Case Studies and Review of Jackhammer Esophagus

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INTRODUCTION

Jackhammer esophagus is a hypercontractile esophageal motor disorder. Jackhammer esophagus is defined by high-resolution manometry (HRM) when high amplitude, high speed waves of contractions occur that have a distal contractile integral (DCI) greater than 8000 mm Hg/cm/s. (1, 2) We present a series of five cases with their clinical and manometric features plus a review of the subject.

METHODOLOGY

We reviewed reports from high-resolution esophageal manometry performed in the physiology unit of Hospital San Ignacio in Bogota last year for diagnoses of esophageal jackhammer. Five cases were found. Patients’ digestive symptoms were recorded and any additional studies such as upper gastrointestinal tract endoscopy (UDE), barium enemas and HRM were reviewed. Relevant variables for
analysis of cases were then compiled. Finally, a search was conducted in PubMed with search terms of Jackhammer esophagus and hypercontractile esophagus. We generated a text with which to review which is appended at the end of the list of references.

CASES

First Case

The patient was a 63-year-old man who had suffered acute myocardial infarction in 2014 that compromised three vessels. It was initially treated with coronary stenting which was followed with surgical myocardial revascularization and two coronary bridges. The patient consulted a physician because of persistent chest pain with atypical features that was accompanied by dysphagia for solids, but no impacted food was found. Cardiological studies ruled out a cardiogenic origin. Upper digestive endoscopy was normal. HRM found 1 of 10 waves with a DCI of 8,351 mm Hg/cm/s. Other waves measured over 5,000 mm Hg/cm/s (Tables 1 and 2 and Figure 1).

Table 1. Clinical and endoscopic characteristics of patients diagnosed with jackhammer esophagus.

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>n</th>
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<tbody>
<tr>
<td>Symptom</td>
<td></td>
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<tr>
<td>Regurgitation</td>
<td>3/5</td>
</tr>
<tr>
<td>Pyrosis</td>
<td>3/5</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>3/5</td>
</tr>
<tr>
<td>Chest pain</td>
<td>2/5</td>
</tr>
<tr>
<td>Diagnosis of GERD</td>
<td>3/5</td>
</tr>
<tr>
<td>Endoscopic Findings</td>
<td></td>
</tr>
<tr>
<td>Hiatal Hernia</td>
<td>2/5</td>
</tr>
<tr>
<td>Esophagitis</td>
<td>1/5</td>
</tr>
<tr>
<td>Esophageal Diverticulum</td>
<td>1/5</td>
</tr>
<tr>
<td>Presbyesophagus</td>
<td>2/5</td>
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</tbody>
</table>

Second Case

The patient was a 45-year-old woman with typical symptoms of gastroesophageal reflux disease (GERD) caused by heartburn and regurgitation without dysphagia or chest pain that was refractory to treatment with proton-pump inhibitors (PPIs). Upper endoscopy showed hiatal hernia and esophagitis. Given that she had been refractory to treatment, HRM was performed. It found that three of every 11 waves had DCIs above 5,000 mm Hg/cm/s, and one of every 11 measured more than 8000 mm Hg/cm/s (12,562). Intrabolus pressure during the HRM was normal (Tables 1 and 2 and Figure 2).

Figure 1. 63-year-old male patient with DCI of 8,351 mm Hg/cm/s with large amplitude waves and full relaxation of the lower esophageal sphincter. A) High resolution manometry. B) Conventional manometry.

Figure 2. 45-year-old woman with DCI of 12,562 mm Hg/cm/s with large amplitude, high velocity waves. A) High resolution manometry. B) Conventional manometry.
Third Case

The patient was a 72-year-old man with typical symptoms of gastroesophageal reflux disease (GERD) who also had occasional dysphagia for solids. Upper endoscopy showed no abnormalities, but a barium enema showed presbyesophagus and apparent esophageal diverticulum. HRM found three waves with DCIs over 5,000 mm Hg/cm/s, one with a DCI of 8,809 mm Hg/cm/s. There were also six multi-peak waves which reinforced the diagnosis of esophageal jackhammer (Tables 1 and 2 and Figure 3).

