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CASE REPORT | ESOPHAGUS

The Role of Automatically Generated Chicago Classification in Delayed Achalasia Diagnosis

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ABSTRACT

Achalasia is an esophageal motility disorder characterized by a lack of peristalsis and an increased lower esophageal sphincter pressure that does not relax with swallowing. High-resolution manometry (HRM), a valuable diagnostic tool for esophageal disorders, often comes with software for automated study interpretation. Although helpful, there are certain caveats in the diagnostic criteria for achalasia which the software may miss. We highlight 2 patients in whom software analysis of HRM studies resulted in misdiagnosis of achalasia as manometrically detected nonachalasia esophagogastric junction outflow obstruction and emphasize the importance of manual interpretation of HRM data by clinicians.

INTRODUCTION

Esophageal achalasia is a primary esophageal motility disorder characterized by a lack of esophageal peristalsis and an increased lower esophageal sphincter pressure with impaired relaxation of the sphincter on swallowing.[1] Patients with esophageal achalasia generally present with dysphagia to liquids, solids, or both. Achalasia is categorized into 3 recognized subtypes based on manometric patterns: quiescent esophageal body (Type I), intermittent isobaric panesophageal pressurization (Type II), and premature or spastic distal esophageal contractions (Type III).[2] Unfortunately, under certain circumstances, achalasia across all 3 subtypes may be misdiagnosed as manometrically detected nonachalasia esophagogastric junction outflow obstruction (EJOO), a vague clinical entity that serves as a clinical dilemma for physicians and results in unnecessary testing with inadequate treatment.[2,3] We describe 2 such cases and highlight the importance of achalasia subtype classification via high-resolution manometry (HRM).

CASE REPORT

Patient 1: A 58-year-old woman with a medical history significant for hypertension presented as a referral for second opinion for her persistent spasm-like chest pain. The patient's discomfort was associated with dysphagia on consumption of liquids. She had previously undergone a motility study at an outside institution and was subsequently diagnosed with an outflow obstruction for which she underwent empiric esophageal dilation. The patient, however, did not experience symptomatic improvement. Reviewing the patient's previous motility report, it was noted that the HRM software marked the patient's swallow attempts as premature contractions with an average distal contractile integral (DCI) of 455 mm Hg, generating the diagnosis of outflow obstruction (Figure 1). However, on obtaining the actual report and conducting a reanalysis of the imaging, it was concluded that the patient displayed failed swallow attempts on HRM. The swallow attempts that were labeled premature were, in reality, failed swallow attempts because they were premature with a DCI less than 450 mm Hg based on the Chicago classification criteria. Based on the patient's symptoms, HRM, and radiological studies, she was diagnosed with Type I achalasia and referred for Heller myotomy. The patient subsequently experienced significant improvement in symptoms, roughly 2 years after her initial presentation to an outside facility.

Patient 2: A 66-year-old man with a medical history significant for hypercholesterolemia presented for evaluation after experiencing symptoms of dysphagia for liquids and solids for 2 years with associated significant weight loss, recurrent aspiration pneumonia, and failure to thrive. HRM performed 2 years earlier at an outside facility identified esophageal outflow obstruction based on a computer-generated HRM analysis which interpreted the patient's swallow attempts as premature contractions (Figure 2). However, on our analysis

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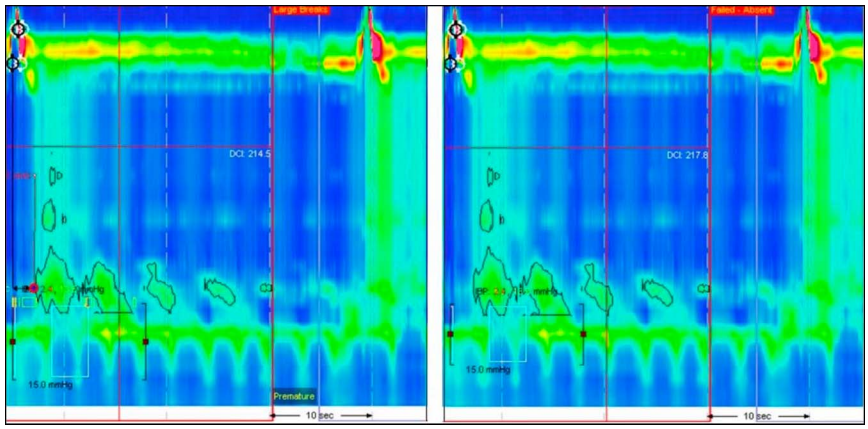

Figure 1. High-resolution manometry software marked the patient's swallow attempts as premature contractions with large breaks with an average DCI of 455 mm Hg (left). Manual analysis of the patient's high-resolution manometry identified failed swallow attempts with a DCI less than 450 mm Hg with an IRP greater than 15 mm Hg (right). DCI, distal contractile integral; IRP, integrated relaxation pressure.

of the study, panesophageal pressurization was identified on over 20% of swallow attempts and the swallow attempts were reclassified as failed with panesophageal pressurization. Based on the patient request, HRM was repeated and Type II achalasia was confirmed. The patient successfully underwent Heller myotomy with subsequent complete resolution of his symptoms. He was able to gain weight with restored quality of life.

