

# Peroral Endoscopic Myotomy for the Treatment of Achalasia in a Patient with Esophageal Varices. A Case Report

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## ABSTRACT

Achalasia is very uncommon, and rarely does achalasia co-exist with esophageal varices. We present a 62-year-old woman who was diagnosed with both achalasia and esophageal varices in December 2014 and had a past history of hematemesis. The patient's achalasia symptoms' Eckardt score was 9, and her hepatic function was Child-Pugh grade A6. After comprehensive assessment of the patient's health and discussion of the pros and cons of various therapies for achalasia, the patient underwent a peroral endoscopic myotomy. She was symptom-free after the operation and had no recurrence of achalasia symptoms at 20-month follow-up. No adverse events were reported. Peroral endoscopic myotomy for achalasia with esophageal varices has not been previously reported in the English literature.

**Key words:** achalasia – esophageal varices – gastric varices – liver cirrhosis – peroral endoscopic myotomy.

**Abbreviations:** BTI: Botulinum toxin injections; EUS: Endoscopic ultrasonography; GEJ: Gastroesophageal junction; HIFU: High-intensity focused ultrasound; LHM: Laparoscopic Heller myotomy; PD: Pneumatic dilation; POEM: Peroral endoscopic myotomy; TIPS: Transjugular intrahepatic portosystemic shunt.

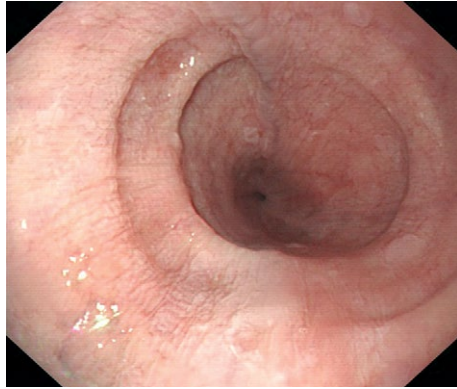
## INTRODUCTION

Achalasia affects approximately 1 in 100,000 individuals [1], while co-existing esophageal varices with achalasia are quite rare. Ten cases have been reported in the English medical literature, seven of which received treatment [2-5]. However, these treatments often involved various risks and complications, such as risk of hemorrhage after thoracotomy, dysphagia recurrence requiring subsequent myotomy after botulinum toxin injections (BTI), and hepatic encephalopathy after transjugular intrahepatic portosystemic shunt (TIPS). These treatment strategies may alleviate symptoms effectively, but may also cause the patient physical or mental stress. Peroral endoscopic myotomy (POEM) is a minimally invasive endoscopic

approach to the Heller myotomy for achalasia. Over the past 6 years, research has verified its safety and effectiveness to treat achalasia [1, 6]. Additionally, the endoscopic approach allows the physician to visualize varices, which may permit better avoidance of bleeding complications and easier implementation of precise electrocautery. We report a rare case of achalasia with esophageal varices that was treated with POEM. This is the first published report of such a case in China.

## CASE REPORT

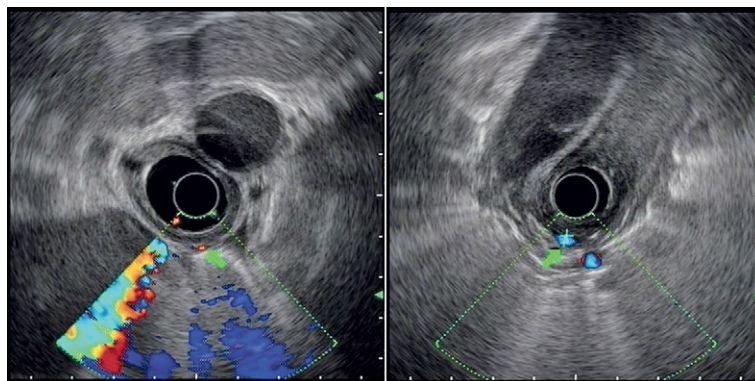
A 62-year-old woman presented with dysphagia accompanied by occasional retrosternal pain, vomiting, and a nocturnal cough for six years. Examination by endoscopy showed esophagitis. The patient was also diagnosed with hepatitis B cirrhosis. She was diagnosed with achalasia and esophageal varices due to hepatitis B cirrhosis in December 2014 and was admitted to our hospital in April 2015 for progression of dysphagia and a nocturnal cough lasting for one month. Between diagnosis and being admitted to the hospital, she had lost 12.5 kg and had two episodes of hematemesis, totaling 100 ml. After a series of examinations, the diagnosis of achalasia with esophageal varices from hepatic B cirrhosis was confirmed. Gastroscopy showed a dilated and atonic esophagus with a large liquid food residue and two pewter columns of grade 1 esophageal varices (Fig. 1). Esophageal ultrasound



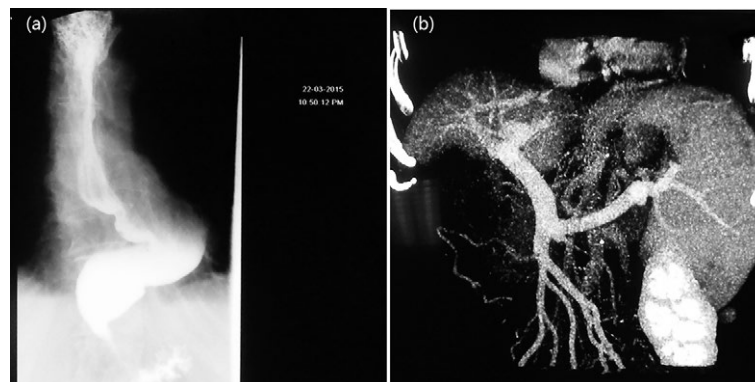
**Fig. 1.** Gastroscopy showed a dilated and atonic esophagus with two pewter columns of grade 1 esophageal varices.

