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Research review

Whose patient is it? The path to multidisciplinary management of achalasia

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ARTICLE INFO

Article history:
Received 31 July 2017
Received in revised form
20 September 2017
Accepted 22 February 2018
Available online xxx

Keywords:
Achalasia
Heller myotomy
Esophageal manometry
Ambulatory pH monitoring
High-resolution manometry
Chicago classification

ABSTRACT

In the past decade, the introduction of high-resolution manometry and the classification of achalasia into subtypes has made possible to accurately diagnose the disease and predict the response to treatment for its different subtypes. However, even to date, in an era of exponential medical progress and increased insight in disease mechanisms, treatment of patients with achalasia is still rather simplistic and mostly confined to mechanical disruption of the lower esophageal sphincter by different means. In addition, there is partial consensus on what is the best form of available treatments for patients with achalasia. Herein, we provide a comprehensive outlook to a general approach to the patient with suspected achalasia by: 1) defining the modern evaluation process; 2) describing the diagnostic value of high-resolution manometry and the Chicago Classification in predicting treatment outcomes and 3) discussing the available treatment options, considering the patient conditions, alternatives available to both the surgeon and the gastroenterologist, and the burden to the health care system. It is our hope that such discussion will contribute to value-based management of achalasia through promoting a leaner clinical flow of patients at all points of care.

Introduction

Achalasia is a motor disorder of the esophagus characterized by aperistalsis and impaired relaxation of the lower esophageal sphincter (LES) due to inflammatory and degenerative changes in neurons with loss of inhibitory innervation of the esophageal wall. This disease mechanism leads to chronic dysphagia, regurgitation, and chest pain.[1] The introduction of high-resolution manometry (HRM) and the classification of achalasia into subtypes has made possible to accurately diagnose the disease and predict the response to treatment for its different subtypes.[2,3] However, even to date, in an era of exponential medical progress and increased insight in disease mechanisms, our understanding of primary achalasia remains somewhat limited with treatment options that are rather simplistic and do not provide definitive cure. Cure is in

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https://doi.org/10.1016/j.jss.2018.02.047

fact aimed at decreasing the resting pressure in the LES by mechanical disruption through surgical myotomy or pneumatic dilatation (PD). Treatment alternatives, albeit with much less efficacy, are typically reserved to patients who are too friable to undergo therapeutic endoscopy or surgery and include topical injection of botulinum toxin (BT) or oral administration of smooth muscle relaxants. Importantly, selection of patients for surgical and endoscopic interventions remains controversial, and there is no interdisciplinary consensus on the optimal management of achalasia.

Our goal is to provide an update into the evaluation process of patients with achalasia to discuss current diagnostic challenges and to advocate for finding the right treatment alternatives available to both the surgeon and the gastroenterologist. Our premise is that a true multidisciplinary approach to the management of achalasia is the best way to maximize patient outcomes and minimize burden to the health care system.

Clinical evaluation and diagnostic challenges

The evaluation process of patients complaining of a combination of solid and liquid dysphagia, heartburn, atypical chest pain, and regurgitation attributed to achalasia aims to confirm the absence of peristalsis, abnormal relaxation of the LES, and assess the degree of esophageal dilatation/tortuosity or the presence of epiphrenic diverticula. The typical workup for achalasia therefore includes symptomatic evaluation, upper endoscopy, barium esophagram, and HRM. Ambulatory pH monitoring is usually reserved to challenging patients who failed previous treatments.[4] These patients are often diagnosed with refractory gastroesophageal reflux disease (GERD) and treated with acid-reducing medications, although about half do not have reflux but achalasia or other motility disorders.[5]