<table>
<thead>
<tr>
<th>Case</th>
<th>Highest DCI</th>
<th>Waves with DCI &gt; 8,000 mm Hg/cm/s</th>
<th>Waves with DCI &gt; 5000 mm Hg/cm/s</th>
<th>Multi-peak Waves</th>
<th>IRP mm Hg</th>
<th>Presión Intrabolus mm Hg</th>
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<tr>
<td>1</td>
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<td>1 of 10</td>
<td>10 of 10</td>
<td>0</td>
<td>7.9</td>
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<td>2</td>
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<td>1 of 11</td>
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<td>0</td>
<td>2.5</td>
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</tr>
<tr>
<td>3</td>
<td>8,809</td>
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<td>3 of 11</td>
<td>6 of 11</td>
<td>12.5</td>
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<td>4</td>
<td>16,285</td>
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<td>8 of 10</td>
<td>3 of 10</td>
<td>6.4</td>
<td>23.1</td>
</tr>
</tbody>
</table>

Fourth Case

A 73-year-old male patient with a long-standing history of GERD without dysphagia or chest pain and with typical symptoms that had become refractory to PPIs. Endoscopic revealed a hiatal hernia and presbyesophagus without esophagitis. HRM found that eight out of twelve waves had multiple peaks of more than 5000 mm Hg/cm/s and that four had DCIs above 8,000 mm Hg/cm/s. the highest DCI was 16,285 mm Hg/cm/s. This patient also had a high intrabolus pressure of 26 mm Hg. (Tables 1 and 2 and Figure 4).

Figure 3. A 72-year-old male patient with DCI of 8,809 mm Hg/cm/s with multi-peak waves (at least three peaks) of large amplitude and duration. A) High resolution manometry. B) Conventional manometry.

Figure 4. Male patient with DCI of 16,285 mm Hg/cm/s with multi-peak waves of large amplitude and high velocity with complete relaxation of the lower esophageal sphincter. A) High-resolution manometry. B) Conventional manometry.
Fifth Case

A 41-year-old female patient who had suffered symptoms of occasional globus pharyngis without dysphagia or chest pain for two years. Upper endoscopy found nothing of significance. HRM showed the 8 of 10 waves had DCIs above 5,000 mm Hg/cm/s and that three of ten waves were multi-peak including one with a DCI above 8,000 (8,258 mm Hg/cm/s) (Tables 1 and 2 and Figure 5).

TOPIC REVIEW

Updating the Classification of Esophageal Contractility Disorders

The International Working Group for Gastrointestinal Motility and Function, led by Dr. Peter Kahrilas of Northwestern University in Chicago, Illinois, has taken the responsibility of collecting the available evidence and building consensus criteria for defining esophageal contractility disorders. The latest version of its classification (v.3 Chicago 2015) establishes five groups of results which are based on analysis of functional status of lower esophageal sphincter (LES) and peristalsis. Whether the functional status of the LES is altered is determined by measurement of the integrated relaxation pressure (IRP). (1, 2) It should be our physiology unit still uses the older method for reporting HRM results because it does not yet have the equipment needed for interpreting and classifying esophageal disorders according to Chicago v.3. (1) Based on the new criteria (Table 3) the following three groups of esophageal manometric anomalies have been established:

1. Outflow tract disorders include achalasia Types I, II and III, and outflow tract obstruction (no change from the previous version).

Figure 5. Female patient of 41 years with a DCI of 8258 mm Hg/cm/s. A) High resolution manometry. B) Conventional manometry.
2. Major peristalsis disorders do not have altered IRP (<15 mm Hg). They include diagnoses of distal esophageal spasms with more than 20% of contractions occurring prematurely, aperistalsis (absence of peristaltic waves), and jackhammer esophagus when at least two waves with DCIs over 8,000 mm Hg/cm/s occur. The previous Chicago classifications’ criteria for this disorder of a single wave of high contractility is still used in Colombia. The new classification eliminates the diagnosis of nutcracker esophagus since this manometric alteration occurs frequently in healthy patients and is not a real disorder of esophageal contractility.

