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Peroral Endoscopic Myotomy for Refractory Dysphagia in Esophageal Absent Contractility: A Case Report

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Mounir El bied [1, 2], Ahlam Boutaleb [1, 2], Mohamed Amine El Amghari [1, 2], Hajar Kirami [1, 2], Mehdi Zouaoui [1, 2], Mbarek Azouaoui [1, 2], Youssef Hnach [3, 2], Nourdin Aqodad [1, 2], Hassan Seddik [4, 5]

1. Department of Hepato-Gastroenterology, Souss-Massa Territorial Health Group (GST), Agadir, MAR 2. Department of Hepato-Gastroenterology, Faculty of Medicine and Pharmacy of Agadir, Ibn Zohr University, Agadir, MAR 3. Department of Hepato-Gastroenterology, Oued Eddahab Military Hospital, Agadir, MAR 4. Department of Gastroenterology II and Digestive Endoscopy, Mohammed V Military Teaching Hospital, Rabat, MAR 5. Department of Gastroenterology and Gastrointestinal Endoscopy, Faculty of Medicine and Pharmacy of Rabat, Mohammed V University, Rabat, MAR

Corresponding author: Mounir El bied, mounirelbied27061998@gmail.com

Abstract

Esophageal absent contractility is characterized by absent esophageal body contractions despite preserved relaxation of the esophagogastric junction. We describe a patient with severe dysphagia who underwent peroral endoscopic myotomy (POEM). A 55-year-old man presented with progressively worsening dysphagia since childhood, food regurgitation, and heartburn (Eckardt score: 8). Upper gastrointestinal endoscopy showed a megaesophagus with food retention and no resistance to passage through the cardia. A conventional barium swallow also showed a megaesophagus, without the distal “bird’s beak” appearance of achalasia. High-resolution manometry showed failed contractions during all 10 supine and five upright swallows, with median integrated relaxation pressures of 13.34 mmHg and 9 mmHg, respectively. A subsequent timed barium esophagram was normal. Twenty-four-hour esophageal pH monitoring performed off proton pump inhibitor therapy showed an acid exposure time of 2%. Dietary measures and a five-day course of metoclopramide did not improve dysphagia. POEM was performed, with a favorable clinical response. At the five-month follow-up, the patient was eating solid food, his Eckardt score was 0, and his weight had increased from 54 to 65 kg. Control manometry at four months showed reduced lower esophageal sphincter resting pressure and integrated relaxation pressures. This case documents short-term improvement in dysphagia and weight after POEM despite persistent absent contractility. Longer follow-up is needed to assess the durability of this response and post-operative reflux.

Categories: Therapeutics, Internal Medicine, Gastroenterology
Keywords: absent contractility, dysphagia, esophageal aperistalsis, high-resolution manometry, lower esophageal sphincter, peroral endoscopic myotomy, timed barium esophagram

Introduction

Esophageal absent contractility is a motility disorder characterized by a complete absence of esophageal body contractions despite preserved relaxation of the esophagogastric junction (EGJ). According to the Chicago Classification version 4.0, it is defined by 100% failed peristalsis (distal contractile integral <100 mmHg·s·cm) with a normal median integrated relaxation pressure (IRP) in both supine and upright positions [1]. Although a normal IRP distinguishes this pattern from classic achalasia, additional testing may be necessary when clinical, radiological, and manometric findings are discordant [1].

Peroral endoscopic myotomy (POEM) has been explored in a small number of patients with absent contractility, with preliminary reports suggesting symptomatic improvement in selected cases [2]. However, management remains challenging because no established medication reliably restores esophageal peristalsis, and treatment generally focuses on nutritional support, symptom control, and associated reflux [3]. POEM may facilitate esophageal emptying by reducing resistance at the EGJ, but it does not directly restore esophageal body contractions. Its role in this setting remains uncertain, particularly when objective testing does not demonstrate persistent EGJ obstruction [2,3]. We report the diagnostic assessment and short-term clinical and manometric outcomes of POEM in a patient with severe dysphagia and absent esophageal contractility.

