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Seminars in
ULTRASOUND
CT and MRI

Achalasia With Tortuous Megaesophagus

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Case report

A 45-year-old male presented with a 1-year history of severe dysphagia, reflux, and malnutrition with an unintentional weight loss of 60 pounds. Posteroanterior and lateral radiographs of the chest (Figs. 1 and 2) revealed a markedly dilated, redundant, and torturous esophagus (arrows in Fig. 1; asterisks in Fig. 2-5). The diagnosis of achalasia was confirmed by upper endoscopy, which showed complete esophageal aperistalsis, with a tight, nonreactive esophagogastric junction. The patient failed initial laparoscopic myotomy, requiring a feeding gastrojejunostomy tube. One year later, his symptoms persisted, and preoperative computed tomography demonstrated a markedly redundant and tortuous megaesophagus, with proximal air-fluid levels denoting distal obstruction (Figs. 3-5). The dilated esophagus compressed and shifted the trachea (short arrows in Figs. 3 and 4). Multiplanar CT reconstruction showed the entire course of the megaesophagus, with a narrowed esophagogastric junction demonstrating the classical "bird's beak" appearance of achalasia (arrowheads, Fig. 5). The patient subsequently underwent definitive surgical management with a McKeown esophagectomy, from which he is currently recovering.

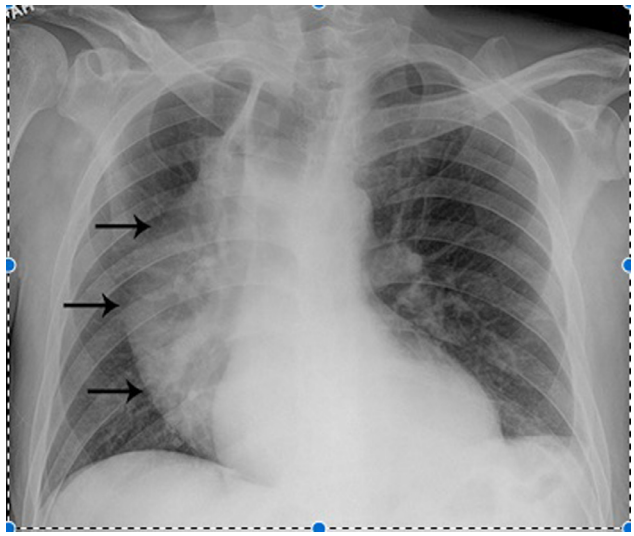

Figure 1 Posteroanterior radiograph of the chest x-ray shows a widened mediastinum, with a well-defined opacity extending to the right hemithorax (arrows), suggestive of a dilated esophagus.

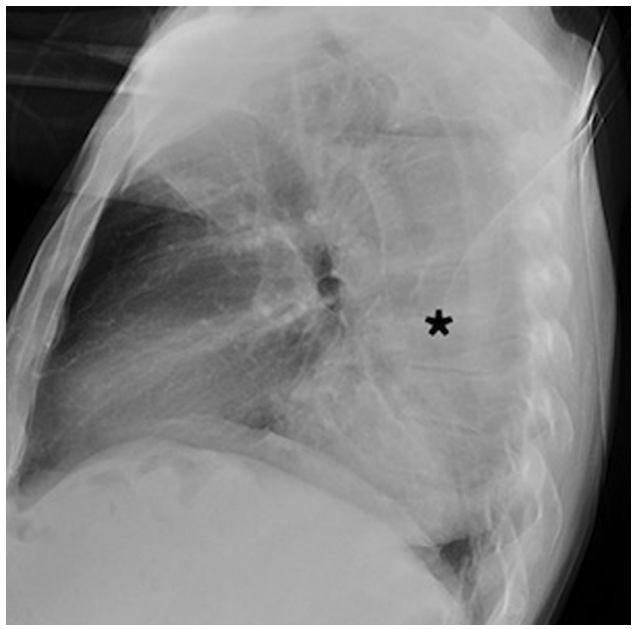

Figure 2 Lateral chest x-ray shows an increased density along the posterior mediastinum (asterisk), suggestive of a dilated esophagus.

