A Case Report of Jackhammer Esophagus

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Introduction
The patients with presentation of noncardiac chest pain or dysphagia can have hypertensive esophageal peristaltic contractions. Jackhammer esophagus is a term describing a subset of this entity who have extremely high-amplitude contractions and intact peristalsis. The extreme distal contractile integral (DCI) value of Jackhammer esophagus on manometric studies has not been encountered in control subjects, and can distinguish this specific diagnosis from other dysphagia entities. Here, we present a rare case of Jackhammer esophagus with initial presentation of chronic dysphagia.

Case History
A 60-year-old Caucasian female visited the gastroenterological clinic for a history of progressive dysphagia. Patient stated that the symptoms started 9 years ago and got worse after a gastric bypass surgery done in 2011. She described the dysphagia as being initially to solids (food and pills) and then it progressed to include liquids, especially after food gets lodged in the chest. She also reported chest pain and odynophagia only after food gets “stuck”. She had no associated nausea but inserting her finger into her throat to induce vomiting would usually relieve her symptoms. She had no abdominal pain, heartburn, gas, constipation, diarrhea, rectal bleeding or change in bowel habits. She was recommended to eat small meals at slower rates and was placed on Omeprazole 20 mg daily by her primary care physician, but without any appreciable relief of her dysphagia.

On review of systems, patient denied fever, weight loss, chest pain, cough or dyspnea. She did not report any neurological or respiratory symptoms. She is a current smoker with 20 year history of smoking at least half a pack daily. She denies any alcohol or drug use and drinks at least 1 caffeine drink a day. Past medical history includes diabetes mellitus (DM Type 2), hyperlipidemia, and myocardial infarction with stent placement in 2007. Past Surgical history includes gastric bypass in 2001 and 2 hiatal hernia repairs. Her home medications included metformin and glipizide, atorvastatin and metoprolol for cardiovascular diseases, and omeprazole for dysphagia.

Upon physical examination (PE), the patient was afebrile with stable vital signs. She weighed 90 Kg with a BMI of 33.07. On abdominal exam, there was a midline surgical scar; abdomen was soft, non-distended and non-tender, no masses or organomegaly were palpable. Bowel sounds were present and normal. Other exam findings were unremarkable. Her laboratory results indicated normal complete blood count and comprehensive metabolic panel except high blood glucose of 157 mg/dL.

Patient had esophagogastroduodenoscopy (EGD), which showed a small hiatal hernia with changes of mild esophagitis at the gastro esophageal junction with normal middle third of the esophagus. Her previous gastric surgery left her with a cardia and fundus which appeared normal and then a patent gastrojejunosotomy. She also had an intact gastric antrum pylorus and duodenum. Pathology results reported the change of intestinal metaplasia in the esophageal mucosa, which is consistent with Barrett’s esophagus without neoplasia or dysplasia, and chronic gastritis with mild gastritis changes. Her barium swallow showed a 3cm sliding hiatus hernia without signs of reflux or Schatzki ring and gastric emptying assessed scintigraphically by an isotope-labelled meal showed rapid gastric emptying.

She had an esophageal motility study with high-resolution manometry (HRM) and esophageal pressure topography (EPT). The mean distal contractile integral (DCI) value of each contraction sequence following based on 15 wet swallows was identified as well as the maximal DCI contraction amplitudes for any one swallow. The swallow with the greatest DCI was further characterized for maximal peristaltic amplitude. Mean basal lower esophageal sphincter (LES) pressure and Integrated Relaxation Pressure (IRP) were also assessed. This patient had normal upper and lower esophageal sphincter pressure, but the relaxation of LES was incomplete, less than 50% with swallows. Following wet swallows there was 100% peristalsis, but the contraction amplitudes were extremely elevated, with a mean DCI of 8,620 mmHg-cm-s and the maximum DCI of 25,942 mmHg-cm-s (normal <8,000 mmHg-cm-s). There was one contraction amplitude exceeded DCI of 20,000 mmHg-cm-s. Most contractions have double or triple peaks (73%, normal <10%), the diagnosis of “Jackhammer esophagus” was given based on current Chicago classification of HRM. (Figure 1) A barium swallow with a 13mm barium tablet noted the barium tablet lodged in mid esophagus with small amount of fluid ingestion, but the tablet passed with more fluid ingestion.