Case Presentation

A 55-year-old man presented with distal esophageal dysphagia to solids, beginning in childhood and gradually worsening. He also reported food regurgitation and heartburn, without other reported digestive or extra-digestive symptoms. His baseline Eckardt score was 8, and his weight before treatment was 54 kg. He had stopped smoking (10 pack-years) and drinking alcohol 15 years earlier. There was no diabetes or implicated medication use.

A previous barium swallow had been described as showing megaesophagus (Figure 1). Subsequent thoracic computed tomography showed esophageal dilation and retained food. Upper gastrointestinal endoscopy later demonstrated esophageal dilation and food stasis without resistance at the cardia. No esophagitis or candidiasis was identified on endoscopic assessment. Esophageal biopsies showed no eosinophilic infiltration.

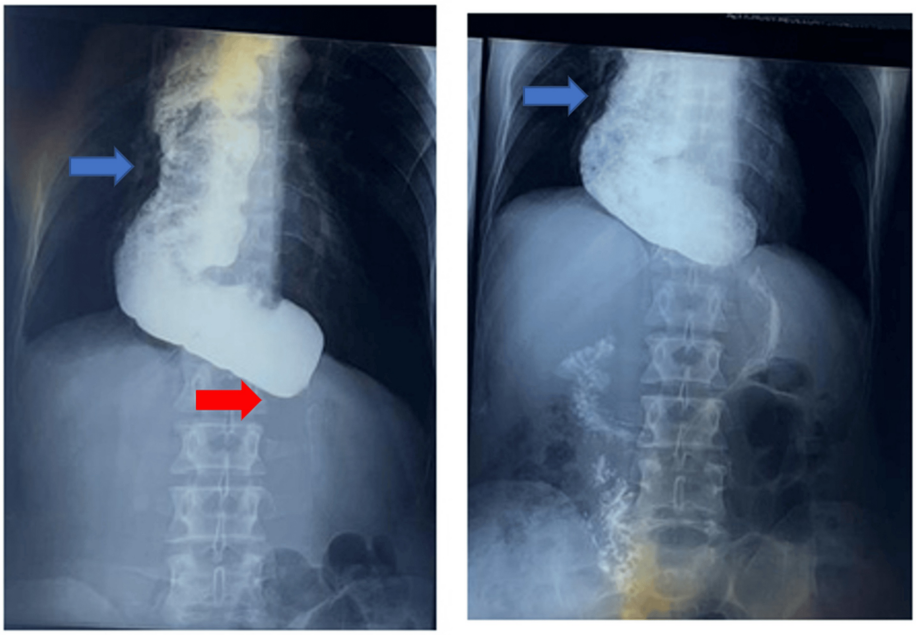


FIGURE 1: Barium esophagram showing megaesophagus (blue arrows) without an abrupt distal cutoff of the barium column at the lower esophagus (red arrow).

High-resolution esophageal manometry was performed using a Solar GI system (Hengelo, the Netherlands: Medical Measurement Systems B.V. {Laborie}). The protocol included 10 supine swallows, five upright swallows, and a rapid drink challenge. The system-specific upper limits of normal for median integrated relaxation pressure were 22 mmHg in the supine position and 15 mmHg in the upright position.

The esophagogastric junction had type I morphology, with no hiatal hernia. Basal upper and lower esophageal sphincter pressures were normal (Figure 2). During 10 supine swallows, the median integrated relaxation pressure (IRP) was 13.34 mmHg (normal for the Laborie system {Portsmouth, New Hampshire: Laborie Medical Technologies Corp.}: <22 mmHg), and esophageal contractions were absent in all swallows. During five upright swallows, the median IRP was 9 mmHg (normal for the Laborie system: <15 mmHg), again with no esophageal contractions. Figures 3, 4 show representative supine and upright swallows, respectively. Provocative testing showed pan-esophageal pressurization and an IRP of 14 mmHg during the rapid drink challenge (Figure 5). These provocative findings alone were insufficient to establish type I achalasia. Overall, the manometric findings suggested esophageal absent contractility.