Discussion

Achalasia is derived from the Greek word "khalasis," translated as "not loosening or relaxing."[1] Achalasia is a rare esophageal motility disorder that is usually idiopathic in origin. It affects equally males and females, without racial predilection, and across all ages, with a peak incidence between the ages of 30 and 60.[2] Achalasia is characterized by incomplete relaxation of the lower esophageal sphincter (LES) on swallowing and aperistalsis of the esophageal body.[1-4]

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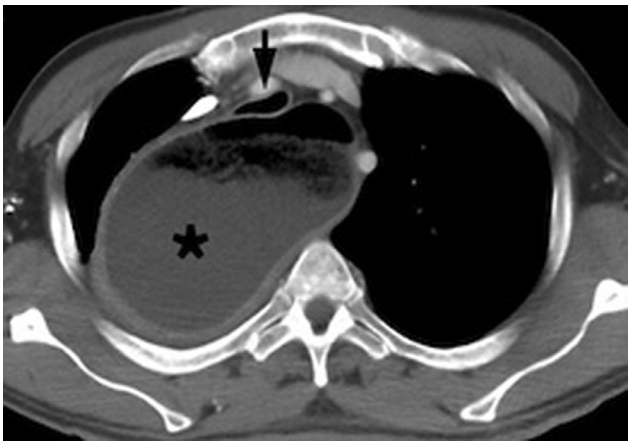

Figure 3 Axial CT demonstrates a markedly dilated and tortuous megaesophagus (asterisk), with proximal air-fluid level denoting distal obstruction. The dilated esophagus compresses and shifts the trachea anteriorly and to the right (short arrow).

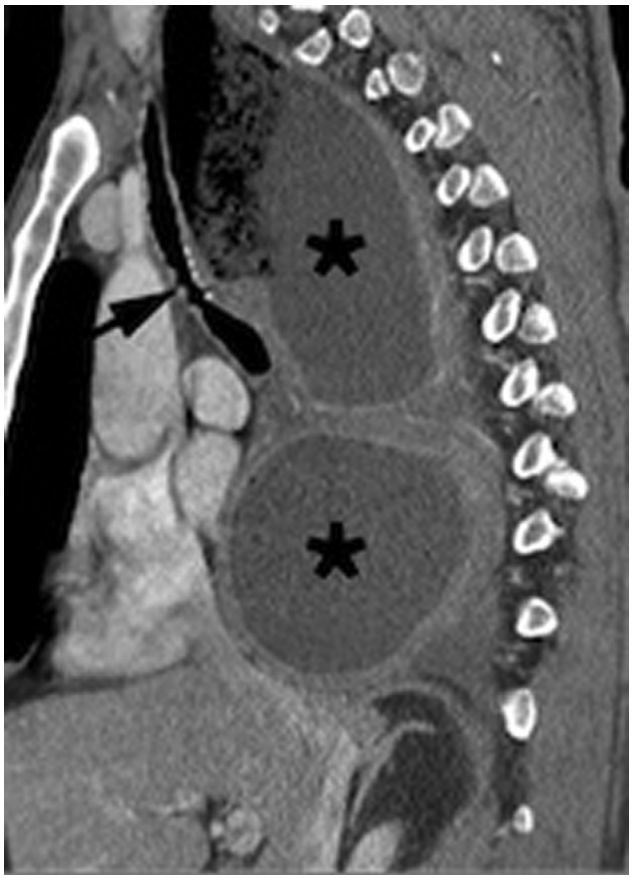

Figure 4 Sagittal CT demonstrates a markedly dilated and tortuous megaesophagus (asterisks), with proximal air-fluid level denoting distal obstruction. The dilated esophagus compresses and shifts the trachea anteriorly and to the right (short arrow).

