Clinical outcomes and safety of high-resolution manometry guided superficial partial circular muscle myotomy in per-oral endoscopic myotomy for Jackhammer esophagus: Two cases report

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Abstract

BACKGROUND
Jack hammer esophagus is a relatively rare disease and to date, there is no dramatic treatment option. Recently, conventional per-oral endoscopic myotomy (POEM) have expanded their area into Jackhammer esophagus. However, several complications such as post procedure motility disorders (e.g., passage disturbance) are issues after POEM. To overcome these issues, we here introduced high-resolution manometry (HRM)-guided superficial partial circular muscle myotomy, which involves cutting only the superficial layer of the esophageal circular muscle.

CASE SUMMARY
We report two cases of patients with Jackhammer esophagus who were treated with HRM-guided extremely superficial partial circular muscle myotomy during POEM. Case 1 was a 53-year-old female with medication-refractory odynophagia and case 2 was a 47-year-old man who presented with chest pain. They were diagnosed with Jackhammer esophagus using HRM, and the hypercontractile segments of the esophagus were identified. HRM-guided extremely superficial partial circular muscle myotomy was performed while preserving the lower esophageal sphincter. Therefore, the circular and longitudinal muscle layers are preserved but hypercontractile movements are reduced, even after POEM. Patients’ clinical symptoms dramatically improved right after POEM, and 6-mo follow-up HRM revealed completely resolved status. During a 1-year follow-up period, patients were still in good health and remained symptom free.

CONCLUSION
HRM-guided superficial partial circular muscle myotomy may be a promising treatment option for Jackhammer esophagus.
alternative to conventional POEM for treating Jackhammer esophagus with improved efficacy.

Key words: Jackhammer esophagus; Hypercontractile; Partial circular muscle myotomy; Case report

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Core tip: Jack hammer esophagus is a relatively rare disease and to date, there is no dramatic treatment option. Recently, conventional per-oral endoscopic myotomy (POEM) have expanded their area into Jackhammer esophagus. However, several complications such as post procedure motility disorders (e.g., passage disturbance) are issues after POEM. To overcome these issues, we here introduced high-resolution manometry-guided superficial partial circular muscle myotomy, which involves cutting only the superficial layer of the esophageal circular muscle for two patients.

INTRODUCTION

Jackhammer esophagus, also referred to as hypercontractile peristalsis, is a rare esophageal motility disorder characterized by hypertensive but normally propagated peristaltic contractions[1-3]. The manometric criteria for Jackhammer esophagus are an initial average peristaltic amplitude > 180 mmHg in the distal esophagus using conventional manometry[1,3,4] > 20% of swallows having a distal contractile integral (DCI) value > 8000 mmHg.s.cm, and normal latency on high-resolution manometry (HRM)[2,5-8]. The therapeutic options for Jackhammer esophagus are pharmacologic agents such as nitrates, phosphodiesterase 5 inhibitors, low-dose antidepressants, proton pump inhibitors, and endoscopic botulinum toxin injection into the esophageal body. However, the efficacy of these methods is not satisfactory[1-3,9,10]. Per-oral endoscopic myotomy (POEM) has been used as an alternative treatment to overcome the limitations of the above therapies. However, there are still concerns regarding post-POEM complications, such as passage disturbance and sigmoid esophagus[2,11-14].

Jack hammer esophagus is a relatively rare disease and to date, there is no definitive and dramatic treatment options, including medication, endoscopic treatments or surgical treatments.

To reduce the risk of complications after conventional POEM[11,19], we introduced HRM-guided extremely superficial partial circular muscle myotomy during the POEM procedure for two Jackhammer esophagus cases. Partial circular muscle myotomy involves cutting only the superficial layer of the esophageal circular muscle. Therefore, the circular and longitudinal muscle layers are preserved but hypercontractile movements are reduced, even after POEM. Moreover, hypercontractile segments were specifically targeted and measured though HRM.

Between April 2016 and August 2018, a total of 350 patients underwent HRM and 8 were diagnosed with Jackhammer esophagus in our hospitals. Two patients with medication-refractory Jackhammer esophagus underwent partial circular muscle myotomy during POEM. Herein, we describe two patients who presented with Jackhammer esophagus and were successfully treated using HRM-guided partial circular muscle myotomy during POEM.