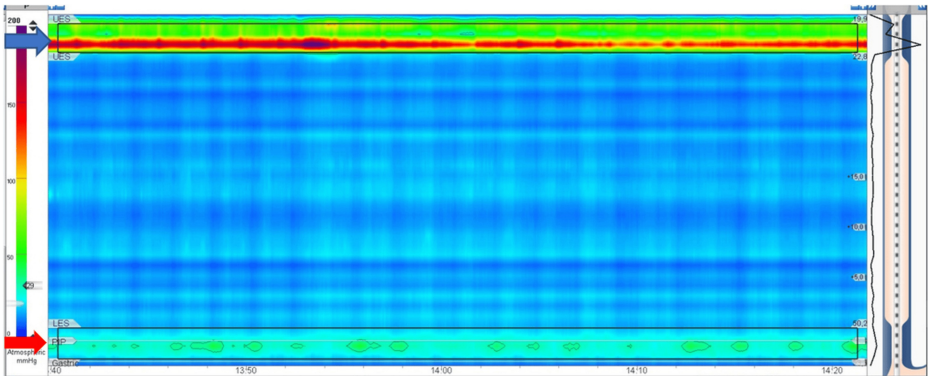


FIGURE 2: Basal high-resolution esophageal manometry tracing.

The red arrow indicates the resting pressure of the lower esophageal sphincter, and the blue arrow indicates the resting pressure of the upper esophageal sphincter.

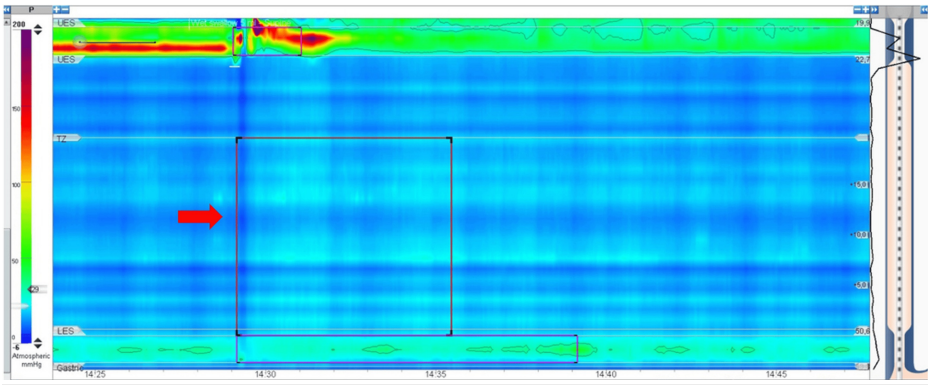


FIGURE 3: Representative supine swallow on high-resolution manometry, showing failed esophageal body contraction (red arrow).

Median supine IRP: 13.34 mmHg across 10 swallows.

IRP: integrated relaxation pressure.

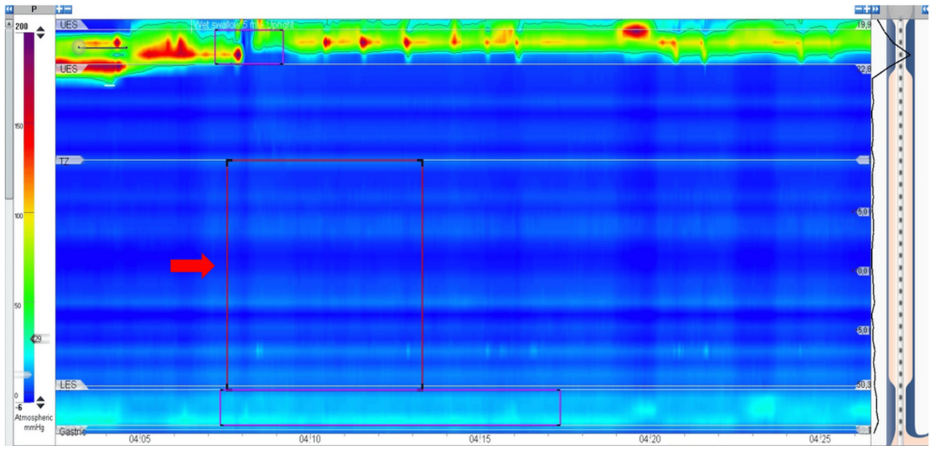


FIGURE 4: Representative upright swallow on high-resolution manometry, showing failed esophageal body contraction (red arrow).

