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Letter to the Editor

Progression of Jackhammer Esophagus to Type III Achalasia and Improvement After Extended Myotomy

Introduction

Jackhammer esophagus is manometrically defined as at least 2 swallows with distal contractile integral ≥ 8000 mmHg·sec·cm. Achalasia is an uncommon primary esophageal motor disorder of unknown etiology characterized by incomplete or absent relaxation of the lower esophageal sphincter and loss of esophageal peristalsis.[1] According to Chicago classification version 3.0[2] there are 3 types of achalasia. Type III or spastic achalasia is characterized by premature contractions with distal contractile integral > 450 mmHg·sec·cm with > 20% of swallows. Early recognition of the disease is improved with high-resolution manometry (HRM).

Since its introduction by Inoue et al,[3] peroral endoscopic myotomy (POEM) has become a first-line established treatment for achalasia when an expert operator is available.[4] It is especially useful in type III achalasia, which is associated with the worst outcome with all the classical treatments.[5]

Case Report

A 66-year-old patient who presented with a history of daily intermittent esophagus dysphagia for solids and liquids over 15 months associated with food regurgitation, retrosternal pain and weight loss. This clinical condition had only a partial response to medical therapy. Gastroscopy with esophageal biopsies, barium esophagogram and 24-hour pH monitoring did not show any significant pathological findings. Solid state HRM (Manoscan, Medtronic Inc, Minneapolis, MN, USA) with impedance showed a hypercontractile esophagus and impaired relaxation in some of the swallows. Viscous bolus clearance was also impaired (Fig. 1A).

After botulinum toxin injection the retrosternal pain improved but the dysphagia got worse. A new HRM showed a hypertonic lower esophageal sphincter with a lack of relaxation (integrated relaxation pressure > 15 mmHg) with panesophageal pressurization associated and hypercontractile contractions. These changes suggested a progression to a spastic achalasia (Fig. 1B). A thoracic computerized tomography was normal. Surgical myotomy was proposed to the patient as POEM was not available. The myotomy was prolonged according to the HRM findings. A 16 cm Heller myotomy was performed associated to an anti-reflux technique (Toupet).

The symptoms improved, especially the dysphagia. Postsurgical HRM with impedance showed the disappearing of the previous hypercontractile waves preserving normal peristalsis until the upper limit of the myotomy. Also was observed a better relaxation of the gastroesophageal junction, with an integrated relaxation pressure at the high limit of normal because of the anti-reflux surgery. However, an incomplete bolus clearance was observed in all the swallows (Fig. 2).

Discussion

In the literature, there are multiple case reports[6,7] and a few prospective studies that have demonstrated progression from certain esophageal motility disorders to achalasia, including jackhammer esophagus. According to the study of Huang et al[8] patients with concomitant jackhammer esophagus and impaired EGJ relaxation are more likely to develop achalasia, especially if they present with intermittent bolus clearance in the impedance exam and with dysphagia as the presenting symptom. If these patterns are found, as in our case, more aggressive treatment may be warranted to avoid deterioration of esophageal function. However, more studies are needed to confirm these findings. Spastic disorders of the esophagus seem to have a good response to longer miotomy.[9]

The underlying mechanism behind this progression is still unknown, but there are some hypotheses that suggest that all esophageal motility disorders represent a spectrum[6] rather than a unique