CASE PRESENTATION

Chief complaints

Case 1: A 53-year-old woman was referred to our hospital for odynophagia and regurgitation.
Case 2: A 47-year-old man was referred to the gastrointestinal department for atypical chest pain for 6 mo.

History of present illness
Case 1: She had previously presented to a local hospital and had been prescribed oral proton pump inhibitors and nitroglycerin for several months, but her symptoms did not improve.

Case 2: He was previously presented to the cardiovascular department for atypical chest pain. However, his symptom was not improved even after stent angiography and administration of anti-angina medications and oral proton pump inhibitors for several months. After then he was referred to the gastrointestinal department.

History of past illness/ Personal and family history
Case 1: She had no known medical or surgical history. Her family history was negative.

Case 2: He had two-vessel cardiovascular disease with angina and a stent was inserted 5 years ago. Despite use of a patent stent, cardiovascular medications including nitrates, and a 3-mo trial of proton pump inhibitors, atypical squeezing pain in the epigastric region remained. His family history was negative.

Physical examination upon admission
Case 1: Physical examination on admission revealed no abnormal palpable mass on head and neck area.

Case 2: Physical examination on admission revealed no abnormal findings.

Laboratory examinations
Case 1: There were no abnormal findings on electrocardiogram, and laboratory tests including total blood count, liver function, renal function, and other basic chemical tests were normal.

Case 2: Laboratory tests were normal, including total blood count, liver function, renal function, and other basic chemical tests.

Imaging examinations
Case 1: Esophagography (barium radiography) showed spasmodic contraction of the distal esophagus and a narrowing of the esophageal cavity (Figure 1). HRM showed high-amplitude distal esophageal contractions with a DCI value > 8000 mmHg.s.cm for 6 of a total of 10 swallows. The highest DCI value was 13553 mmHg.s.cm (Figure 1). HRM showed high-amplitude distal esophageal contractions located 25-38 cm from the incisors according to the distance gauge of the pressure measuring tubes.

Case 2: On upper endoscopy, no abnormal findings were reported. Barium radiography showed spasmodic contractions of the distal esophagus and a narrow esophageal cavity (Figure 2). HRM showed high-amplitude distal esophageal contractions with a DCI value > 8000 mmHg.s.cm for 8 of a total of 10 swallows (Figure 2B). The highest DCI value was 21024 mmHg.s.cm. POEM was performed. HRM showed high-amplitude distal esophageal contractions located 28-39 cm from the incisors according to the distance gauge of the pressure measuring tubes.

FINAL DIAGNOSIS
Case 1: The final diagnosis was medication refractory Jack hammer esophagus without involvement of low esophageal sphincter.

Case 2: The final diagnosis was medication refractory Jack hammer esophagus without involvement of low esophageal sphincter.

TREATMENT
Case 1: The patient underwent HRM-guided superficial partial circular muscle myotomy (Figure 3). Since HRM showed high-amplitude distal esophageal contractions located 25-38 cm from the incisors, we performed partial circular muscle myotomy of the esophageal muscle on the right side (Figure 3). We preserved the lower esophageal sphincter.
Figure 1. The examination of 53-year-old woman conducted before superficial circular muscle per-oral esophageal myotomy. A: Pre-treatment high-resolution esophageal manometry (HRM) image depicted a jackhammer esophagus patient with distal contractile index over 8000 mmHg.s.cm; B-D: Barium radiography showed spasmodic contraction of the distal esophagus and a narrowing of the esophageal cavity (C). Post-treatment HRM revealed distal contractile integral of 980 mmHg.s.cm (D). Post-treatment esophagogram showed improved.

**Case 2:** The patient underwent HRM-guided superficial partial circular muscle myotomy (Figure 3). Since HRM showed high-amplitude distal esophageal contractions located 28-39 cm from the incisors, we performed partial circular muscle myotomy of the esophageal muscle on the right side (Figure 3). We preserved the lower esophageal sphincter.

We performed HRM-guided superficial partial circular muscle myotomy of the esophageal muscle. Since HRM showed high-amplitude distal esophageal contractions located 28-39 cm from the incisors, we performed partial circular muscle myotomy of the esophageal muscle on the right side (Figure 3) and preserved the lower esophageal sphincter.