Median upright IRP: 9 mmHg across five swallows.

IRP: integrated relaxation pressure.

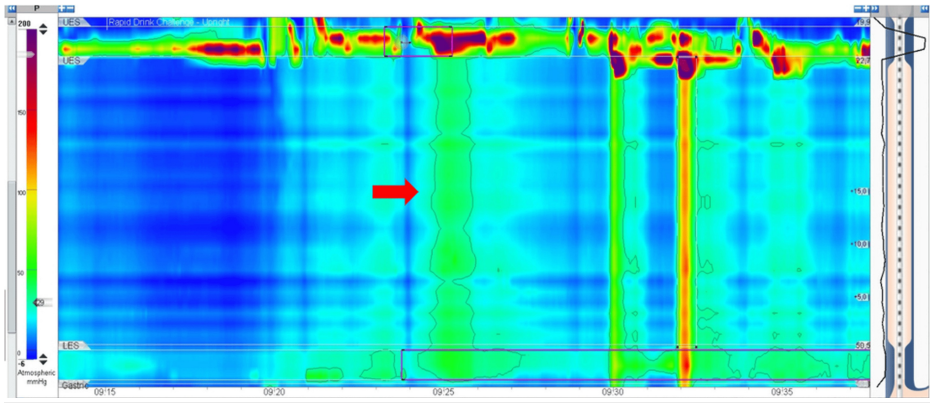


FIGURE 5: Rapid drink challenge on high-resolution manometry, showing pan-esophageal pressurization (red arrow).

IRP during the provocative maneuver: 14 mmHg.

IRP: integrated relaxation pressure.

The clinical presentation and high-resolution manometry findings supported a diagnosis of esophageal absent contractility. The timed barium esophagram showed a residual barium column measuring 3 cm at 1 min and 1 cm at 2 min, with complete clearance by 5 min, indicating preserved esophageal emptying without persistent radiographic evidence of esophagogastric junction obstruction. As part of the etiologic evaluation of esophageal absent contractility, an immunological workup was performed to investigate a possible underlying systemic autoimmune disease as follows: antinuclear antibodies by indirect immunofluorescence were <1:80, the erythrocyte sedimentation rate was 9 mm/h, and C-reactive protein was 0.1 mg/L. The patient had no diabetes or relevant medication exposure. Ambulatory 24-h esophageal pH monitoring off proton pump inhibitors showed an acid exposure time of 2%, arguing against pathological acid exposure.

The patient received dietary and lifestyle advice, including eating small, frequent meals, cutting food into small pieces, chewing thoroughly, taking sips of water with solid food, and remaining upright during and after meals. A five-day trial of metoclopramide 10 mg daily did not improve his dysphagia. Given the persistence of severe symptoms, peroral endoscopic myotomy (POEM) was performed. The procedure was carried out using an Olympus EVIS X1 endoscope (Tokyo, Japan: Olympus Corporation), a TT knife, and an ERBE VIO 3 electrosurgical unit (Tübingen, Germany: Erbe Elektromedizin GmbH).

Endoscopy showed no retained food or suspicious esophageal mucosal lesion. The cardia was located 45 cm

from the incisors and could be traversed with slight resistance. Saline tinted with methylene blue was injected at 37 cm from the incisors, followed by a mucosal incision at the same level. A submucosal tunnel was extended to 2.5 cm beyond the cardia. The myotomy began 2 cm distal to the mucosal incision and had a total length of 8.5 cm, including a 2.5-cm gastric extension beyond the cardia. It was selective for the first 2.5 cm and full thickness for the remaining 6 cm. The mucosal entry was closed with six 11-mm clips. The procedure lasted 70 min, with no immediate complications (Figures *6A*-*6H*).

