Dysphagia Aortica from Endovascular Leak After Thoracic Endovascular Aneurysm Repair

by Sonal Gandhi, Malorie K. Simons, Joshua A. Sloan

Achalasia is an esophageal motility disorder that causes peristaltic dysfunction and failure of the lower esophageal sphincter (LES) to relax upon swallowing. Pseudoachalasia is a mimicker of achalasia, but its pathophysiology and treatments are different. We present a case of an elderly woman with dysphagia. Manometry was most consistent with type II achalasia. Ultimately, the diagnosis of dysphagia aortica causing pseudoachalasia from a large, compressing thoracic aorta endoleak after a thoracic endovascular aneurysm repair (TEVAR) was made. This diagnosis altered her management course, avoiding possible intervention such as peroral endoscopic myotomy (POEM) versus laparoscopic Heller myotomy (LHM).

INTRODUCTION

Achalasia is a primary esophageal neuromuscular motility disorder of unclear etiology.[1] It results from degeneration of the myenteric plexus causing failure of the lower esophageal sphincter (LES) to relax upon swallowing with esophageal body dysmotility (absent contractility, panesophageal pressurization, or spastic contractions).[1,2] In order to make the diagnosis of achalasia, it is crucial to rule out other mimickers including medication effect and pseudoachalasia in the appropriate patients.[3] Dysphagia aortica (DA) is a rare cause of pseudoachalasia. First described in 1932 DA is due to external compression of the esophagus by ectatic, tortuous or aneurysmal thoracic aorta due to age related degeneration.[4] It is mainly seen in elderly women of short stature and with co-existing conditions such as hypertension and kyphosis. We present a case of an elderly woman with manometric findings consistent with Type II achalasia who was transferred to our center for peroral endoscopic myotomy (POEM). After careful review of clinical history, imaging and manometry, her symptoms were more consistent with DA from a large, compressing thoracic aorta, resulting in pseudoachalasia.

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Case Report

A 75-year-old woman with a history of anti-Jo1 antisynthetase syndrome, steroid dependent interstitial lung disease on home oxygen, chronic pain on long-term opiates, severe pulmonary hypertension, and thoracoabdominal aortic aneurysm status post total thoracic endovascular aneurysm repair (TEVAR) three years prior with recent redo presented with six months of worsening dysphagia to solids and progressing to liquids. This resulted in weight loss, failure to thrive, and recurrent aspiration with declining lung function.

Diagnostic work up included computed tomography (CT) scan which revealed active type II endoleak extending from the TEVAR with lateral compression and displacement of the esophagus (Figure A). Barium esophagram was subsequently performed that showed a delay in esophageal emptying with retrograde flow and evidence of a short segment narrowing at the lower esophageal sphincter (LES) where the TEVAR intersects the LES (Figure B). Upper endoscopy demonstrated esophageal candidiasis without evidence of stenosis, external compression, or tight LES. An endoscopically placed manometry catheter showed no normal peristalsis, panesophageal pressurization in 5/10 swallows, and an integrated

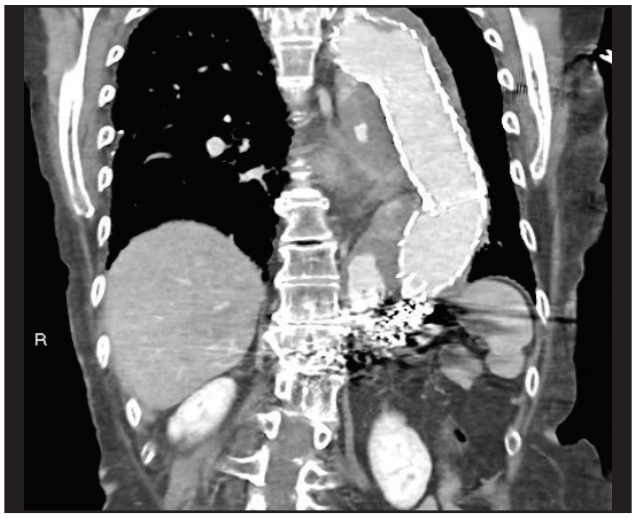

Figure A. Computed tomography (CT) scan of chest revealing active type II endoleak extending from the TEVAR with lateral compression and displacement of the esophagus.

