JJ. Acute upper airway obstruction in achalasia. *Otolaryngol Head Neck Surg* 1999;120:105–108.

Walzer N, Hirano I. Achalasia. *Gastroenterol Clin North Am* 2008;37:807–825, viii.

Westbrook JL. Oesophageal achalasia causing respiratory obstruction. *Anaesthesia* 1992;47:38–40.