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Case Report

Progression of Jackhammer Esophagus to Type II Achalasia

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It has been suggested that patients with certain motility disorders may progress overtime to develop achalasia. We describe a 66 year-old woman who presented with dysphagia for solids and liquids for a period of 18 months. Her initial workup showed normal endoscopy and non-specific esophageal motility disorder on conventional manometry. Six months later, due to persistence of symptoms, the patient underwent a high resolution esophageal manometry (HREM) demonstrating jackhammer esophagus. The patient was treated with a high dose proton pump inhibitor but without resolution of her symptoms. During the last year, the patient reported repeated episodes of food regurgitation and a significant weight loss. A repeat HREM revealed type II achalasia. Multiple case reports, and only a few prospective studies have demonstrated progression from certain esophageal motility disorders to achalasia. However, this report is the first to describe a case of jackhammer esophagus progressing to type II achalasia.
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Key Words
Esophageal achalasia; Esophageal motility disorders; Esophagus; Manometry

Introduction

Achalasia is a primary esophageal motor disorder of unknown etiology characterized by selective loss of inhibitory neurons in the esophageal wall, resulting in insufficient relaxation of the lower esophageal sphincter and loss of esophageal peristalsis.[1,2] Jackhammer esophagus is a newly introduced term to describe patients with at least 20% of the swallows with distal contractile integral (DCI) ≥ 8000 mmHg.sec.cm.[3] Several reports have described patients with diffuse esophageal spasm (DES), non-specific esophageal motor disorder (NSEMD), nutcracker esophagus, and gastroesophageal reflux disease (GERD) progressing to achalasia.[4-9] Although no causal relationship has been identified, these reports suggest that the different esophageal motor disorders represent a spectrum rather than unique and stable disorders. We describe, for the first time, a case of a patient who progressed from jackhammer esophagus to type II achalasia.

Case Report

A 66 year-old woman with a history of atrial fibrillation, hypertension, gastritis, and hypothyroidism presented to our clinic in July 2014 with progressive dysphagia for solids and liquids over 18

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months. Her symptoms occurred daily and resulted in an 11-kg weight loss over this period of time. She denied heartburn, regurgitation, choking or coughing during eating, chest pain, vomiting, abdominal pain, or change in bowel habits. Her medications included levothyroxine, metoprolol, pantoprazole, and warfarin. In June 2013, she underwent a barium swallow, upper endoscopy, and conventional manometry. The barium swallow showed a mildly dilated esophagus with tertiary contractions, delayed emptying of the esophagus, and a narrowed gastroesophageal junction (Fig. 1). The upper endoscopy demonstrated mild-moderate antral gastritis, normal appearing esophageal mucosa, and it was noted that the patient might have a tight gastroesophageal junction. The patient then underwent conventional manometry which was consistent with NSEMD. The patient was initiated on pantoprazole 40 mg daily. Unfortunately, after completing these tests she missed 3 follow-up appointments and was seen in our clinic only one year later.

Given her progressive dysphagia symptoms and weight loss, a high resolution esophageal manometry (HREM) was performed (Fig. 2). The study showed a normal median integrated residual pressure of 8 mmHg, normal mean resting pressure of 21 mmHg, normal mean residual pressure of 7 mmHg, and mean DCI of 10 770 mmHg·sec·cm. Of the 10 swallows, 30% were hypercontractile (> 8000 mmHg·sec·cm), 50% were normal and 20% simultaneous. Overall, the findings were consistent with jackhammer esophagus based on the Chicago Classification of Motility Disorders.[3] Pantoprazole was increased to 40 mg twice daily with plan to add a pain modulator if her symptoms recurred.

Unfortunately, she was lost to follow-up again until she was seen in our clinic a year later in June 2015. She continued to have worsening dysphagia, lost an additional 7 kg, and reported new onset of occasional post-prandial chest tightness. At this point, a timed barium esophagram was performed (Fig. 3). This study demonstrated diffuse esophageal dilatation with retained secretions and barium with only 0-20% change in esophageal volume after 5 minutes. In addition, tertiary peristaltic waves within the distal esophagus, limited emptying of the contrast into the stomach and an air-fluid level were also noted. A repeat HREM was performed (Fig. 4) revealing a median integrated residual pressure of 71.5 mmHg, mean resting pressure of 97 mmHg, and mean residual pressure of 90 mmHg. There was panesophageal pressurization with every swallow and 80% of the swallows demonstrated prolonged pan-pressurization, all consistent with type II achalasia. Therapeutic options were discussed with the patient, and she elected to undergo Heller myotomy. The patient was referred to General Surgery but presented urgently to GI clinic 2 weeks later for worsening dysphagia, dehydration, weakness, inability to tolerate oral intake, and an additional 7-kg weight loss. The patient was admitted to the hospital and was noted to develop acute kidney injury. The patient underwent aggressive volume resuscitation. In addition, a nasogastric tube was placed and she was started on tube feeds to optimize her nutritional status. A week after discharge the