Diagnosis can be difficult to establish, and many patients have symptoms for many years prior to correct diagnosis and treatment. The condition is characterized by dysphagia, heartburn, regurgitation or vomiting, and noncardiac chest pain. Associated signs and symptoms include cough or asthma,

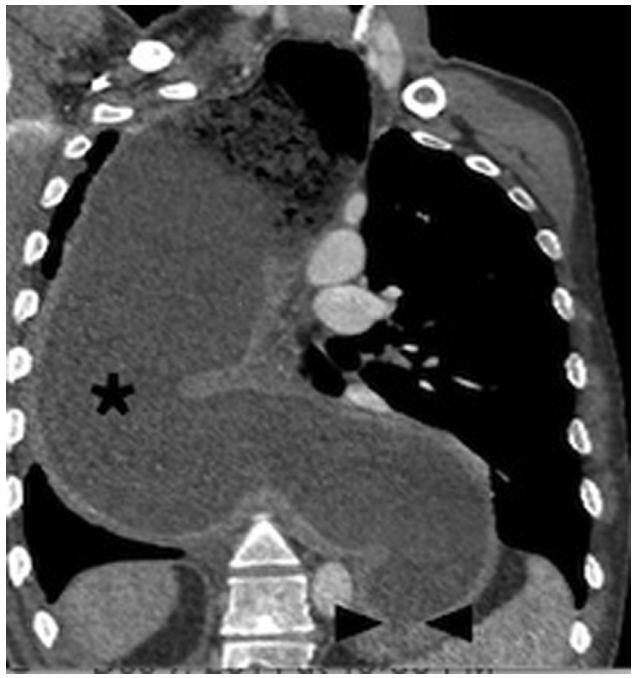

Figure 5 Multiplanar reformat CT demonstrates a markedly dilated and tortuous megaesophagus (asterisk), with proximal air-fluid level denoting distal obstruction. The dilated esophagus compresses and shifts the trachea anteriorly and to the right. The narrowed esophagogastric junction shows the classical "bird's beak", typical appearance of achalasia (arrowheads).

chronic aspiration, hoarseness or sore throat, and unintentional weight loss.[1,3,4] The diagnosis of achalasia should be considered after an endoscopy in patients with these symptoms does not reveal a mechanical obstruction or an inflammatory cause of the esophageal symptoms.[1-4] Manometry should then be performed if achalasia is suspected. Esophageal manometry assesses esophageal pressures and contractions and has become the standard for diagnosing and classifying achalasia. The 2 classic manometric findings in the diagnosis of achalasia are aperistalsis of the esophageal body and impaired relaxation of the LES during swallowing.[2]

Barium esophagogram is a helpful modality to establish the diagnosis. Classically, peristalsis is absent, and the distal esophagus usually tapers to a characteristic narrowing, or so-called "bird's beak." This narrowing represents the upper margin of the LES which fails to relax normally. Emptying of barium from the esophagus is poor.[2]

In advanced cases, CT shows uniform dilatation which affects a long segment of the esophagus, without wall thickening and with normal-appearing boundary surfaces and mediastinal fat. The esophagus narrows abruptly at the esophagogastric junction without evidence of an intramural or extrinsic obstructive lesion. The esophageal wall at the site of the narrowing appears normal, in contrast to the focal thickening present in cases of esophageal tumor or esophagitis.[2,3,5]

Most cases of achalasia can be managed conservatively with graded endoscopic pneumatic dilation or botulinum toxin injections to the stenotic gastroesophageal junction, which aim at weakening or ablating the LES. Surgical myotomy and

fundoplication are performed if these treatment options are refractory. Esophagectomy is ultimately reserved as a final option in patients with severe esophageal dilatation, reflux strictures, or failed prior esophagomyotomy.[1,2,4-6]

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