Achalasia in a Sixty-Four-Year-Old Man

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Achalasia is an esophageal motility disorder characterized by increased lower esophageal sphincter pressure and absence of peristalsis in the lower esophagus. Patients typically present with complaints of progressive difficulty swallowing over a period of several years. Diagnosis is confirmed by esophageal manometry. Complications of achalasia include esophagitis, aspiration and possibly an increased risk of esophageal carcinoma. Medical treatment options include pneumatic dilatation, esophageal bougienage, nitrates, calcium channel blockers and botulinum toxin injections. The primary method of surgical treatment is the Heller myotomy, in which longitudinal incisions are made in the muscle fibers of the lower esophageal sphincter to reduce sphincter pressure. Frequently, a fundoplication is performed in addition to the myotomy to decrease the likelihood of development of gastroesophageal reflux.

In recent years, the Heller myotomy has been performed both thoracoscopically and laparoscopically. An additional development has been the placement of an endoscope in the esophagus to provide transillumination during surgery; intraoperative endoscopy allows improved assessment of the depth of myotomy incisions and reduces the risk of esophageal perforation. The case report below describes a 64-year-old-man with achalasia who presented with persistent dysphagia despite multiple attempts at medical treatment. A laparoscopic Heller myotomy with Toupet fundoplication was performed with subsequent eradication of symptoms. A discussion of the epidemiology, etiology, clinical presentation, diagnosis and treatment of achalasia follows the case report.

INTRODUCTION

Achalasia is an esophageal motility disorder characterized by three abnormalities of smooth muscle function: (1) failure of the lower esophageal sphincter to relax completely following deglutition, (2) normal to increased lower esophageal sphincter pressure and (3) absence of peristaltic contractions in the esophageal body [1]. The most common clinical manifestation of achalasia is dysphagia, which may be accompanied by chest pain. Treatment of this condition involves disruption of non-relaxing smooth muscle in the area of the lower esophageal sphincter.

A case of severe achalasia and its treatment in a 64-year-old man is presented below.

CASE REPORT

G.W. is a 64-year-old white male who presented to the GI surgical service at Yale New Haven Hospital for evaluation of difficulty swallowing. At that time, the patient described progressive difficulty swallowing over the seven-month period prior to admission. He stated that this dysphagia was most pronounced when he attempted to swallow

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Abbreviations: LES, lower esophageal sphincter; EG, esophagogastric.
solids. The patient had initially been able to swallow liquids successfully and had been drinking nutritional supplements; more recently, he had begun experiencing dysphagia with liquids as well and reported regurgitating solidified beverages.

Upon the recommendation of his internist, the patient consulted a gastroenterologist. Five months prior to admission, a barium swallow, endoscopy and esophageal manometry were performed. The barium swallow demonstrated severe distal esophageal narrowing. The endoscopy demonstrated normal mucosa without evidence of neoplasm or any obstructive intraluminal masses. Manometry revealed a lower esophageal sphincter (LES) pressure of 33 mm Hg (normal LES pressure ranges from 10 to 20 mm Hg) as well as absent peristalsis in the distal esophagus. Contractions occurred simultaneously throughout the distal esophagus and were of low amplitude. The patient was diagnosed with achalasia on the basis of his clinical presentation and the above findings.

Esophageal balloon dilatations were performed three times within the six-month period prior to admission. The patient reported temporary decreases in dysphagia but stated that the difficulty swallowing returned within days to weeks following each episode of dilatation.

The patient reported a twelve pound weight loss since six months prior to admission. He denied loss of appetite and any gastrointestinal complaints other than difficulty swallowing. One month prior to admission, he was offered treatment options including further balloon dilatation, therapy with botulinum toxin or elective surgical myotomy. After consideration, the patient chose the surgical procedure. Following discontinuation of coumadin (the patient had been taking coumadin since a pulmonary embolism one year prior), the patient was admitted for IV heparin therapy. After cessation of heparin, he underwent a laparoscopic Heller myotomy and Toupet partial fundoplication. These procedures were performed via five separate laparoscopic port sites using a 30-degree angle telescope for full visualization of the region of the esophagogastric (EG) junction. Intraoperative endoscopy was performed during the myotomy, which permitted precise control of gastric insufflation and transillumination of the myotomy site. A 270-degree fundoplication was created, and seven sutures were used to secure the two fundal components of the wrap to the edges of the myotomy. At the conclusion of the myotomy, the mucosa bulged freely along its entire length, which extended 5 cm proximal and 2 cm distal to the EG junction (Figure 1). The patient's post-operative course was remarkable only for an episode of angina accompanied by T wave inversions on ECG. The patient's ECG reverted to normal, and he did not have a significant increase in cardiac enzymes. He began eating a soft diet on post-operative day two and was discharged to home on post-operative day four. At follow-up two weeks later, the patient was eating a normal diet without difficulty. He reported occasional heartburn symptoms, which were controlled by daily administration of an H₂ receptor antagonist.

