ACG CASE REPORTS JOURNAL

CASE REPORT | ESOPHAGUS

Ineffective Esophageal Motility Progressing into Distal Esophageal Spasm and Then Type III Achalasia

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ABSTRACT

The clinical significance of minor esophageal motility disorders is unclear, though they typically carry a benign course. Distal esophageal spasm progressing to achalasia has been reported, although it appears to be rare. We report a case of a patient with dysphagia and chest pain who was found to have ineffective esophageal motility on high-resolution manometry, which developed into distal esophageal spasm and then progressed to type III achalasia.

INTRODUCTION

The introduction of high-resolution manometry (HRM) has enhanced our ability to characterize esophageal motility disorders. Some symptomatic patients evaluated with HRM demonstrate motility patterns that are also observed in asymptomatic controls. These esophageal motility diagnoses, such as ineffective esophageal motility (IEM), are considered minor esophageal motility disorders and their clinical significance remains unclear, although they typically represent a good prognostic indicator with a benign clinical course.[1,2] Distal esophageal spasm (DES) is a rare, major esophageal motility disorder (ie, not observed in asymptomatic controls) that is thought to involve dysfunction of inhibitory innervation. While DES may share pathophysiological features with achalasia, the progression from DES to achalasia appears to be rare.

CASE REPORT

A 58-year-old male was referred to our esophageal clinic in 2007 for dysphagia and chest pain. He reported a history of dysphagia for solid foods and occasionally for liquids for many years, but denied heartburn, regurgitation, nausea, vomiting, anorexia, or weight loss. He was previously placed on lansoprazole for his dysphagia without significant improvement. His medications were lansoprazole, bupropion, simvastatin, and niacin. Esophagogastroduodenoscopy (EGD) showed some tortuosity in the esophagus, and esophageal HRM was consistent with IEM based on Chicago Classification v3.0, with a median integrated relaxation pressure (IRP) of 5.5 mm Hg, the shortest distal latency (DL) among the 10 test swallows of 9.1 s, and 7 of 10 ineffective swallows (distal contractile integral $<$450 mm Hg·s·cm); the remaining 3 supine swallows were normal (Figure 1).[1] Gastroesophageal reflux disease was suspected as the cause of his symptoms, and his proton pump inhibitor therapy was intensified.

The patient returned in 2012 with worsening dysphagia, occurring with every meal and occasionally for liquids. EGD showed a corkscrew configuration in the middle and distal thirds of the esophagus, with a slight resistance to passage through the lower esophageal sphincter (LES) (Figure 2). HRM was consistent with DES (DL $<$4.5 s in 10 of 10 swallows, median IRP of 12.5 mm Hg), and he was placed on sublingual nitroglycerin on as-needed basis

ACG Case Rep J 2016;3(4):e183. doi:10.14309/crj.2016.156. Published online: December 21, 2016.