**OUTCOME AND FOLLOW UP**

**Case 1:** After the procedure, the patient’s symptoms dramatically improved and post-POEM HRM was within the normal range. During a 1-year follow-up period, patients were in good health and remained symptom free.

**Case 2:** After the procedure, the patient’s symptoms dramatically improved and post-POEM HRM was within the normal range (Figure 2C). During a 6-mo follow-up period, patients were in good health and remained symptom free.
DISCUSSION

We here report two patients with Jackhammer esophagus who were successfully treated with HRM-guided superficial partial circular muscle myotomy during POEM. After the procedures, both patients reported improve symptoms with no side effects. The current cases suggest that HRM-guided superficial partial circular muscle myotomy may be a potential treatment option for Jackhammer esophagus with a relatively low rate of post-procedure complications as compared to conventional POEM.

Jackhammer esophagus is rare and severe disease\(^2,16,17\). Jackhammer esophagus is extremely high amplitudes contractions and within normal limit of peristatic contractions\(^4\). Treatment strategy for Jackhammer esophagus includes medication for smooth muscle relation (nifedipine), anti-reflex medication, and pneumatic dilatation of LES\(^4\). Because of the rare incidence of Jackhammer esophagus, proper evaluation of incidence is not easy, the medication refractory Jackhammer esophagus has been increasing. Recently for medical refractory Jackhammer esophagus, POEM was introduced\(^11\).

POEM is the first clinically efficacious natural orifice transluminal endoscopic surgery (NOTES) with an endoscopic safety profile\(^1,15,18,19\). However, despite its safety profile, post-POEM complications are not rare\(^6,7,9,20\). Conventional POEM for Jackhammer esophagus is associated with several side effects including post-procedure sigmoid esophagus and ineffective esophageal motility\(^15,19\). It remains debated whether the lower esophageal sphincter should be cut to prevent symptom...
Figure 3 Procedures of high-resolution manometry guided superficial partial circular muscle myotomy. A: We first detect the hypercontractile lesion through high-resolution manometry (HRM), and in this patient’s case, HRM showed high-amplitude distal esophageal contractions located 25-38 cm from the incisors according to the distance gauge of the pressure measuring tubes. B: Therefore, we performed superficial partial circular muscle myotomy of the esophageal muscle on the right side. (B) Superficial partial circular muscle myotomy during POEM. Remnant circular muscle.

Recent systemic review showed that the pooled rate of clinical success in patients of Jackhammer esophagus for POEM was 89.6%. The success rates of both the length > 10 cm, and the length < 10 cm were 91.1% and 89.1%, respectively. There are several researches on the symptom in patients with Jackhammer esophagus and the pre-peak and post-peak phase of contraction. In these regards, to distinguish the contractile integral components of pre-peak and post-peak phase contractile activity is important to treat Jackhammer disease. However, there are still concerns regarding post-POEM complications for medication refractory Jackhammer esophagus after POEM, such as passage disturbance and sigmoid esophagus.

To improve treatment efficacies and reduce the complications in the treatment of Jack hammer esophagus after POEM, we focused two issues: (1) To measure the accurate segments which are hypercontractile in the esophagus; and (2) To conserve the esophageal motility after POEM procedures. HRM-guided superficial partial circular muscle myotomy which we introduced is a modified type of conventional POEM, and this method is expected to reduce side effects and increase treatment efficacy. Because partial circular myotomy during POEM involves cutting only the superficial layer of the circular esophageal muscle and not the full thickness of the muscle nor the full circular muscle layer, even after POEM, the previously diagnosed Jackhammer esophagus consists circular and longitudinal layer of muscle with its nature but reduced hypercontractile movements. Moreover, it is not necessary to cut the lower esophageal sphincter in a partial circular myotomy when low esophageal sphincter is not involved. This method not only reduces the occurrence development after the procedure.
of side effects associated with conventional POEM, including partial or full thickness POEM, but also improves treatment efficacy.

CONCLUSION

HRM-guided superficial partial circular muscle myotomy during POEM may be a promising alternative to conventional POEM for the treatment of patients with Jackhammer esophagus who are refractory to conventional medical therapy, which is associated with improved efficacy and safety profile.

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