While in the past, the subjective interpretation of the presence and severity of many symptoms including dysphagia and regurgitation were used during the initial clinical evaluation; today, the Eckardt score (ES) is being increasingly adopted as a more objective clinical adjunct.[6] The ES provides a simple, reliable, validated, quantitative evaluation of the severity of symptoms of achalasia, and it helps in the objective postoperative assessment of treatment outcomes. The ES is based on the presence and severity of four elements—weight loss, dysphagia, regurgitation, and retrosternal pain—and ranges from 0 to 12. It is widely accepted that a postoperative ES of three or less represents an excellent treatment result, while a score greater than three is indicative of treatment failure. During the initial evaluation, the clinician should also inquire about respiratory symptoms. Today, there is strong evidence that cough, hoarseness, wheezing, and pneumonia in these patients may be caused by macroaspirations/microaspirations of retained food in the esophagus. It has been shown that after surgical treatment of achalasia, respiratory symptoms improve.[7]

The diagnostic testing includes barium esophagram, upper endoscopy, and esophageal function tests. Barium esophagram provides information about the esophageal anatomy and esophageal emptying (timed video esophagram). The characteristic features of the barium swallow in patients with achalasia are the bird's beak appearance of the very distal esophagus, loss of primary peristalsis, delayed esophageal emptying and, in severe cases, the presence of an air-fluid level. Other suggestive features include a dilated or sigmoid esophagus and the presence of epiphrenic diverticula, as these are almost always caused by a primary esophageal motility disorder, such as achalasia.[4,8]

The main purpose of diagnostic upper endoscopy is to rule out squamous cell esophageal cancer and pseudoachalasia. Pseudoachalasia is a condition caused by esophageal adenocarcinoma infiltrating the neural plexus of the esophagus, resulting in a manometric picture similar to achalasia.[9] Like the barium esophagram, an upper endoscopy alone is not sufficient to diagnose achalasia. However, suggestive features during endoscopy are retention of food or fluid in the esophageal body and presence of an epiphrenic diverticulum.

The gold standard diagnostic test of achalasia is esophageal manometry. With conventional manometry, it is possible to detect the absence of esophageal peristalsis and failure of the LES to relax during swallows. However, the recent introduction of HRM has allowed the identification of three subtypes of achalasia based on the patterns of esophageal contractility and peristalsis. According to the Chicago Classification (CC), achalasia can be distinguished as type I (classic achalasia, lack of peristalsis and contractility), type II (lack of peristalsis, but panesophageal pressurization is detected), and type III (spastic achalasia, lumen-obliterating contractility with impaired peristalsis).[2] The major clinical relevance of CC is its ability to predict the clinical outcomes of treatments depending on the subtype with a high degree of accuracy and reliability.[10] Rohof *et al*., in the European achalasia trial comparing laparoscopic Heller myotomy (LHM) with PD outcomes, reported a different success rate based on the achalasia subtype, supporting the concept that the classification may be useful to determine the treatment of choice. Results suggested that achalasia type I and III are important predictors of treatment failure compared to type II, with type III being the one with worse outcomes even after LHM.[11] This is important, as results of the CC may serve as the first point of care in the clinical flow process in which to choose a treatment option and discuss it with the patient together with the multidisciplinary team (gastroenterologists and surgeons). Recently, the CC has been updated (version 3) and the new classification includes refined morphological and contractility criteria with a better characterization of esophagogastric junction outflow obstruction and the distinction of additional major motility disorders not observed in normal subjects (distal esophageal spasm, hyper-contractile esophagus, and absent contractility).[12]

Ambulatory pH monitoring is usually reserved for patients who have failed previous treatments and for patients with GERD refractory to proton pump inhibitor therapy. It is important to review the tracings of these patients. In fact, the combination of dysphagia and heartburn combined with a pathologic amount of esophageal acid exposure on pH monitoring may prompt a referral for an antireflux operation. On a more careful review, the pH-monitoring tracings of such patients may reveal a false-positive study due to pseudo-reflux and suggest that the presence of abnormal esophageal acid exposure is not due to actual reflux from the stomach but stasis and fermentation of food in a nonemptying esophageal body.[5]