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CASE REPORT | ENDOSCOPY

Safe Use of POEM in a Patient With Decompensated Cirrhosis and Severe Achalasia

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ABSTRACT

Patients with both achalasia and decompensated cirrhosis can often present a therapeutic challenge because portal hypertension has generally been considered a contraindication to definitive therapies for achalasia. This case report depicts a patient who presented with progressive dysphagia, weight loss, and large-volume ascites; was diagnosed with type II achalasia and decompensated cirrhosis without esophageal varices; and underwent peroral endoscopic myotomy after preprocedural transjugular intrahepatic portosystemic shunt placement. Our case highlights the importance of multidisciplinary care and need for definitive therapies for these complex patients at high risk of malnutrition and sarcopenia.

KEYWORDS: cirrhosis; achalasia; peroral endoscopic myotomy; third space endoscopy; sarcopenia

INTRODUCTION

Peroral endoscopic myotomy (POEM) has become a widely accepted therapy for the management of advanced achalasia.[1,2] Compensated cirrhosis with portal hypertension has generally been considered a contraindication to POEM.[3] Despite this, previous case report series have reported successful POEM in patients with cirrhosis (both with and without esophageal varices).[4–6] We present a case of a patient with decompensated Child C cirrhosis and achalasia who underwent successful POEM without immediate complications. To our knowledge, this is the first case report to describe large-volume ascites that was managed peri-POEM with transjugular intrahepatic portosystemic shunt (TIPS) and temporary ascites drain placement.

CASE REPORT

A 59-year-old man with a history of achalasia and suspected nonalcoholic steatohepatitis decompensated cirrhosis was transferred to a tertiary care center with 3 weeks of intractable regurgitation, vomiting, and associated weight loss.

The patient reported a diagnosis of achalasia many years ago but with overall mild symptoms and, at baseline, to be able to tolerate both solids and liquids. At the time of his current presentation, the patient described approximately 4 months of abdominal distension and dysphagia that had worsened over the past month accompanied by a 10 lb weight loss (Eckardt score 9: dysphagia and regurgitation with each meal, daily chest pain, and less than 5 kg weight loss). Initial cross-sectional imaging was notable for ascites with evidence of portal hypertension including perisplenic and epigastric varices, as well as a fluid-distended distal esophagus (Figure 1). He required multiple large-volume paracenteses, which were negative for spontaneous bacterial peritonitis. During his hospitalization, he suffered from ongoing vomiting with food intolerance, with subsequent esophagram demonstrating a markedly distended esophagus with marked obstruction at the gastroesophageal junction suggestive of achalasia. Based on his persistent symptoms and radiographic findings, he underwent upper endoscopy that showed no evidence of esophageal varices and received botulinum toxin injection to the lower esophageal sphincter. Unfortunately, he did not experience symptomatic improvement and was not a candidate for percutaneous endoscopic gastrostomy placement because of large-volume ascites. Therefore, he was started on total parenteral nutrition before being transferred to a tertiary care center on day 15 after initial presentation for consideration of advanced therapies for his achalasia.

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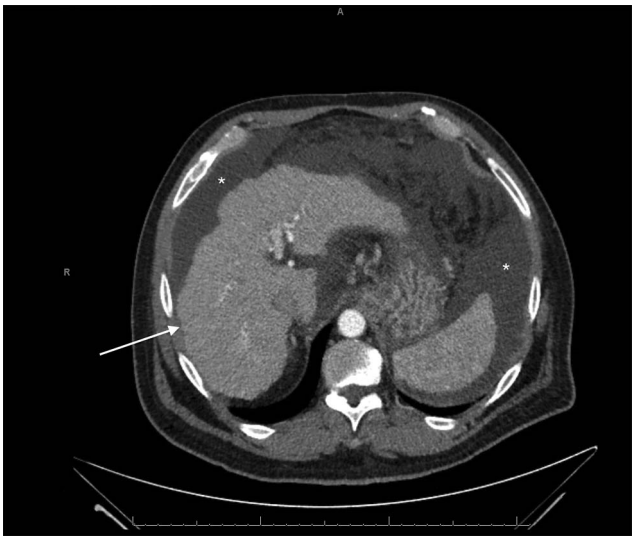

Figure 1. Admission computed tomography of the abdomen and pelvis demonstrating a cirrhotic liver (white arrow) with evidence of portal hypertension and large-volume ascites (white asterisks).