The impaired relaxation of the LES suggested a degree of “esophageal outlet obstruction,” therefore the therapy plan was to dilate the LES and thus improve esophageal emptying. Esophageal dilation was performed using a through the scope technique with a 20mm balloon dilator (60 French chamber). After positioning endoscopically at the GE junction, the dilator was inserted via the endoscopic channel and then the balloon was inflated on 3 separate occasions, 1 minute each time. (Figure 2) The dilation site was examined and had mild mucosal trauma as expected. No other com—

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Discussion
The “Jackhammer Esophagus” is an uncommon phenomenon identified in non-cardiac chest pain patients. In one study in the literature, 4.1% of all patients referred for manometric evaluation met the criteria of “Jackhammer Esophagus” diagnosis. These patients usually have normal distal contractile latency and normal peristaltic contractile velocity, but have extreme hypercontractility in the smooth muscle portion of the esophagus, particularly in the distal third of the esophageal body with accompanying multipeaked characteristics.

In the literature, the majority of patients (70%) with hypercontractility presented with dysphagia or chest pain, which could also be accompanied by gastroesophageal reflux symptoms. Our patients present with chronic dysphagia for 7 months before the diagnosis of Jackhammer Esophagus. In addition, the finding of Barrett’s esophagus indicated that GE reflux was occurring.

Radiographic findings are quite variable in patients suspected of Jackhammer esophagus. The barium radiographic picture includes a broad spectrum from reporting “tertiary” or non-propagating contractions to question of “spastic contractions” to entirely normal. Thus, these tests are neither specific nor sensitive for Jackhammer Esophagus.

Manometric study is the main diagnostic method for Jackhammer esophagus. Since the introduction of high-resolution manometry (HRM) and esophageal pressure topography (EPT), a mean distal contractile integral (DCI) calculation in mm Hg·s·cm has supplemented the traditional peristaltic contractile amplitudes to better quantitate and define distal esophageal contractility. The Chicago classification defined two DCI based diagnoses for hypertensive peristalsis: A mean DCI value of 5,000 to 8,000 mm Hg·s·cm was described as “nutracker esophagus” corresponding to conventional water-perfused manometry; while “spastic nutracker” had at least a single contraction with DCI value of more than 8,000 mm Hg·s·cm in single or multipeaked contractions. This latter entity with extremely high-amplitude contractions and intact peristalsis was also referred to as “Jackhammer esophagus.”

Hypertensive lower esophageal sphincter (LES) represents a primary esophageal motility abnormality of the hypercontractile type. The hypertensive LES is defined as a resting, midrespiratory pressure greater than 45 mmHg based upon conventional manometry (CM) studies performed in normal volunteers.

The patients with Jackhammer esophagus can present as mechanical esophagogastric junction outflow obstruction, GERD, or primary esophageal muscle hypercontractility. The intact latency characteristics with preserved peristalsis also distinguishes this diagnosis from achalasia and diffuse esophageal spasm.

Currently, no specific therapies have been suggested for patients with Jackhammer esophagus, since both the actual pathophysiology and the relation of the motility findings to symptoms remain obscure. Some prospective controlled observations suggested that some pharmacologic treatments aimed at inhibiting smooth muscle contractions or injecting Botulinum toxin to relax smooth muscle may be beneficial. Recently, interventions aimed at dilating, stretching, or even “breaking” smooth muscle at the level of the LES have been utilized in the name of decreasing the resistance to emptying of the esophagus. Hence, the need for excessive esophageal contractions to overcome resistance would be addressed. The two approaches are by an endoscopically positioned balloon Ortnerian dilatation with a large diameter balloon. Other potential therapies include surgical or endoscopic myotomy. Our case illustrated the most conservative approach with an endoscopically positioned balloon of 20mm size.
(60 F). The pneumatic dilation is 30 to 40 mm size. Our patient has been benefited from this initial approach, but in the future a pneumatic dilation approach may be required.

In conclusion, Jackhammer esophagus is an uncommon but well recognized clinical entity characterized by extreme hypercontractility in the smooth muscle portion of the esophagus. This entity should be considered when assessing a presentation of noncardiac chest pain and unexplained dysphagia. Manometric study is essential to confirm this diagnosis. Intervention to relax the LES can be performed in a subset of patients with impaired relaxation of the lower esophageal sphincter and not responding to medical management targeting inhibition of esophageal smooth muscle.

REFERENCES


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