DISCUSSION

Achalasia should be suspected in those with dysphagia to liquids and/or solids and in those with symptoms of regurgitation unresponsive to an adequate trial of proton-pump inhibitor therapy. The Chicago classification of esophageal motility, initially published in 2009 to help categorize esophageal motility disorders using HRM, has become an invaluable diagnostic aid. As per the latest version of the Chicago classification (V3.0), published in 2015, features of achalasia include a mean integrated relaxation pressure greater than or equal to 15 mm Hg (or upper limit of normal) and an absence of normal peristalsis on HRM.[2] All currently available HRM systems provide computer-generated study interpretations that, although helpful, are not infallible. The Chicago classification V3 system provides the criteria of each type of achalasia, with fine italic print about common errors which the HRM computer-generated reading in our cases fell victim to.[2] In our first case, the computer-generated study misclassified the HRM data as "premature contractions," when in reality, they were failed peristalsis with DCI values less than 450 mm Hg·s·cm, satisfying the criteria for failed peristalsis and thus combined with clinical picture and diagnostics for Type I achalasia. In our second patient, the computer-generated analysis misidentified esophageal pressurization as contractions and subsequently calculated a DCI, a value that should not have been calculated in Type II achalasia, as per the Chicago classification system.[2] Additional errors caught on manual re-evaluation of the HRM data from other cases include incorrect marking of the lower esophageal sphincter because of the presence of artifact and spastic swallow attempts mislabeled as rapid or ineffective swallow attempts resulting in an initially incorrect diagnosis.

The 2 patients described had previously been diagnosed with "outlet obstruction." However, a diagnosis of EJOO cannot be made without sufficient evidence of peristalsis that was not met in our cases. EJOO is a confusing entity which some experts believe requires a barium study for evidence of distal esophageal

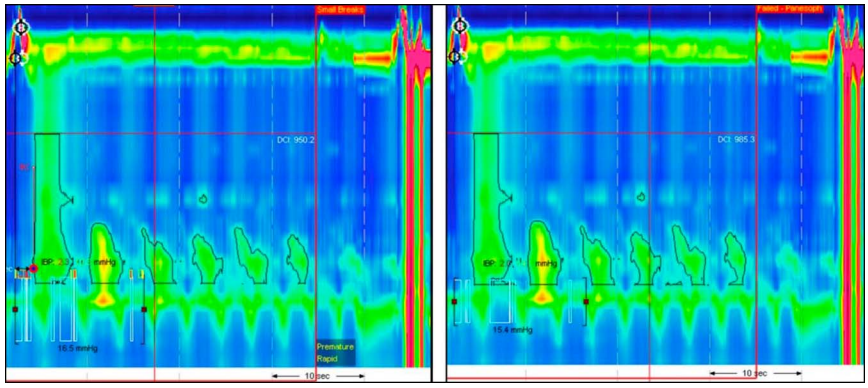

Figure 2. Computer-generated high-resolution manometry analysis showed premature and rapid small breaks on swallow attempts (left). However, on manual data analysis, panesophageal pressurization was identified and the swallow attempts were reclassified as failed attempts (right).

pressurization or elevated intrabolus pressure.[4–6] It is imperative that when physicians note study terms such as rapid swallow attempts, swallow attempts with small or large breaks, and fragmented swallow attempts, they review the study data to ensure sufficient evidence of contractions or peristalsis may downgrade the diagnosis from achalasia to a potential EJOO.

Ultimately, HRM is a powerful tool in the motility examination arsenal. However, the successful application of HRM requires a responsible and well-trained operator who understands potential pitfalls of the evaluation and possesses proper clinical judgment. Reliance on computer-generated study data places the clinician at a high risk of misdiagnosis. In one study series, computer-generated diagnosis based off of the HRM raw data resulted in the correct diagnosis of achalasia in only 30% of cases.[7] Physicians using HRM should use the study data in combination with all endoscopic, radiologic, and physical findings to identify a diagnosis rather than evaluate the manometric data alone. Any data that does not correlate with other patient findings should be carefully reanalyzed. Although the Gastroenterology Core Curriculum currently recommends 2 levels of training in HRM: basic and advanced (50 proctored study administrations and interpretations for competency assessment), there are, at present, no accredited advanced training programs in motility and HRM.[8] Given the growing complexities and advancements in HRM technology, it may behoove the gastroenterological community to develop more thorough and official training requirements in the field to minimize medical malpractice and improve patient care.

DISCLOSURES

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