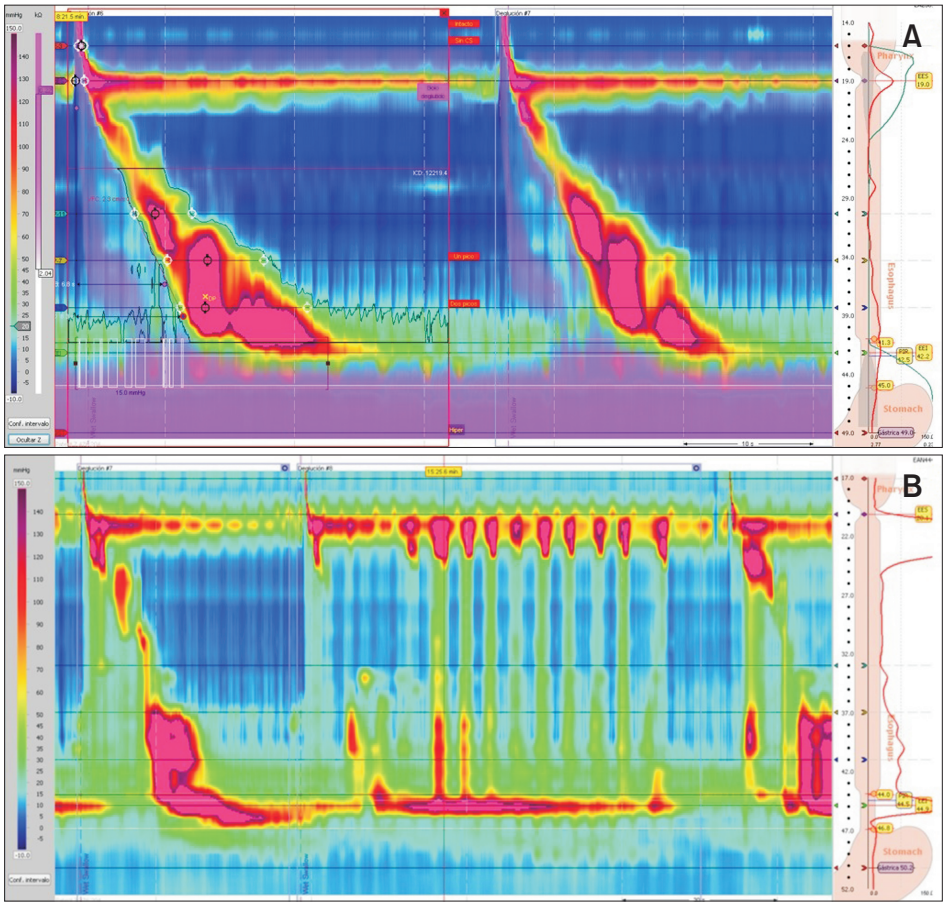

Figure 1. (A) High-resolution manometry (HRM) with impedance initially performed on the patient that shows 2 swallows. Mean distal contractile integral (DCI) is 8882.6 mmHg·sec·cm (> 8000 mmHg·sec·cm) and integrated relaxation pressure (IRP) is 14.8 mmHg (< 15 mmHg). The study is consistent with jackhammer esophagus. Liquid bolus clearance was complete but viscous was incomplete. (B) HRM after treatment with botulinum toxin. DCI persists over 8000 mmHg·sec·cm but IRP is now 33.6 mmHg (> 15 mmHg) and panpressurization appears. These new findings suggest a progression from the previous motor disorder to type III achalasia.

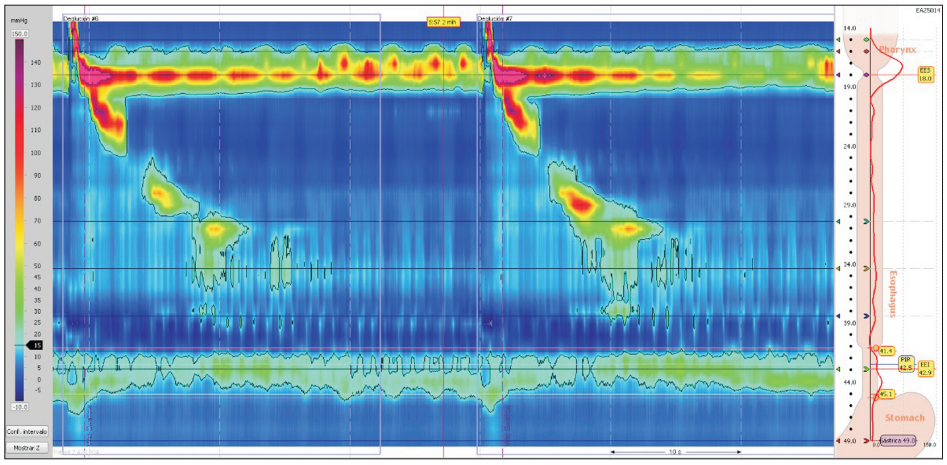

Figure 2. High-resolution manometry performed after extended myotomy. No hypercontractile waves are observed (distal contractile integral < 8000 mmHg·sec·cm), with absence of peristalsis in the myotomy zone that has a length of 16 cm. Integrated relaxation pressure is 14.8 mmHg and bolus clearance is incomplete related to the absent peristalsis below the myotomy.

and stable disorder, where patients may progress over time from one to another. Therefore, it is very important to follow these patients who were diagnosed with non-achalasia motility disorders to assess for progression or a shift to a different esophageal motility disorder.

It is also important to exclude opioid and ethanol consumption as they can favor the development of esophageal motility disorders such as achalasia type III,[10] but our patient refused the consumption of any of these substances.

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1. Vaezi MF, Pandolfino JE, Vela MF. ACG clinical guideline: diagnosis and management of achalasia. Am J Gastroenterol 2013;108:1238-1249.
2. Kahrilas PJ, Bredenoord AJ, Fox M, et al. The Chicago classification of esophageal motility disorders, v3.0. Neurogastroenterol Motil 2015;27:160-174.
3. Inoue H, Minami H, Kobayashi Y, et al. Peroral endoscopic myotomy (POEM) for esophageal achalasia. Endoscopy 2010;42:265-271.
4. Akintoye E, Kumar N, Obaitan I, Alayo QA, Thompson CC. Peroral endoscopic myotomy: a meta-analysis: a meta-analysis. Endoscopy 2016;48:1059-1068.
5. Salvador R, Provenzano L, Capovilla G, et al. Extending myotomy both downward and upward improves the final outcome in manometric pattern III achalasia patients. J Laparoendosc Adv Surg Tech A Published Online First: 20 Mar 2019. doi: 10.1089/lap.2019.0035.
6. Abdallah J, Fass R. Progression of jackhammer esophagus to type II achalasia. J Neurogastroenterol Motil 2016;22:153-156.
7. Usai Satta P, Oppia F, Piras R, Loriga F. Extrinsic autonomic neuropathy in a case of transition from diffuse esophageal spasm to achalasia. Clin Auton Res 2004;14:270-272.
8. Huang L, Pimentel M, Rezaie A. Do jackhammer contractions lead to achalasia? a longitudinal study. Neurogastroenterol Motil Published Online First: 23 Sep 2016. doi: 10.1111/nmo. 12953.
9. Khan MA, Kumbhari V, Ngamruengphong S, et al. Is POEM the answer for management of spastic esophageal disorders? a systematic review and meta-analysis. Dig Dis Sci 2017;62:35-44.
10. Schindler V, Runggaldier D, Bianca A, et al. Opioid treatment and excessive alcohol consumption are associated with esophagogastric junction disorders. J Neurogastroenterol Motil 2019;25:205-211.

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