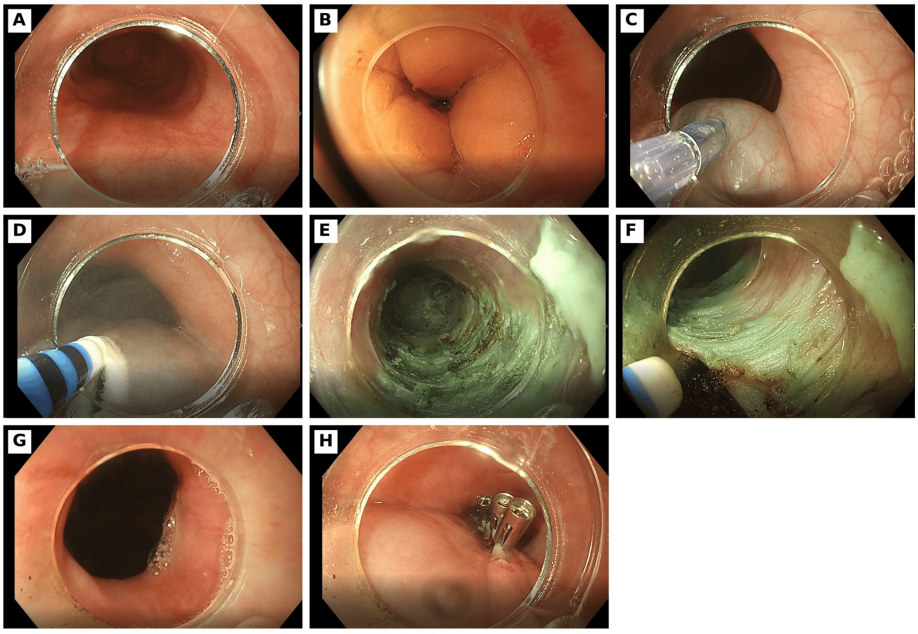


FIGURE 6: Endoscopic stages of peroral endoscopic myotomy (POEM).

(A) Esophagus before POEM. (B) Esophagogastric junction before POEM. (C) Submucosal injection. (D) Mucosal incision. (E) Submucosal tunnel. (F) Myotomy. (G) Esophagogastric junction after POEM. (H) Closure of the mucosal entry with clips.

At the latest clinical follow-up, approximately five months after POEM, he was eating solid food without dysphagia, regurgitation, chest pain, reported reflux symptoms, or belching. His Eckardt score was 0, and his weight had increased from 54 to 65 kg (gain of 11 kg). He was taking a proton pump inhibitor once daily. Control esophageal manometry at four months showed a decrease in basal lower esophageal sphincter pressure from 18 to 5 mmHg, indicating lower esophageal sphincter hypotonia after POEM. The median supine integrated relaxation pressure (IRP) decreased from 13.3 to 4.14 mmHg, and the median upright IRP decreased from 9 to 0.84 mmHg (Table *1*, Figures *7A*-*7C*). Esophageal contractions remained absent after POEM.

Parameters	Before POEM	Four months after POEM
LES resting pressure	18 mmHg	5 mmHg
Median supine IRP	13.3 mmHg	4.14 mmHg
Median upright IRP	9 mmHg	0.84 mmHg

TABLE 1: Esophageal manometry before and four months after POEM.

POEM: peroral endoscopic myotomy; LES: lower esophageal sphincter; IRP: integrated relaxation pressure

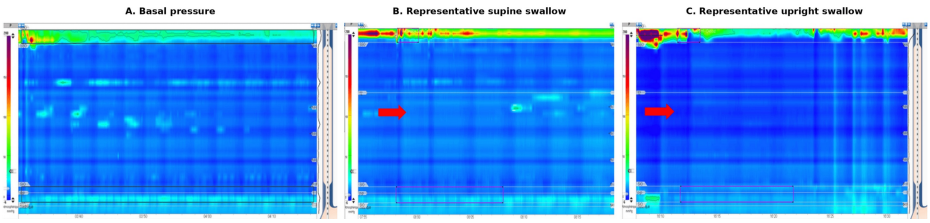


FIGURE 7: Follow-up high-resolution esophageal manometry four months after peroral endoscopic myotomy.

(A) Basal pressure tracing showing lower esophageal sphincter hypotonia (resting pressure: 5 mmHg). (B) Representative supine swallow and (C) representative upright swallow, both showing persistent absence of esophageal body contraction (red arrows).