3. Minor disorders of peristalsis are characterized by normal IRP associated with ineffective waves (more than 50%) in what is now called ineffective motility disorder. If more than 50% of the waves are effective, but are fragmented in equal percentage of cases, fragmented peristalsis is diagnosed. (2)

**DEFINITION OF JACKHAMMER ESOPHAGUS**

Jackhammer Esophagus is a hypercontractile motor disorder of the esophagus which is diagnosed with HRM when esophageal waves have high amplitude and high speed so that the DCI measures higher than 8,000 mm Hg/cm/s. (1,2) It may be associated with outflow obstructions or abnormalities of the lower esophageal sphincter. (3,4)

**PHYSIOPATHOLOGY**

The hypercontractile characteristic of jackhammer esophagus results from temporal asynchrony between contractions of the circular and longitudinal muscle layers of the muscularis and is probably due to excessive cholinergic activity. (5) The detection of these abnormalities in diabetic patients with autonomic neuropathy by Loo et al. supports this hypothesis of excess cholinergic stimulation. (6) In this study, multi-peak contractions were also more frequent in diabetic patients with neuropathy than in control subjects or in diabetics without neuropathy. This may also occur in patients with jackhammer esophagus in whom thickness of the esophageal smooth muscles has also been observed increased. (7)

A clinical and pathophysiological relationship exists between hypercontractile esophageal disorders and GERD. (8) A study by Crespin et al. has found that a large proportion of patients (69.2%) with hypercontractile esophageal disorders also have GERD symptoms of regurgitation and/or heartburn and that 53% had abnormal exposure to acidic pH as measured by esophageal pH monitoring. (9) Some of these patients underwent Nissen fundoplication with resolution of symptoms, decreased exposure to acid pH and, especially, normalization of esophageal peristalsis. The authors conclude that the symptoms of patients with hypercontractile disorders, typical reflux symptoms, and acidity related to GERD, improve with treatment that reduces exposure to acid pH and that this treatment also solves the esophageal contractility disorders.

**CLINICAL SIGNS AND SYMPTOMS**

Dysphagia, chest pain, regurgitation and epigastric pain are all associated with hypercontractile esophageal disorders but not specific to these disorders. When they are present, other disorders such as cardiac pathologies which may have lethal potential should be ruled out before hypercontractile esophageal disorders are considered. (10) Richter and Castell conducted a study that found that less than 5% of patients with these symptoms had peristalsis disorders demonstrable by esophageal manometry. (11) The combi-
ned prevalence of distal esophageal spasms, spastic esophageal achalasia and jackhammer esophagus is only about 2%. (12, 13) The deterioration of transit of esophageal boluses may be the cause of spastic contractions and may explain dysphagia. Chest pain is probably due to altered contractions, and hypersensitivity can be explained by the perception of acidity in patients without demonstrable evidence of reflux. (14) These patients frequently present epiphrenic diverticula which may occur as a result of hypercontractile disorders. The presence of a diverticulum could also explain the symptoms of dysphagia or regurgitation. (15)

DIAGNOSIS

Upper Digestive Tract Endoscopy

The first examination to be conducted as part of the initial study of the symptoms reported by the patient is an upper digestive tract endoscopy even though the results are generally normal. Sometimes endoscopy shows abnormal contractions or changes in the esophageal anatomy. If there is high suspicion, biopsy samples must also be taken to rule out eosinophilic esophagitis, especially when dysphagia is a prominent symptom. (16)

Esophageal Manometry

Esophageal manometry is the diagnostic gold standard for study of abnormal esophageal motility, even more so with the advent of high-resolution manometry. High-resolution manometry is superior to conventional manometry for assessment of the gastroesophageal junction and for quantifying contractile amplitude and wave speed through use of the DCI. (17) Conventional manometry cannot simultaneously monitor the motor function of the upper esophageal sphincter (UES), the esophageal body, and the lower esophageal sphincter (LES) with each swallow while the high-resolution manometry provides this possibility with a full spatiotemporal representation of motor functions of the esophagus. (18) To avoid false hypercontractility waves, there should be intervals of 20 seconds to 30 seconds between each swallow. As has been documented, these small ranges waves with higher DCIs. The criteria for diagnosis of esophageal jackhammer have been discussed earlier in this article.