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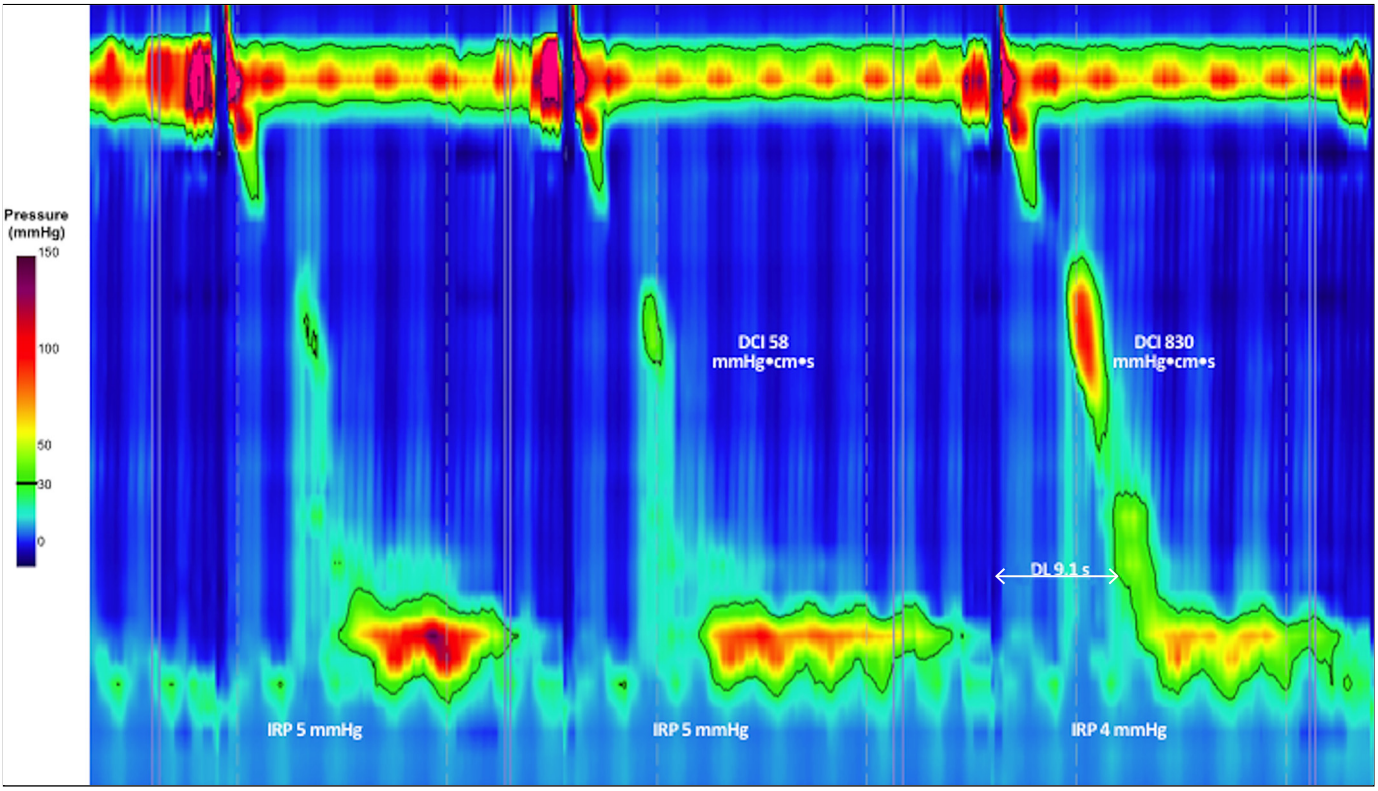

Figure 1. High-resolution manometry consistent with ineffective esophageal motility. Two failed swallows and one normal swallow are included. IRP = integrated relaxation pressure; DL = distal latency; DCI = distal contractile integral.

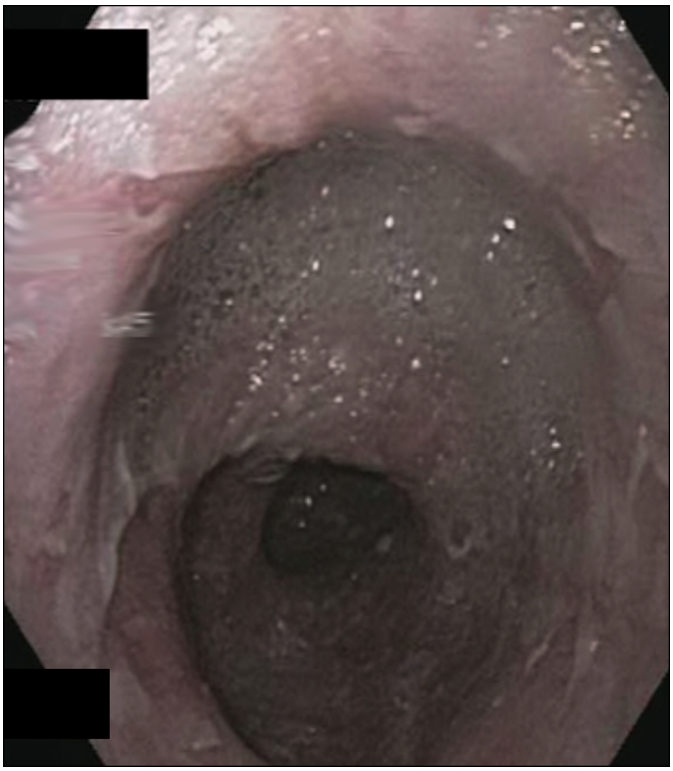

Figure 2. Upper endoscopy shows corkscrew esophagus.

(Figure 3). In 2014, he returned with chest pain and progressive dysphagia for both solids and liquids and occasionally with saliva, in addition to intermittent heartburn and regurgitation. EGD again showed a corkscrew configuration, and passage of the endoscope into the stomach was associated with a "pop" sensation. Timed barium esophagram showed a corkscrew appearance of the middle and distal esophagus with esophagogastric junction narrowing and a residual contrast column at 5 minutes (Figure 4). A repeat HRM was consistent with type III achalasia with a median IRP of 22 mm Hg and DL <4.5 s in 7 of 10 swallows (Figure 5). The patient underwent peroral esophageal myotomy with an extended myotomy (up to 14 cm proximal of the squamocolumnar junction), with significant improvement in his symptoms. At his 6-month follow-up he reported only rare dysphagia and chest pain with complete esophageal bolus clearance by 1 minute on the timed barium esophagram.