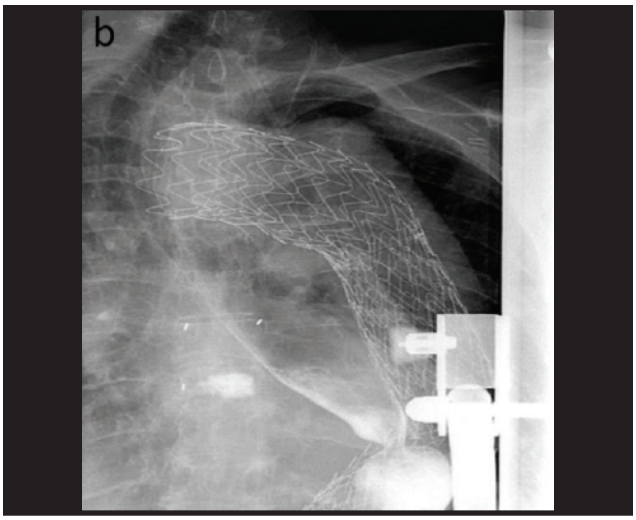

Figure B. Barium esophagram showing a delay in esophageal emptying with retrograde flow and evidence of a short segment narrowing at the LES where the TEVAR intersects the LES.

relaxation pressure (IRP) of 25.4 (Figure C). After a multidisciplinary discussion with the therapeutic endoscopists, motility and neurogastroenterology specialists, and vascular surgeons, her clinical presentation was deemed most consistent with pseudoachalasia due to endovascular leak causing compression as opposed to primary achalasia, thus limiting the utility of therapy directed towards achalasia. Additionally, the risk of endovascular repair was felt to outweigh benefits.

The patient opted for conservative management of her dysphagia, which improved with treatment for esophageal candidiasis and dietary modification to liquids, and pureed foods. After a lengthy hospital stay, she wished to focus her attention on comfort, and she was discharged home with hospice care.

Discussion

Pseudoachalasia is a diagnostic entity that is indistinguishable from primary achalasia. Most reports are from a malignant paraneoplastic effect, or circumferential compression and infiltration of the LES.[5,6] DA, itself, is an uncommon cause of dysphagia due to compression of the LES; however, DA resulting in pseudoachalasia is exceedingly rare. To date, there is only one case report of a patient initially thought to have achalasia but then found to have DA from a thoracic aortic aneurysm causing pseudoachalasia.[5] To our knowledge, we believe we present the first reported case of a TEVAR endoleak causing pseudoachalasia.

This case posed a significant diagnostic challenge. Manometrically the patient's diagnosis was that of type II achalasia. With the advent of high resolution manometry (HRM), achalasia can be separated into three subtypes (Type I, II and III) based upon the esophageal pressure topography and the Chicago Classification.[1,2] Diagnosing the correct achalasia phenotype has important prognostic and therapeutic implications. For example, Type II achalasia, defined by absent peristalsis, an elevated IRP and panesophageal pressurization in a minimum of 2/10 reclining swallows, responds to pneumatic dilation, POEM, or Heller myotomy.[3]

Our patient was a high risk surgical or POEM candidate due to her known cardiopulmonary disease, chronic steroids use, and position of the active endovascular leak near the LES; however, there were multiple factors that made her achalasia diagnosis questionable. First, opiates have been shown to cause dysphagia and manometry findings comparable to achalasia.[7] Once opiates are stopped, esophageal motility returns to normal. For our patient, ceasing all opiate use would have resulted in significant morbidity and thus was a last resort. Secondly, her history was more consistent with pseudoachalasia: sudden onset of symptoms, marked weight loss, and an esophageal compression at the site of the endovascular leak.