Esophageal pH Monitoring

Given the relationship of GERD and hypercontractile disorders of the esophagus in patients with typical reflux symptoms (heartburn and regurgitation), measurement of pH in the distal third of the esophagus should be considered before defining treatment. While esophageal pH monitoring is not a perfect method, it allows accurate assessment of the degree of esophageal acid exposure and also correlate patient symptoms with reflux episodes.

TREATMENT

Given jackhammer esophagus’s low prevalence, there is no consensus on management of this condition. Nevertheless, since the study by Crespin et al. and other recent evidence have reinforced the idea that typical GERD symptoms can be related to jackhammer esophagus, initial management of patients who have both should be directed toward decreasing exposure to acid with either medications or surgery. (9) In contrast, for patients with dysphagia and chest pain who do not have GERD symptoms or elevated exposure to acid, management should try to decrease the amplitude of esophageal contractility through relaxation of smooth muscle tissue while also optimizing relaxation of the lower esophageal sphincter. Ideally, management should be by stages: first, seek control of symptoms with medication, either monotherapy or combination therapies; and second, consider the benefit of surgical or endoscopic treatment for refractory cases (Table 4).

MEDICAL MANAGEMENT

Proton-Pump Inhibitors (PPIs)

Proton-pump inhibitors (PPIs) should always be considered the drug of first choice. We recommend using empirical therapeutic at a double dose for 8 weeks. This recommendation is addressed especially to patients with symptoms of GERD, esophagitis and exposure to acid reflux confirmed by esophageal pH monitoring. This monotherapy can be effective for controlling symptoms and may even solve the esophageal motor disorder. (9, 19,20)

Smooth Muscle Relaxants

Nitrates and calcium antagonists appear to reduce the pressure of the lower esophageal sphincter and amplitude of esophageal contraction. These drugs have been studied for use in achalasia and distal esophageal spasms with slight improvements of symptoms and improvements in manometric findings, but have not yet been scientifically tested for treatment of jackhammer esophagus. Another group of drugs with effects similar to those of nitrates are 5-phosphodiesterase inhibitors such as Sildenafil, but evidence for their use in treating hypercontractile disorders is also weak,
Endoscopic injection of botulinum toxin has proven useful for relief of chest pain in various studies. The dose varies from 80 U to 260 U, and it may be injected at different sites within the esophagus or at the gastroesophageal junction. Some patients may suffer recurrences in which cases repeated injections of botulinum toxin are required. (26,27)

POEM (Peroral Endoscopic Myotomy)

POEM (peroral endoscopic myotomy) has been proposed for treating patients with hypercontractile esophageal disorders that are refractory to medical management and who have chest pain and dysphagia. Success has been recently reported for patients with nutcracker esophagus, diffuse esophageal spasms and jackhammer esophagus although no data are available regarding long-term outcomes. (28, 29)

Surgical Management

For patients with both GERD and jackhammer esophagus, Nissen fundoplication is an alternative therapeutic option to PPIs. Retrospective studies have shown improvement in symptoms and diminished impairment of esophageal contractility. (9) At present, despite case reports, Heller’s myotomy is not considered to be standard management for patients with hypercontractile disorders, so risks and benefits of this intervention must be carefully assessed.
CONCLUSION

Jackhammer esophagus is a rare disorder of esophageal contractility. The clinical picture of patients who suffer from this disorder varies from GERD symptoms to chest pain and dysphagia. The gold standard for diagnosis is high resolution esophageal manometry. According to the criteria of the previous version of the Chicago classification, a single altered wave indicates a diagnosis of jackhammer esophagus, but according to the new 2015 Chicago v.3 version, there must be at least two waves with DCIs over 8,000 mm Hg/cm/s. For patients who have this disorder, the manometric path also includes intrabolus pressures and high amplitude multi-peak high waves that reinforce the diagnosis of jackhammer esophagus. Similarly, and as has been mentioned in larger series, a large proportion of these patients had symptoms typical of GERD refractory to treatment with PPIs, which is consistent with the evidence suggesting a cause and effect relationship between the two entities. There is no consensus about treatment, but it appears that reduction of exposure to acid improves symptoms and motor disorders. If the predominant symptoms are chest pain and dysphagia, management with neuromodulators and smooth muscle relaxants should also be provided. Surgical and endoscopic management should be reserved for patients with severe and refractory symptoms.

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REFERENCES

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