(EUS) also exhibited a mild varix (Fig. 2). The gastric fundus had no lesions or varices. A barium esophagogram and manometry both confirmed the diagnosis of achalasia (Fig. 3a). Computed tomographic portal angiography found a series of problems: cirrhosis, splenomegaly, ascites, and portal hypertension without obvious gastric-esophageal or left gastric varices (Fig. 3b). There were no abnormal cardiac or pulmonary findings. The patient had an Eckardt score of 9 for her symptomatic achalasia. Her hepatic function was Child-Pugh grade A6. After considering the various treatment options, we decided to perform POEM.

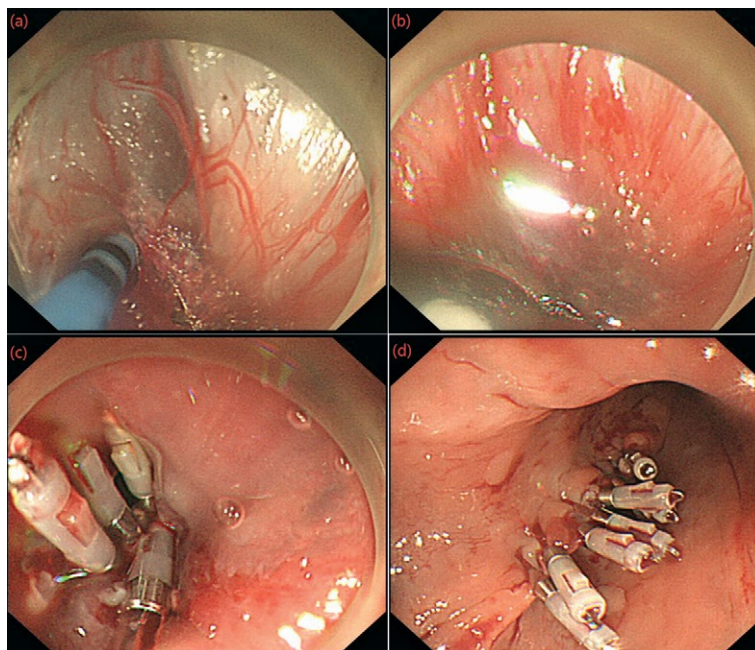
After obtaining informed consent, we conducted the operation. The patient was placed in the supine position and general anesthesia was administered. Carbon dioxide was used for insufflation. A relatively “low risk” window to conduct submucosal injection was chosen at the anterior wall of the median esophagus because of the esophageal varices at the right and posterior walls (Fig. 1). A mixture of methylene blue, saline, and epinephrine was injected into the anterior esophageal wall 9 cm from the gastroesophageal junction (GEJ). A 2 cm longitudinal mucosotomy was performed after raising a submucosal wheal. Then we created the submucosal tunnel that extended past the GEJ and 3 cm onto the gastric cardia, carefully avoiding the varices and giving adequate hemostasis. A rich vascular network appeared at 2 cm in front of the GEJ, leading to a mild hemorrhage that was alleviated with adequate hemostasis by electrotome thermocoagulation (Fig. 4a, b). Under direct endoscopic visualization, a selective myotomy of circular muscle layer was performed with a high-intensity focused ultrasound (HIFU) knife, starting 2 cm below the mucosal entry point and ending 2 cm distal to the GEJ, taking care to preserve the longitudinal muscle layers. We found a small lesion on the mucosa near the cardia and closed it with three endoscopic clips. Smooth passage of the endoscope through the GEJ confirmed an adequate myotomy and showed no perforations or active bleeding. The mucosotomy was then closed with endoscopic clips (Fig. 4c, d). The whole operation lasted 115 minutes. Additionally,



**Fig. 2.** EUS revealed mild esophageal varices.



**Fig. 3.** a: barium esophagogram demonstrated the delay of barium passing and a “bird-beak” appearance with a dilated esophagus. b: computed tomographic portal angiography presented a series of problems: cirrhosis, splenomegaly, ascites, portal hypertension without obvious gastric-esophageal or left gastric varices.