**DISCUSSION**

**Epidemiology**

Achalasia is a rare disorder with an incidence of less than one per 100,000 people per year in the USA, and there is no definite sex or race predilection [2]. The relative contribution of genetic and environmental factors toward the development of achalasia is unclear. Previous suggestions of autosomal recessive inheritance remain unsubstantiated [3]. A large retrospective study of hospitalizations for achalasia demonstrated geographical variation in the frequency of achalasia among Medicare patients, with high rates in the South and low rates around the Great Lakes and the Pacific U.S. [2].
Etiology

Achalasia appears to result from esophageal denervation; histological examination of tissue from patients with achalasia demonstrates degenerative changes in the ganglion cells of the myenteric plexus, the dorsal motor nucleus of the vagus nerve and the fibers of the vagus nerve itself [4]. In addition, inflammatory changes with lymphocytic infiltration are also evident in association with the myenteric plexus [5]. Impairment of relaxation of the LES is probably due to decreased function of nerves that inhibit contraction of the sphincter. The neurotransmitters involved most likely include nitric oxide and vasoactive intestinal peptide [6]. The etiology of the neuronal degeneration in patients with achalasia is unknown; hypotheses include damage secondary to infection with a virus such as varicella zoster or an autoimmune process [5].

Clinical Presentation

Achalasia is usually characterized by an indolent course; symptoms are present for an average of seven years before medical help is sought [4]. The most common presenting symptom is dysphagia, which is present in 82 percent to 100 percent of patients [4, 7]. This dysphagia may be intermittent, may occur with ingestion of solids, liquids, or both, and usually becomes progressively worse over time [7]. Regurgitation of stagnant, undigested food occurs in 56 to 94 percent of patients [4] and can cause aspiration with nocturnal cough, pneumonia or lung abscesses. Chest pain occurs in 50 percent of patients; achalasia is diagnosed in 10 percent of patients with noncardiac chest pain. The pain may be caused by esophageal dilation proximal to the LES, esophagitis or esophageal spasms.
In one series, heartburn was reported in 44 percent of patients with achalasia [9]. This subternal burning discomfort is purported to be secondary to fermentation of static food in the esophagus by bacteria rather than by gastroesophageal reflux [10]. Furthermore, weight loss is also often a presenting symptom of achalasia. Weight loss can occur as a result of inadequate esophageal emptying in addition to decreased intake due to fear of dysphagia and chest pain.

Previously, achalasia has been discussed in terms of two distinct entities: classic and vigorous achalasia. Patients with relatively high amplitude esophageal contractions were said to have vigorous achalasia, which was thought to be associated with chest pain. A retrospective study performed in 1991, which compared radiographic and clinical findings as well as response to balloon dilatation in patients with classic and vigorous achalasia, did not find any significant difference between the two groups [11]. The distinction has been utilized less frequently in recent years.

Diagnosis

Esophageal manometry represents the gold standard for diagnostic confirmation of achalasia. Manometric evaluation of esophageal pressure in patients with achalasia typically demonstrates normal peristaltic contractions in the upper esophagus. The distal esophagus exhibits low amplitude contractions, which occur simultaneously rather than in a peristaltic wave pattern. In addition, the LES shows a failure of relaxation and often a high resting pressure. The LES resting pressure is usually elevated to about 35 mm Hg (normal, 10 to 20 mm Hg) [7].

Barium esophagrams of patients with achalasia typically demonstrate a "bird's beak" appearance of the distal esophagus at the region of esophageal narrowing, and a dilated esophagus resembling the sigmoid colon is often seen proximally [12]. Chest radiographs can show a widened mediastinum due to esophageal dilation, an air-fluid level in the posterior mediastinum due to stasis of swallowed food and liquid in the esophagus and the absence of a gastric air bubble (50 percent of patients) [12].

