ESOPHAGEAL AND LUNG INSTITUTE VISITING LECTURER SERIES

George Liebler Thoracic Oncology Lectureship

Lecture Overview:
The purpose of this lecture series is to bring in thought leaders in the field of esophageal and lung surgery who are innovative in the field. The series is designed for the exchange of ideas and to energize and rejuvenate practitioners, faculty, trainees, and staff.

Friday, December 4, 2015

Keynote Speaker: Wayne Hofstetter, MD

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To Register:
Online registration is preferred. To register online or download a brochure, please go to www.agh.org. Select Conference Schedule, then scroll to find the ELI Lecture Series.
Esophageal Motility Disorders

Introduction

The esophagus is a highly organized muscular tube, with an antigravity peristaltic muscle layer such as longitudinal and circular muscles, both contracting synchronously and working together to deliver food bolus from the throat to the stomach without any resistance. Esophageal motility disorders are caused by lack or disorganization of esophageal muscle functions, and typically manifest as dysphagia (difficulty swallowing and sticking) and non-cardiac chest pain. Abnormal esophageal motility can lead to aspiration. In general, esophageal motility disorders are classified into primary esophageal motility disorders and secondary motility disorders, which is idiopathic or secondary to a known condition, such as gastroesophageal reflux disease (GERD) and connective tissue disorders. Primary esophageal motility disorders are the most common, divided into esophageal spasm (type II) and achalasia with esophageal spasm (type III). The correct terminology is idiopathic or secondary to a known condition, such as GERD and connective tissue disorders. Primary esophageal motility disorders are the most common, divided into esophageal spasm (type II) and achalasia with esophageal spasm (type III).

Table 1. Synopsis and treatment of achalasia and other motility disorders

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Achalasia</td>
<td>Classification as a primary motility disorder due to incomplete distal relaxing LES, with an unknown etiology, and typically manifests as dysphagia (difficulty swallowing, food sticking), regurgitation of undigested food, and weight loss.</td>
</tr>
<tr>
<td>Other spastic motility disorders</td>
<td>Other spastic motility disorders include hypercontractile esophagus (type IV), distal esophageal spasm (DES), and esophageal spasm (type II). Other spastic motility disorders are characterized by increased esophageal contractility, which may lead to symptoms such as dysphagia, chest pain, and regurgitation.</td>
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<tr>
<td>Pseudachalasia</td>
<td>A clinical syndrome similar to achalasia, characterized by incomplete distal LES relaxation, but absence of demonstrable anatomic lesions that can cause pseudoachalasia.</td>
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<tr>
<td>POEM</td>
<td>Peroral endoscopic myotomy (POEM) is a novel endoscopic procedure that enables transmural myotomy over the entire length of the intrathoracic esophagus, improving esophageal emptying and dysphagia.</td>
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</tbody>
</table>

Achalasia

Achalasia is the most common primary esophageal motility disorder with the estimated prevalence of 0.5-1 per 100,000 people. It is characterized by esophageal body and incomplete relaxation of the lower esophageal sphincter (LES) with an unknown etiology, and typically manifests as dysphagia (difficulty swallowing, food sticking), regurgitation of undigested food, and weight loss.

Table 1 shows a "bird's-eye" view of a ball that is stuck in the throat, sometimes tortuous esophagus body in the upright position (Figure 1), which is noted that the esophagus body in the upright position may not present or be concealed in a "bird's-eye" view. Recently, the lower esophageal sphincter is observed in a "bird's-eye" view, which is the lower esophageal sphincter body in the upright position. The height of residual esophageal column correlates with the severity of regurgitation and esophageal emptying.

Diagnosis

The diagnosis of achalasia can be suspected based on clinical presentation, barium esophagram and upper endoscopy, and confirmed by upper gastrointestinal manometry. A lower esophageal sphincter is a radiographic test and the best initial diagnostic tool. A barium esophagram shows a symmetrical tapering at the gastroesophageal junction and is observed in approximately 2-4% of patients with a suspicious diagnosis of achalasia. The most common etiology of pseudoachalasia is a migratory inflammatory infiltrate of the LES in pseudoachalasia. The symptoms are older and have a longer history of dysphagia, and often have a low prevalence of pseudoachalasia. Further study is needed as such endoscopic ultrasound and CT scan may not necessarily confirm the diagnosis of achalasia.

Eosophageal Manometry

Eosophageal manometry is a gold standard to confirm the diagnosis of achalasia. Characteristics of achalasia include incomplete relaxation of the LES and aperistalsis of the esophageal body. Since the introduction of high-resolution manometry, it is now possible to be been both in esophageal body and LES. The International Swallowing and Esophageal Motility Consensus Conference (ISEM) has classified achalasia into three presentations type I and achalasia esophageal spasm (type II) and achalasia with esophageal spasm (type III) (Figure 2). A type II achalasia approach has to be tailored to the patient’s treatments.

Treatment

Achalasia is first treated with dilation using a bougie or tubes. Some form of treatments can improve the symptoms of achalasia. The success rate of balloon dilation within 6 months is approximately 75% but symptoms return in half of patients approximately 35 months. The previous studies have demonstrated that the success rates improve with increased sizes of balloons (74% at 30 mm, 86% at 35 mm and 92% at 40 mm); however the success rates decline over time (46%, 1-6 months; 17%, 8-12 months; 6%, after 12-20 months and 36% after 2 years) (4). Repeat balloon dilation may have been proposed to achieve a long-term maintenance of symptomatic relief. However, the International Pseudoachalasia Survey has demonstrated that balloon dilation is added in the form of a pseudoachalasia. There is no evidence on the recurrence of pseudoachalasia.

POEM

POEM is a promising treatment option for both achalasia and spastic esophageal motility disorders. The treatment strategy for spastic motility disorders remains controversial, although endoluminal treatment is similar to those for achalasia. Hypercontractile esophagus may be managed with POEM, the most common complication is a perforation of the esophagus and is also characterized by a “corkscrew” or “double helix” appearance. These data suggest that POEM is equivalent to a conventional myotomy over the entire length of the intrathoracic esophagus. However, the success rates for POEM are slightly less than those for achalasia. The international POEM Survey has demonstrated slightly less efficacy of POEM for type II and type IV achalasia but excellent efficacy for hypertensive LES using balloon occlusion and hypercontractile esophagus. Hypercontractile esophagus on manometry, and is also characterized by a “corkscrew” or “double helix” appearance. The clinical criteria for POEM is defined as a simple physiologic evaluation of esophageal emptying for EGJ outflow obstruction, distal esophageal spasm (DES) and Jackhammer esophagus. (Right bottom) typical manometric findings of achalasia; minimal esophageal pressurization (type I, classic), achalasia with esophageal spasm (type II), achalasia with esophageal spasm (type III).

Other spastic motility disorders

Other spastic motility disorders include hypercontractile esophagus (type IV), distal esophageal spasm (DES), and esophageal spasm (type II). Other spastic motility disorders are characterized by increased esophageal contractility, which may lead to symptoms such as dysphagia, chest pain, and regurgitation. These symptoms may be improved with POEM, but symptoms may recur within 1-2 years. Other studies have demonstrated that long-term efficacy of POEM is inferior to pneumatic dilation or surgical myotomy. The most common side effect of POEM is scarring or perianal perforation (up to 4%).

Pseudoachalasia

As previously mentioned, the most common complications of pseudoachalasia are older and have a shorter history of dysphagia, and often have a low prevalence of pseudoachalasia. Further study is needed as such endoscopic ultrasound and CT scan may not necessarily confirm the diagnosis of achalasia.