DISCUSSION

IEM, defined as 50% of swallows being weak (distal contractile integral <450 mm Hg·s·cm) on HRM is a minor esophageal motility disorder characterized by weak esophageal contractions that may result in abnormal esophageal bolus clearance. A recent study that followed a cohort of symptomatic patients with minor esophageal motility disorders on HRM by

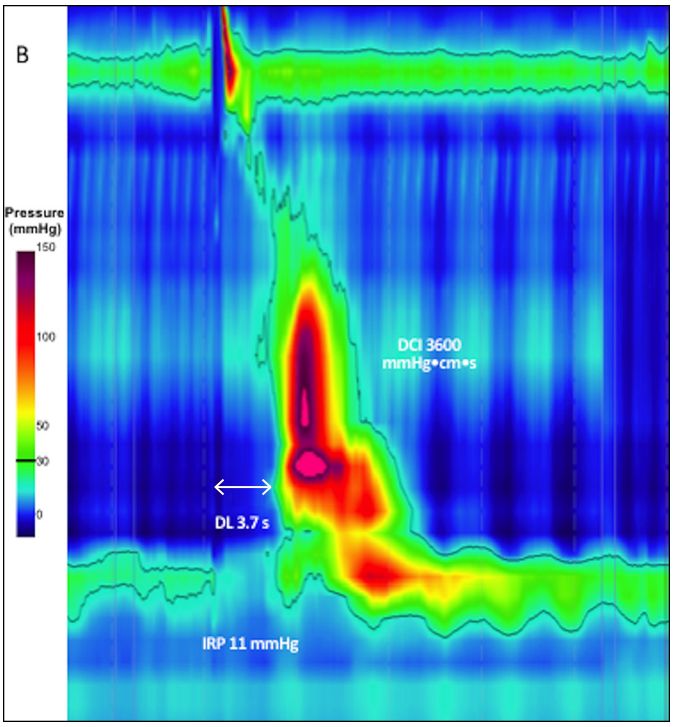

Figure 3. High-resolution manometry consistent with distal esophageal spasm (DL 3.7 s; IRP 11 mm Hg). IRP = integrated relaxation pressure; DL = distal latency.

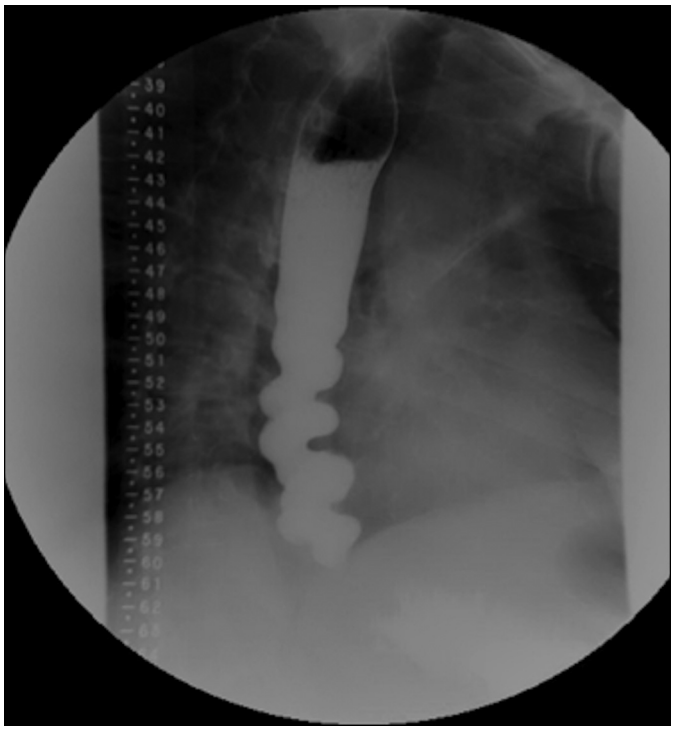

Figure 4. Timed barium esophagram shows corkscrew appearance of the middle and distal esophagus with esophagogastric junction narrowing and a residual contrast column at 5 minutes.

assessing their symptoms using a standardized questionnaire found that these patients were largely asymptomatic after a mean follow-up period of 6.4 years.[2] No follow-up manometry was performed in that cohort, yet the benign course of these disorders argues against major manometric changes. The HRM criteria for IEM reflects previous criteria established for conventional manometry, although IEM was also sometimes included within a heterogeneous spectrum of esophageal motility patterns identified with conventional manometry labeled non-specific esophageal motility disorders (NEMD).[3-5] Previous studies suggested that NEMD may rarely progress to DES or achalasia.[6,7] In another study, however, 54% (15 of 28 patients) with NEMD who underwent repeat conventional manometry progressed to achalasia after a follow-up period of 4 years.[8] These studies are significantly limited by the clinical heterogeneity of the NEMD classification, which also limits application to our case.