Endoscopy of the upper GI tract in patients with achalasia is usually unremarkable but is mandatory in patients being evaluated for achalasia in order to exclude pseudoachalasia (achalasia secondary to malignancy). Malignancy represents the most ominous of the various causes of secondary achalasia, which include amyloidosis, Chagas' disease, MEN IIb and familial adrenal insufficiency [12]. Malignancy can produce achalasia through a number of mechanisms, including direct invasion of the myenteric plexus and ganglion cells by tumor, damage to the vagus nerve by tumor or a paraneoplastic neuropathy not involving direct neuronal invasion by tumor [12]. There are three clinical findings that should raise suspicion of a responsible malignancy in a patient with achalasia of unknown origin: onset of symptoms after age 60, presence of symptoms for less than one year and weight loss of more than twenty pounds [4].

Complications

The complications associated with achalasia are, for the most part, secondary to food stasis. Retained food and fluid in the esophagus creates a hospitable environment for bacterial or candidal infection, producing esophagitis. Stasis also frequently results in aspiration, which can lead to pneumonia or lung abscesses, as stated above. In addition, the chronic presence of static food in the esophagus is thought to contribute to an increased incidence of esophageal carcinoma in patients with achalasia. A large-scale population-based study conducted in Sweden demonstrated a sixteen-fold increase in the risk of esophageal cancer (primarily squamous cell carcinoma) in patients with, compared to patients without, achalasia [13]. An additional complication of achalasia is the development of epiphrenic diverticula secondary to increased intraesophageal pressure.
Treatment

Medical treatment of achalasia has been directed at decreasing LES pressure in order to improve esophageal emptying and prevent stasis-related complications. The most frequently used non-surgical treatment method is pneumatic dilatation, which consists of rupturing LES sphincter fibers by passing a balloon dilator through the LES and inflating the balloon. Studies examining the overall efficacy of a course of treatment of balloon dilatation in the long-term reversal of symptoms of achalasia have demonstrated clinical improvement in approximately 70 percent of patients [4]. The majority of patients do not obtain lasting relief from the symptoms of achalasia after a single episode of balloon dilatation; in one study, only 20 percent of patients exhibited a decrease in LES pressure and a return of peristalsis following the initial dilatation [14]. However, repeated dilatations produced a response in a greater number of patients. Results were better in patients over age 45 who had experienced symptoms for more than five years. The complications associated with pneumatic dilatation include esophageal perforation, which occurs in two to 10 percent of patients [4, 15]. Less commonly, aspiration pneumonia, localized hematoma, or esophageal tear without perforation occur. Finally, reflux esophagitis was reported as occurring in one to seven percent of patients following balloon dilatation in studies conducted in the 1980s [16-18]; a more recent study questioned the validity of these earlier results and suggested that heartburn symptoms and esophagitis in post-dilatation patients could be due to impaired esophageal clearing rather than reflux [19].

Other nonsurgical methods for the treatment of achalasia include esophageal bougienage, which usually results only in temporary relief from dysphagia and is rarely used. Pharmaceutical intervention is more common: nitrates and calcium channel blockers have been used with some success to decrease LES pressure in patients with achalasia [1]. More recently, botulinum toxin, which inhibits the release of acetylcholine from nerve terminals, has been injected directly into the LES to induce relaxation of the sphincter by decreasing excitatory cholinergic innervation to the sphincter muscle. Early studies demonstrated that treatment with botulinum toxin is safe and effective in the short term. In one prospective study, intraspincteric administration of botulinum toxin produced a decrease in symptoms associated with achalasia in two-thirds of patients for a period of approximately 16 months following initial treatment [20]. However, patients who choose intraspincteric botulinum injections as their mode of therapy require endoscopy on a nearly annual basis; furthermore, the long-term safety and efficacy of repeated botulinum toxin injections has not been evaluated.

Patti et al. conducted a prospective study comparing patients receiving pneumatic dilatation, pharmacotherapy with calcium channel blockers, or both, with patients receiving surgical treatment (myotomy via a thoracoscopic approach) [22]. Treatment results were assessed by telephone interviews with patients every three months for a mean period of 28 months after therapy. Results were classified according to the following schema: “Excellent” signified no dysphagia; “Good” indicated less than one episode of dysphagia per week; “Fair” meant more than one episode of dysphagia per week; and “Poor” represented persistent dysphagia. Within the group receiving medical treatment, 26 percent of patients had “Excellent” or “Good” responses to medical treatment (all of those patients had undergone balloon dilatation at least once), and 74 percent had “Fair” to “Poor” responses. In the surgical group, 87 percent of patients had “Excellent” or “Good” results, and the remaining 13 percent had “Fair” or “Poor” responses to treatment [22].