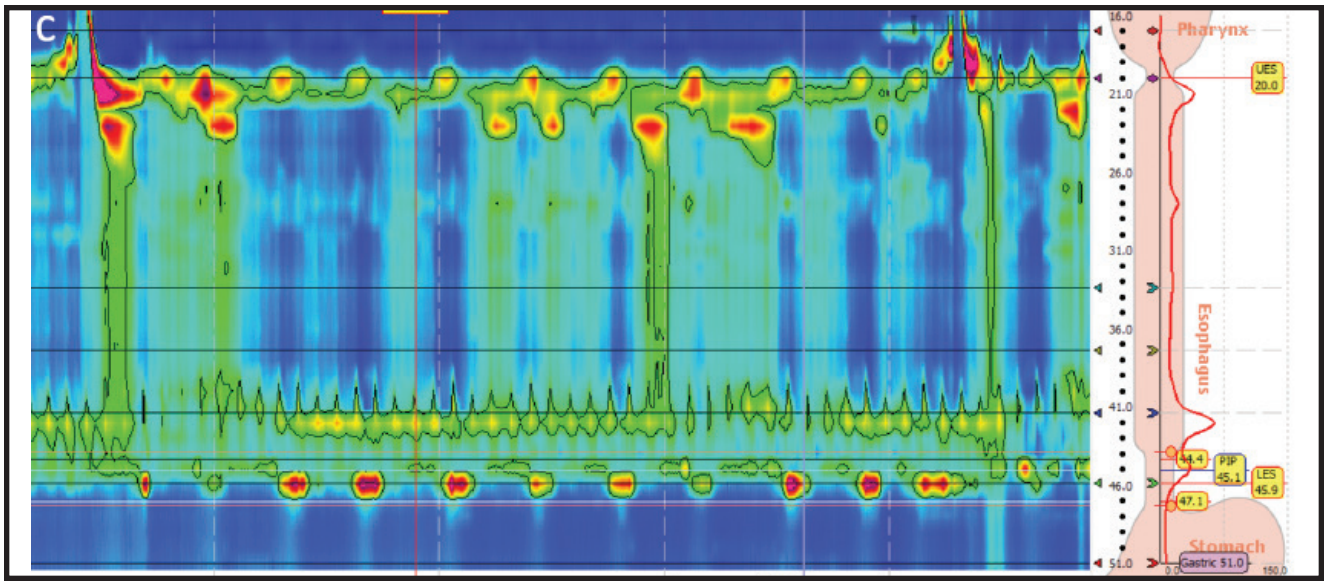

Figure C. Endoscopic manometry catheter showed no normal peristalsis, panesophageal pressurization in 5/10 swallows, and an Integrated Relaxation Pressure (IRP) of 25.[4]

DA is a rare cause of pseudoachalasia, and thus little is known about the underlying pathophysiology. This external compression may result in esophageal dysmotility based on the anatomic impact of the vasculature on the esophagus.[8] In our patient it was the compression by the TEVAR and endoleak that was the most likely cause of her DA and subsequent pseudoachalasia. There was the additional confounder of chronic opioid use that cannot be ignored as a possible contributing factor; however, this would not account for the radiographic findings.

This case highlighted the importance of distinguishing pseudoachalasia from primary achalasia, as treatment options are different. Treatment for pseudoachalasia is aimed at the primary cause. In our case, this would be directed towards weaning off opiates and endovascular repair, for which the latter was deemed to be high risk with uncertain benefit. Achalasia is managed taking into account a number of factors including sex, age, subtype of achalasia, and patient preference with the management choices being laparoscopic Heller myotomy, pneumatic dilation, hydrostatic dilation, POEM, or botulinum toxin injection. To help differentiate pseudoachalasia from true achalasia, amyl nitrate could have been administered during manometry, resulting in LES relaxation in the latter. Unfortunately, this diagnostic option was considered too dangerous for our patient given her severe cardiopulmonary disease. This case highlights the importance of a thorough review of patients' history and clinical presentation in combination with a multidisciplinary approach to decipher the true cause of dysphagia to most appropriately manage complex esophageal disease. ■

References

1. Pandolfino JE, Gawron AJ. Achalasia: a systematic review. Jama. 2015;313(18):1841-1852.
2. Kahrilas PJ, Bredenoord AJ, Fox M, et al. The Chicago Classification of esophageal motility disorders, v3.0. Neurogastroenterol Motil. 2015;27(2):160-174.
3. Khashab MA, Vela MF, Thosani N, et al. ASGE guideline on the management of achalasia. Gastrointest Endosc. 2019.
4. Wilkinson JM, Euinton HA, Smith LF, Bull MJ, Thorpe JA. Diagnostic dilemmas in dysphagia aortica. Eur J Cardiothorac Surg. 1997;11(2):222-227.
5. Beqari J, Lembo A, Critchlow J, Hamden A, Kent MS. Pseudoachalasia Secondary to Thoracic Aortic Aneurysm. Ann Thorac Surg. 2017;103(6):e517-e518.
6. Gockel I, Eckardt VF, Schmitt T, Junginger T. Pseudoachalasia: a case series and analysis of the literature. Scand J Gastroenterol. 2005;40(4):378-385.
7. Ratuapli SK, Crowell MD, DiBaise JK, et al. Opioid-Induced Esophageal Dysfunction (OIED) in Patients on Chronic Opioids. Am J Gastroenterol. 2015;110(7):979-984.
8. Mucklow EH, Smith OE. Dysphagia and unusual radiographic appearances associated with the variable relationships of the aorta and lower oesophagus. J Fac Radiol. 1954;6(2):88-95.