DES and achalasia are thought to share the pathophysiological feature of defective inhibitory neural signaling.[4,9] The notion, however, that DES could progress to achalasia remains a source of debate. Two prospective studies utilizing conventional manometry reported rates of progression from DES to achalasia of 8% (1 of 12 patients) and 14% (5 of 35 patients) over mean follow-up periods of 4.8 and 2.1 years, respectively.[10,11] Limitations of conventional manometry should be recognized when interpreting these studies. DES may have been erroneously diagnosed by either missed impaired deglutitive relaxation related to swallow-associated esophageal shortening (pseudo-relaxation) or by labeling pan-esophageal pressurization (a finding easily identified on HRM) as simultaneous contractions, the typical conventional manometry criterion for DES. The enhanced characterization of esophageal motility characteristics allowed with HRM over conventional manometry appears to improve the sensitivity of diagnosing achalasia and specificity of identifying esophageal spasm.[9,12,13] Beyond improving detection of LES pseudo-relaxation, HRM and its associated metric of LES relaxation, the IRP, provides an improved sensitivity to diagnose achalasia compared with conventional measures of nadir or residual LES pressure.[12,13] Further, measurement of DL provided by HRM is more specific for spastic esophageal motor disorders than the conventional measure of contractile velocity, which means that HRM may also improve the specificity for diagnosing spastic esophageal motility disorders.[9] Although additional investigation utilizing HRM are needed to re-examine the rate of evolution from DES to achalasia, another recent case report that applied HRM suggests that DES can progress to type III achalasia. This case, along with ours, supports the hypothesis that DES may be a stage along the continuum of esophageal inhibitory neuro-degeneration that may eventually develop to type III achalasia.[14]

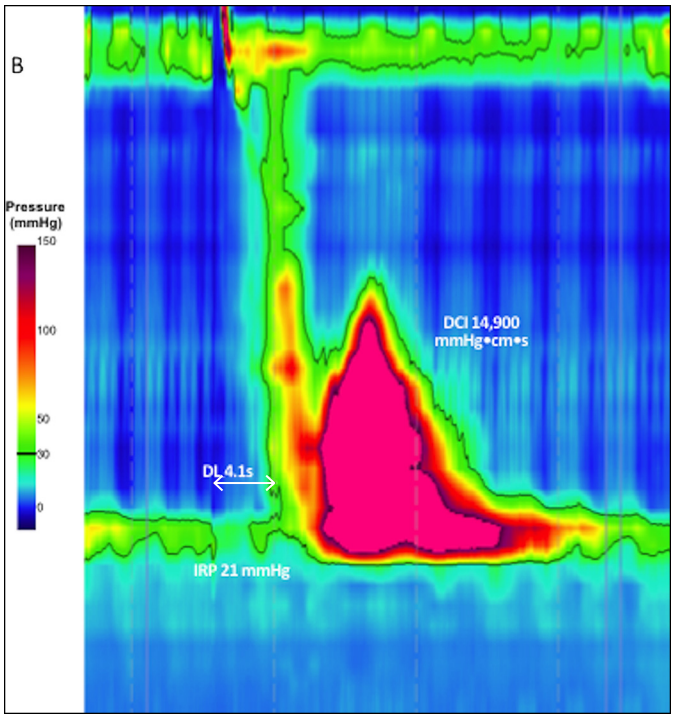

Figure 5. High-resolution manometry consistent with achalasia type III (IRP >15 mm Hg and DL <4.5 s). IRP = integrated relaxation pressure; DL = distal latency.

Although the vast majority of minor motility disorders carry a benign course, our case suggests that IEM may rarely evolve to a major motility disorder such as DES or achalasia on HRM. With this in mind, repeated motility testing in patients with progressive symptoms is warranted.

DISCLOSURES

Author contributions: S. Samo and DA Carlson wrote the manuscript and obtained the images. PJ Kahrilas and JE Pandolfino critically revised the manuscript. S. Samo is the article guarantor.

Financial disclosure: S. Samo, DA Carlson, and PJ Kahrilas have no disclosures to report. JE Pandolfino receives consulting or speaking fees from Medtronic, Sandhill Scientific, Takeda, and Astra Zeneca, and he has received grant funding from Medtronic. This work was supported by T32 DK101363 from the Public Health Service (JE Pandolfino).

Informed consent was obtained for this case report.

Received April 14, 2016; Accepted August 16, 2016

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