The mainstay of surgical treatment for achalasia is the Heller myotomy, in which the muscle fibers of the LES are destroyed by longitudinal incisions. The myotomy extends from approximately 5 to 7 cm above the LES to several centimeters below the EG junction (the appropriate lower border of the myotomy remains subject to debate), and the
mucosa and submucosa bulge outward from the lumen following the full-thickness division of esophageal muscle fibers [21]. The procedure was first described by Heller in 1914 and initially required either a laparotomy or a thoracotomy; more recently, the Heller myotomy has been performed via a laparoscopic or thorascoposcopic approach with an associated decrease in length of hospital stay and post-procedural morbidity. In one study comparing laparoscopic versus open Heller-Dor procedures, the median postoperative hospital stay was four days following laparoscopy and 14 days following laparotomy. The median time elapsed before patients returned to normal activity was 10 days after the laparoscopic procedure and 30 days after the open procedure [24]. One disadvantage of minimal access approaches is the absence of tactile information available during an open procedure. The placement of an endoscope in the esophagus during the myotomy allows better assessment of the depth of the myotomy and decreased the risk of perforation of the esophageal wall [22].

Controversy exists regarding the superiority of the thorascoscopic or the laparoscopic approach. Raiser et al. suggest that the laparoscopic approach provides better access to the gastric fundus, enabling the surgeon to extend the myotomy further onto the stomach and decreasing the difficulty of performing an anti-reflux procedure [21]. Their study demonstrated a statistically significant increase in both the duration of postoperative hospital stay and in time elapsed prior to return to normal activity in patients approached thorascoscopically as opposed to laparoscopically. It is also the impression of the authors that the transabdominal approach is technically simpler and results in faster recovery than the thorascoscopic approach.

The main complication associated with the surgical treatment of achalasia is the development of gastroesophageal reflux, which occurs in approximately 12 percent of patients following Heller myotomy [23]. The likelihood of the development of reflux can be greatly reduced by the concomitant performance of an anti-reflux procedure. The two most frequently employed procedures are the Dor fundoplication, in which a portion of the gastric fundus is pulled anteriorly around the esophagus and fixed to the cut edges of the myotomy, and the Toupet procedure, in which the gastric fundus is wrapped 270 degrees around the esophagus from its posterior aspect and each cut myotomy edge is fixed to the adjacent portion of the fundus. Raiser et al. compared the Heller-Toupet and Heller-Dor procedures performed either laparoscopically or thorascoscopically in terms of post-surgical manometry, pH analysis and clinical course and concluded that the Heller-Toupet procedure performed laparoscopically was the favored approach to surgical intervention for achalasia [21]. At long-term follow up, 33 percent of patients who received the Toupet procedure and 71 percent of patients upon whom the Dor procedure was performed reported some degree of dysphagia; 27 percent of patients described heartburn-like symptoms following the Toupet procedure compared to 57 percent of patients after the Dor procedure. Both differences are statistically significant. The discrepancy in rates of dysphagia following surgery may result from structural differences in the two fundoplications: in the Dor procedure, the gastric fundus covers the myotomy site, limiting the bulging of the mucosa and creating the opportunity for scar formation between the esophageal mucosa and the fundal wrap. In the Toupet procedure, the myotomy is left uncovered so the mucosa is free to bulge outward [21].

The patient discussed in this case report presented with the dysphagia, the most common presenting complaint associated with achalasia. Manometric and radiographic findings were also entirely consistent with the diagnosis of achalasia. His presentation was atypical in terms of the relatively rapid development of clinically significant symptoms, which would raise suspicion of achalasia secondary to malignancy, however pseudoachalasia was ruled out in his case by endoscopic examination. The patient initially received the most frequently used medical treatment for achalasia, balloon dilatation, but three
consecutive courses of dilatation did not significantly reduce his symptoms. Surgical treatment with laparoscopic Heller myotomy and Toupet fundoplication was successful at initial and three-month follow-up in eradicating the patient’s dysphagia. The patient did experience symptoms of heartburn following the procedure, but these were satisfactorily controlled with daily administration of cimetidine. This patient’s case illustrates the effectiveness of surgical intervention in treating this disease. The questions of when to refer a patient for surgical treatment and whether surgery represents a superior method of treating achalasia, are controversial. A subgroup of patients receiving pneumatic balloon dilatation have very satisfactory outcomes and do not require surgery to address residual symptoms. Patients requiring multiple non-operative interventions, who have continued symptoms or incomplete responses, may be considered for transabdominal Heller myotomy with an anti-reflux procedure.

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