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Peroral endoscopic myotomy in pediatric jackhammer esophagus

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Peroral endoscopic myotomy in pediatric jackhammer esophagus

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ABSTRACT

The jackhammer esophagus is a rare hypercontractile disorder and diagnosis is based on high-resolution manometry. Peroral endoscopic myotomy (POEM) of the spastic esophagus segments has been described. We report a pediatric patient with jackhammer esophagus that was treated endoscopically.

Key words: Peroral endoscopic myotomy. Jackhammer esophagus. Pediatric dysphagia. High-resolution manometry.

INTRODUCTION

The jackhammer esophagus is a rare hypercontractile disorder clinically characterized by the presence of recurrent retroesternal pain or dysphagia (1). The diagnosis is based on high-resolution manometry (HRM) defined by distal contractile integrals (DCI) greater

than 8,000 mmHg/s/cm in at least 20% of the evaluated swallows. Despite the physiopathological differences with regard to achalasia, similar therapeutic options have been suggested (2,3). Peroral endoscopic myotomy (POEM) of the spastic esophagus segments has been described (4-6). Here, we report a pediatric patient with jackhammer esophagus that was treated endoscopically.

CASE REPORT

A 14-year-old female patient, 155 centimeters in height, 43 kg in weight (percentile 25), referred to our clinic due to retrosternal pain associated with ingestion and a 5-year evolution of dysphagia. Both gastroesophageal and eosinophilic esophagitis were ruled out; the former via a 24-hour pH-impedance test and the latter, by endoscopic biopsies of the three esophageal thirds after treatment with proton pump inhibitors.

A HRM was performed and increased esophageal contractions were observed in each of the ten liquid and solid swallows studied. Peristalsis was preserved, with an average DCI of 27,000 mmHg/s/cm and a proper relaxation of the lower esophageal sphincter (LES) (Fig. 1).

An endoscopic therapeutic attempt was made via the helical injection of botulinum toxin in the esophageal body. However, there was no response. Inoue and other authors suggest performing a POEM of the spastic segments of the esophagus as a therapeutic option in patients with jackhammer esophagus (7,8). Thus, a POEM of these segments was performed after an antibiotic prophylaxis that was previously determined by the HRM whilst taking care of the LES. There were no complications during the procedure. A liquid diet was started 24 hours later and the patient was discharged on the third day (Fig. 2).

Four months later, the patient was asymptomatic. A control HRM showed the absence of contractions in the smooth muscle with an average DCI of 1,180 mmHg/s/cm (Fig. 3).

DISCUSSION

The manometric criteria for the hypercontractile esophagus varied over time. It started with the “nutcracker esophagus”, defined via conventional manometry by a distal wave

amplitude of ≥ 180 mmHg with preserved peristalsis. Afterwards, this value was increased to 220 mmHg in order to improve the diagnostic specificity.

With the introduction of HRM, a new classification of the motor disorders and new metric units have been incorporated (Chicago International Classification). The DCI allows for a better measurement of the activity of the smooth muscle and the vigor of the contraction. The Chicago International Classification (version 3.0) (9) defines jackhammer esophagus as the presence of hypertensive waves with more than 8,000 mmHg/s/cm in at least 20% of the evaluated swallows. However, this characteristic is not exclusive and can be found in patients with gastroesophageal reflux, outflow obstruction and primary hypercontractility. The diagnostic criteria for Jackhammer esophagus are the same for the pediatric population (10).

Here, we describe the successful endoscopic treatment of a pediatric patient with jackhammer esophagus. It is necessary to perform prospective and randomized studies to confirm these findings.

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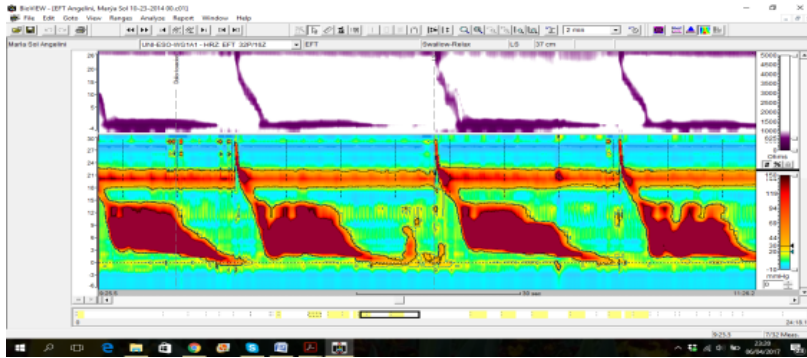


Fig. 1. HRM: increased esophageal contraction. DCI 23,000 mmHg/s/cm.



Fig. 2. POEM. A. Mucosal incision to initiate submucosal tunneling. B. Panoramic view of the submucosal tunnel with a completed longitudinal myotomy. C. Mucosal incision closure with clips.

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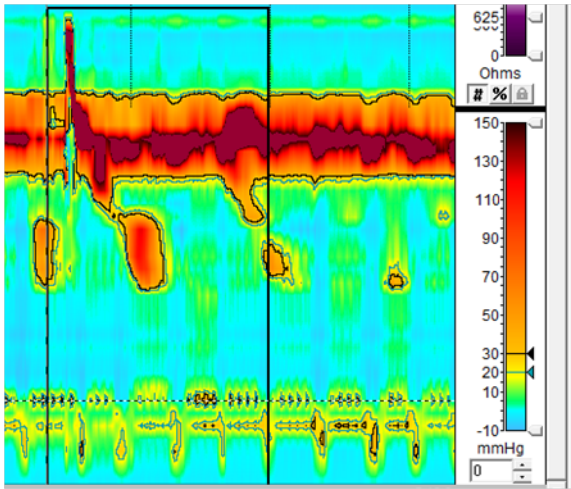


Fig. 3. HRM: no contraction of the smooth muscle after endoscopic treatment.