Discussion

Absent contractility is characterized under the Chicago Classification version 4.0 by 100% failed peristalsis, with a normal median integrated relaxation pressure (IRP) in both supine and upright positions [1]. This pattern is physiologically distinct from classic type I achalasia, in which impaired esophagogastric junction (EGJ) relaxation accompanies absent peristalsis. Nevertheless, a normal IRP does not invariably rule out clinically relevant EGJ dysfunction. This distinction is especially important when dysphagia, marked esophageal dilation, retained food, or provocative pan-esophageal pressurization suggest an achalasia variant [1,4]. In our patient, all 15 standard swallows failed, but both median IRPs were below the Laborie-specific upper limits. The previously documented megaesophagus, food retention, and rapid drink challenge findings prompted a search for occult obstruction. The absence of a distal bird-beak appearance and complete clearance on the timed barium esophagram supports absent contractility, although it cannot completely exclude subtle EGJ dysfunction. Functional lumen imaging probe (FLIP) measurement of EGJ distensibility could have added diagnostic confidence but was unavailable [1,4].

The cause of absent contractility should be considered before treatment. Connective tissue disease, particularly systemic sclerosis, diabetes, severe reflux-related injury, and medication-related dysmotility are among the reported associations [3]. In this patient, there was no diabetes or relevant medication exposure; antinuclear antibodies were below 1:80, and inflammatory markers were low. These findings did not identify a secondary cause, but a negative antinuclear antibody test alone cannot exclude all systemic disease. Endoscopy showed neither erosive esophagitis nor candidiasis, and the off-therapy acid exposure time of 2% was below the threshold for pathological acid exposure on that pre-procedural recording [5]. The predominance of dysphagia rather than proven reflux is clinically relevant as follows: in an observational absent-contractility cohort, higher IRP values within the normal range were associated with a dysphagia-predominant phenotype, while lower values were associated with reflux-predominant symptoms [6]. This association is not a validated criterion for selecting patients for myotomy.

Management initially focuses on nutrition, symptom control, and any treatable cause or complication, because no established medication reliably restores peristalsis [3]. Small, frequent meals, careful chewing, water with food, and an upright posture after eating are practical measures for impaired bolus clearance. Acid suppression is appropriate when reflux is documented or clinically suspected, with endoscopic assessment for mucosal complications when indicated. Evidence supporting prokinetics for dysphagia caused by complete absence of contractions is limited [3]. The lack of benefit from five days of metoclopramide in our patient documents an unsuccessful brief symptomatic trial, rather than proving that all medical treatment had been exhausted. His long-standing severe dysphagia, initial weight of 54 kg, and persistent symptoms despite conservative care led to individualized consideration of an intervention.

The rationale for POEM in absent contractility is to reduce resistance at the EGJ and facilitate gravity-assisted emptying; myotomy does not directly target the underlying loss of esophageal body contractility. This rationale is less compelling when objective testing shows normal barium clearance, as in the present case; EGJ distensibility was not measured. Evidence remains limited. Zhao et al. treated six patients with absent contractility and reported a mean Eckardt score decrease from 6.3 to 4.0 and a mean IRP decrease from 10.8 to 3.7 mmHg; symptomatic improvement was smaller than in their achalasia and EGJ outflow obstruction groups [2]. With such a small uncontrolled sample, those results cannot establish routine efficacy or identify which patients benefit. Our patient's Eckardt score decreased from 8 to 0 at five months, and he gained 11 kg, providing a clinically meaningful short-term observation. Persistent absent contractions on follow-up manometry confirm that symptom relief occurred without recovery of peristalsis. The reduction in EGJ pressure offers a plausible explanation, but neither this single case nor the post-POEM manometry proves that occult EGJ obstruction was present beforehand. Longer clinical and objective follow-up is necessary.