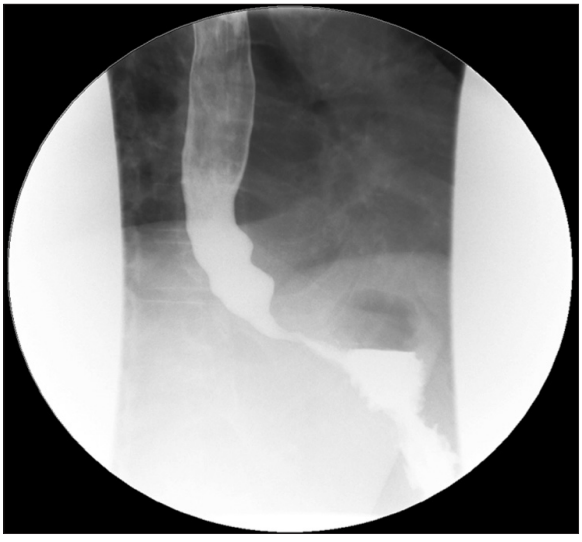

Figure 1. Patient's initial barium swallow which showed a mildly dilated esophagus with tertiary contractions, delayed emptying of the esophagus, and a narrowed gastroesophageal junction.

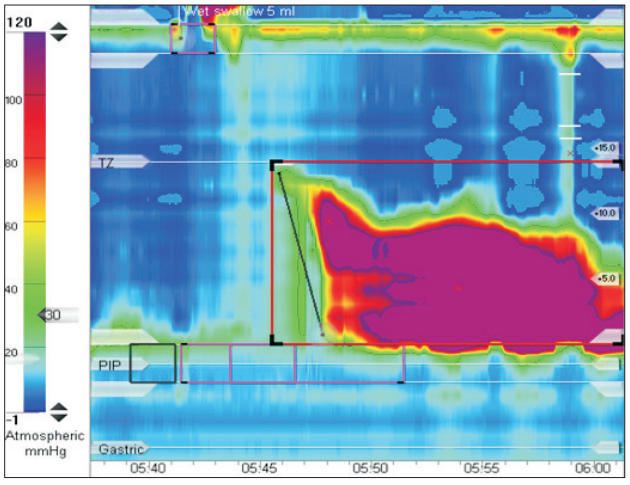

Figure 2. A representative swallow from the patient's initial high resolution esophageal manometry. The median integrated residual pressure was noted to be normal at 7 mmHg, the mean lower esophageal sphincter resting pressure was normal at 20 mmHg, and the distal contractile integral was elevated to 16,534 mmHg·sec·cm. The esophageal manometry was consistent with jackhammer esophagus.

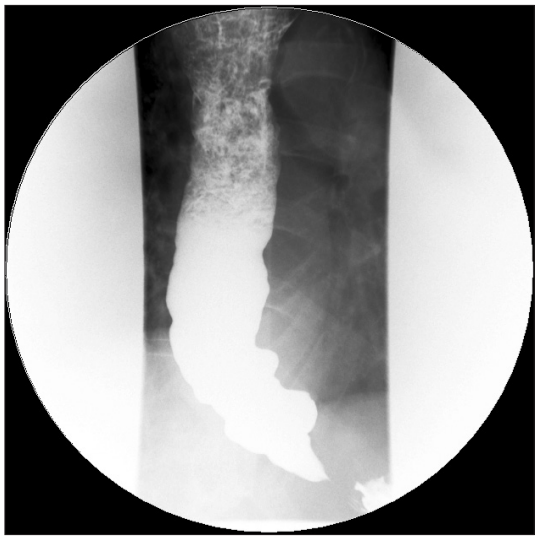

Figure 3. Patient's timed barium esophagram. There is a diffuse esophageal dilatation with retained esophageal secretions and barium with only 0-20% change in volume after 5 minutes. There are tertiary peristaltic waves within the distal esophagus, limited emptying of the contrast into the stomach and an air-fluid level.