**Fig. 4.** a, b: a rich vascular network appeared and adequate electric coagulation hemostasis was given during the formation of the submucosal tunnel. c, d: the mucosal lesion and the mucosotomy were closed with endoscopic clips.

the patient received abdominocentesis at McBurney's point for relief from her pneumoperitoneum. On postoperative day 2, the patient had an abdominal CT scan to rule out the existence of an esophageal-mediastinum fistula or abdominal-esophageal fistula. Octreotide, proton-pump inhibitors, and diuretics were used to control the patient's portal hypertension at the perioperative period. The patient started a liquid diet on postoperative day 6 and was free of dysphagia. At 20-month follow-up, her Eckardt score was zero without any episodes of hematemesis.

## DISCUSSION

This case of a patient with co-existing achalasia and esophageal varices is extremely rare. After a review of the English literature, we found 10 cases, 7 of which received treatment [2, 3, 7]. Two patients underwent surgery due to recurring massive regurgitation of blood with a normal liver [7]. After several successive BTIs without success, a TIPS procedure was performed for portal decompression before proceeding with pneumatic dilation (PD) in the third patient [8]. Three patients were treated with EUS-guided BTI [2, 3, 5]. The last patient received a laparoscopic Heller myotomy (LHM) [4]. The combination of achalasia with esophageal varices from cirrhosis with portal hypertension prompted us to consider the treatment effects and the risk of hemorrhage from a combination of varices, thrombocytopenia, and coagulation abnormalities. The EUS-guided BTI has a low initial success rate and the high rate of achalasia recurrence cannot be ignored. Additionally, repeated treatments with BTI have been shown to make subsequent Heller myotomy more challenging [9]. Although PD after TIPS carries a low risk of hemorrhage, patients need to be monitored for hepatic encephalopathy after TIPS and surgical intervention is required if the patient suffers

from esophageal perforation requiring repair [8]. Gastric varices (GV) were not found in this case, so maybe PD was an alternative treatment. However, our recent research showed that the clinical success of simple PD after six months and over three years follow-up were 82.9% and 62.9% individually, which were significantly lower than 100% ( $P < 0.001$ ) and 85.7% ( $P = 0.029$ ) of POEM. Its long-term retreatment rate (28.6%) was higher than after POEM (2.86%) ( $P = 0.003$ ). This conclusion was consistent with the previous literature [1]. The study suggested that repeated and "graded" dilation brought out good outcomes, but with the diameter increasing, a high perforation rate went up. Considering that the patient was in the early stage of cirrhosis and varices, PD should not be the first line treatment in this case. Encouragingly, POEM has many benefits: high remission rate, low relapse rate, low incidence of complications, ability to directly visualize the esophagus during the procedure, and good control during the operation. Avoiding the esophageal varices and ensuring adequate hemostasis by electric coagulation can be done effectively during the myotomy under endoscopic visualization by an experienced operator.

For this patient, urgent intervention was necessary. With a symptomatic Eckardt score of 9, she was afflicted by serious dysphagia and a persistent nocturnal cough, putting her at risk of a series of subsequent troubles, such as a poorer physical and mental condition, the possibility of developing pulmonary infiltrates, and malnutrition due to achalasia and cirrhosis. Bleeding from the lower esophagus is a long-term complication of achalasia due to the esophagitis induced by persistent residual food and esophageal dilatation, a risk that has been verified both pathophysiologically and clinically [7, 10]. Varices, together with esophagitis, notably increase the risk of hematemesis. Our patient suffered an upper gastrointestinal tract bleed (100 ml) a month prior to POEM that was treated



with pharmaceutical management. In this case, esophageal bleeding is generally thought to be caused by the rupture of an esophageal varix secondary to cirrhosis without endoscopic verification. However, we could not confirm the cause of the patient's previous hemorrhage to have been from her esophageal varices. Considering the low grade (grade 1) esophageal varices present in our patient, the only mildly abnormal blood coagulation function, and the absence of thrombus at a site of previously bleeding varices under endoscopy, in conjunction with the patient's previous diagnosis of esophagitis 6 years prior, we could also not rule out that the hematemesis was the result of esophagitis from long-standing achalasia. The patient's most troubling problem was her symptomatic achalasia; therefore, we chose POEM as the most appropriate treatment for achalasia and pharmacotherapy for her esophageal varices and cirrhosis. While endoscopic band ligation or sclerotherapy can be used to treat esophageal varices, these treatments take time, and patients may develop esophageal scarring, which may further worsen the symptoms of achalasia [5]. The patient underwent successful POEM with octreotide, proton-pump inhibitor, and diuretics for medical management of her varices.

## CONCLUSION

Co-existence of achalasia with esophageal varices is rarely reported, and management of this complicated condition with POEM has not been previously published. Based on our experience, POEM seems to be safer and more effective than other traditional therapies for this type of complex patient. Combined with appropriate pharmaceutical management, POEM may be an alternative option to manage achalasia associated with mild varices from cirrhosis. However, the long-term effects should be monitored, and further research is required regarding the specific indications and therapeutic effects of POEM.

**Conflicts of interest.** The authors declare no conflicts of interest.

**Patient's consent:** obtained.

**Authors' contributions:** H. Z. performed the operation and approved the final version. N.S. and L.Y. drafted and revised the manuscript.

X.W., X.Z., L.Y., H.X. and H. Z. conceived the work, acquired data, and played an important role in interpreting and analyzing the results.

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