Reflux after POEM deserves particular attention in a patient whose esophagus still cannot clear refluxate effectively. At four months, basal lower esophageal sphincter (LES) pressure had fallen from 18 to 5 mmHg, consistent with post-myotomy LES hypotonia; median supine and upright IRPs had fallen to 4.14 and 0.84 mmHg, respectively. Basal LES pressure reflects one element of the resting anti-reflux barrier, whereas IRP measures EGJ relaxation during swallowing. Therefore, a low resting LES pressure provides a physiologically plausible reason for concern, but neither the value of 5 mmHg nor the magnitude of IRP reduction establishes a diagnosis of gastroesophageal reflux disease (GERD) or quantifies an individual patient's reflux risk. In a long-term cohort of patients treated with POEM, predominantly for achalasia, lower post-procedure LES pressure independently predicted reflux esophagitis [7]. Hata et al. found no significant difference in reflux esophagitis or symptomatic GERD between patients with post-POEM IRP below 26 mmHg and those with IRP of 26 mmHg or higher, using Starlet-based criteria [8]. This comparison primarily distinguished normalized from persistently elevated IRP; it did not specifically evaluate profound LES hypotonia or establish the reflux risk associated with very low IRP values. These studies involved different populations and outcomes, and neither validates a post-POEM LES pressure or IRP cutoff for patients with truly absent contractility.

The anti-reflux barrier also depends on crural diaphragm function. In a retrospective study of 200 patients, Yamamoto et al. found that inspiratory augmentation of EGJ pressure, used as a marker of crural diaphragm contraction, decreased three months after POEM ($p=0.011$) [9]. Los Angeles grade B or higher reflux esophagitis was present in 102 patients (51%), who had weaker inspiratory augmentation than patients with no esophagitis or grade A disease ($p<0.001$); the association persisted in multivariable analysis [9]. These findings suggest that impaired diaphragmatic reinforcement may contribute to post-POEM reflux alongside changes in LES pressure. However, this observational association neither establishes causality nor defines a basal LES pressure threshold for reflux. Inspiratory augmentation was not available in the manometric analysis performed with the Solar GI system (Laborie) used at our center and therefore could not be assessed in this patient.

The absence of heartburn and regurgitation at five months is reassuring clinically, but the patient was taking a proton pump inhibitor (PPI) once daily, and symptoms alone may miss objective reflux. His pre-POEM acid exposure time of 2% describes the baseline state and cannot establish the post-operative acid burden. Follow-up should therefore include symptom and nutritional assessment, review of the need for acid suppression, and objective assessment for reflux complications, particularly endoscopy and, if the clinical question remains unresolved, ambulatory reflux monitoring using an on- or off-PPI protocol selected according to the clinical question [5]. This is especially relevant if dysphagia recurs, mucosal injury is found, or acid suppression is to be reduced. The favorable early response must be balanced against the unresolved long-term reflux risk and the limited evidence for POEM in absent contractility.

Conclusions

This case documents a marked short-term improvement in dysphagia and weight after peroral endoscopic myotomy (POEM) in a patient with manometrically defined absent contractility, despite persistent aperistalsis. Careful exclusion of an achalasia variant is essential when clinical, radiological, and manometric findings are discordant. Post-POEM lower esophageal sphincter hypotonia warrants reflux surveillance, but neither low resting sphincter pressure nor reduced integrated relaxation pressure alone establishes post-operative gastroesophageal reflux disease. The five-month follow-up is insufficient to assess the durability of symptom relief or detect late complications, including fibrosis-related recurrent dysphagia and delayed reflux-related injury. Longer follow-up and controlled studies with objective reflux assessment are needed to clarify the efficacy, safety, and role of POEM in this population.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Mounir El bied, Mehdi Zouaoui, Mbarek Azouaoui, Youssef Hnach, Nourdin Aqodad, Ahlam Boutaleb, Hassan Seddik, Hajar Kirami

Acquisition, analysis, or interpretation of data: Mounir El bied, Nourdin Aqodad, Mohamed Amine El Amghari

Drafting of the manuscript: Mounir El bied, Mehdi Zouaoui, Mbarek Azouaoui, Youssef Hnach, Nourdin Aqodad, Ahlam Boutaleb, Hajar Kirami

Critical review of the manuscript for important intellectual content: Mounir El bied, Nourdin Aqodad, Mohamed Amine El Amghari, Hassan Seddik

Supervision: Mehdi Zouaoui, Mbarek Azouaoui, Youssef Hnach, Nourdin Aqodad, Hassan Seddik

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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