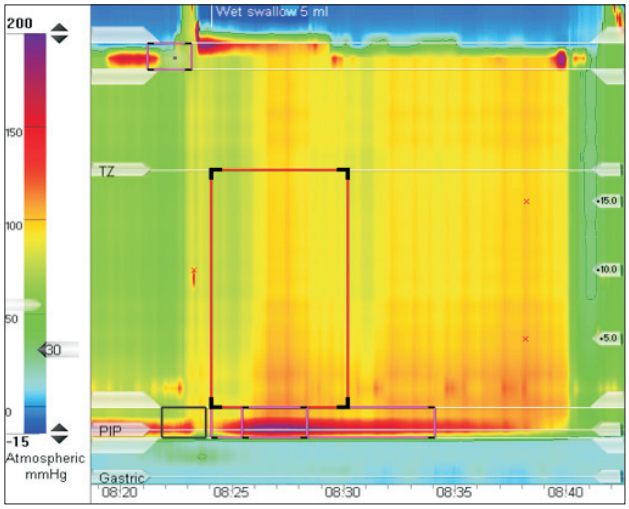

Figure 4. A representative swallow from the patient's repeat high resolution esophageal manometry about a year later. The median integrated residual pressure was noted to be elevated at 143 mmHg and the mean lower esophageal sphincter resting pressure was also elevated at 122 mmHg. There is a 100% prolonged panesophageal pressurization. The esophageal manometry was consistent with type II achalasia.

patient underwent Heller myotomy with Dor fundoplication. On a follow-up visit with her general surgeon, she reported that she was tolerating liquids without any difficulties. The plan at that time was to initiate a soft diet.

Discussion

To our knowledge, this is the first case that describes progression of jackhammer esophagus to achalasia over a period of about one year. Several reports have described the progression of several esophageal motility disorders to achalasia. Smart et al[5] reported 5 patients with GERD who subsequently developed achalasia over a period of 2-10 years. Robson et al[4] reported a patient with GERD who developed DES one year after initial diagnosis and achalasia one year later. Several reports describe the progression of DES to achalasia.[6-8] Khatami et al[10] conducted the first prospective cohort study with 12 patients. They observed the progression of DES to achalasia in only one patient after 10.6 years. They also noted that low esophageal body amplitude contractions was a predictor of this progression. Fontes et al[11] conducted the largest prospective study to date to assess the progression of DES to achalasia. Thirty-five patients with DES were followed for at least one year. Patients with GERD confirmed by pH monitoring or systemic diseases that may affect the esophagus were excluded. Five (14%) of the patients progressed to achalasia at a mean follow-up of 2.1 years. Although demographic characteristics were not predictive of the transition to achalasia, the authors observed that amplitude contractions of less than 50 mmHg was a predictive factor ($P = 0.002$). Anggianash et al[9] described the progression of nutcracker esophagus to achalasia in one patient after 3 years. Paterson et al[12] also described this progression in one patient after 2 years. Vantrappen et al[13] described six patients with NSEMD who progressed to achalasia.

While our patient presented with symptoms that could be considered classic for achalasia, her initial conventional manometry was consistent with NSEMD. The first HREM demonstrated jackhammer esophagus rather than achalasia. Only the second HREM as well as the timed barium esophagram were consistent with achalasia. This progression within one year of an esophageal motor disorder was faster than what has been documented in previous reports.

The underlying mechanism behind this progression is still unknown. However, this case suggests that all esophageal motility disorders represent a spectrum of disorders, where patients may progress over time from one to another. This case also demonstrates the importance of following patients who were diagnosed with non-achalasia motility disorders with a repeat HREM in one year to assess for progression or a shift to a different esophageal motility disorder. Because most of the studies documenting progression

of a motility disorder to achalasia were case reports using conventional manometry, long-term studies evaluating this progression with HREM are needed. Perhaps with the current availability of HREM, researchers may be able to identify predictive factors associated with progression of certain esophageal motility disorders to achalasia.

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Conflicts of interest: None.

Author contributions: Jason Abdallah collected references and prepared the manuscript; and Ronnie Fass edited and supervised the preparation of the manuscript.

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