Vitals at the time of transfer were stable with a temperature of 97.6F, heart rate of 95 beats per minute, blood pressure of 146/80 mm Hg, and pulse oximeter of 96% on room air. Relevant laboratory results were notable for albumin 2.2, total bilirubin 2.0, international normalized ratio 2.14, and platelets 71,000 b/L and were otherwise normal (MELD-Na 19, MELD 18). He underwent confirmatory testing with chest computed tomography and repeat upper endoscopy, which showed a dilated and fluid-filled esophagus with narrowing at 40 cm from the incisors that could only be traversed with a pediatric endoscope. No esophageal or gastric varices were noted on this repeat upper endoscopy, although mild portal hypertensive gastropathy was identified. He also underwent endoscopic ultrasound, which showed a normal-appearing mucosa in the stomach and lower esophagus without mass lesions or disruption of the muscularis propria, but did note marked but discrete homogeneous and circumferential thickening of the submucosa and muscularis propria layers at 42–30 cm from the bite block (Figure 2). High-resolution esophageal manometry was also performed and revealed an elevated resting pressure with abnormal integrated relaxation pressure (median integrated relaxation pressure = 47.3 mm Hg), failed peristalsis, and panesophageal pressurization (Figure 3). Studies performed were all consistent with a diagnosis of type II achalasia. After multidisciplinary discussion, we proceeded with TIPS placement before POEM (Figure 4). He underwent successful TIPS placement on day 13 of this admission with reduction in the portosystemic gradient from 21 mm Hg to 4 mm Hg and drainage of 7 L of clear yellow ascites. Given persistent ascites the following week after TIPS placement, despite aggressive intravenous diuresis, a temporary ascites drain was placed before proceeding with POEM. Four days later, he underwent successful POEM with a 12 cm myotomy. A fluoroscopic esophagram the day following POEM did not show any leak, and he had near resolution of symptoms. He was tolerating a soft diet before discharge, 5 days after POEM. Approximately 1 year after POEM, the patient was doing well without symptoms and endorsed a 30 lb weight gain (Eckardt score 0) (Figure 5).

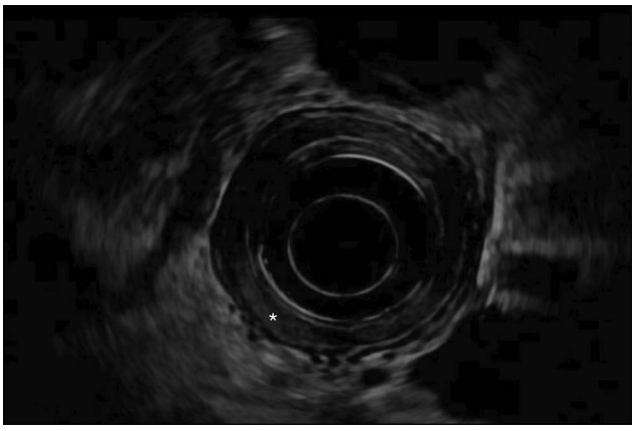

Figure 2. Endoscopic ultrasound revealed circumferential thickening of the submucosa and muscularis propria layers (white asterisk), also consistent with type II achalasia.

DISCUSSION

Cirrhosis has been previously considered a potential contraindication to certain therapies, such as pneumatic dilation and POEM, for achalasia.[3,7] To our knowledge, this is the first case report to describe large-volume ascites that was managed peri-POEM with TIPS and temporary ascites drain placement. Periprocedural TIPS placement has been reported in other nonhepatic surgeries.[8] In our case, TIPS placement was performed to resolve portal hypertension and, therefore, to mitigate the theoretical risk of variceal bleeding and reduce ascites.

Recently, the literature has elucidated an association between cirrhosis and esophageal motility disorders, believed to be due to autonomic dysregulation in the setting of increased nitrous oxide production.[9] This case demonstrates a potential management approach to successfully treat achalasia in this subset of patients. Patients with achalasia typically present with weight loss and are often malnourished.[10,11] Sarcopenia and frailty are important yet potentially modifiable complications that predict morbidity and mortality in patients with cirrhosis. While these complications are multifactorial, adequate caloric and protein intake are important factors that contribute to frailty and sarcopenia in this patient population.[12] Both frailty and sarcopenia are also more prevalent in patients with decompensated cirrhosis compared with compensated cirrhosis.[12] Presumably, patients with both achalasia and decompensated cirrhosis are at even more significant risk of malnutrition and sarcopenia given the individual risk within each of these populations. Therefore, expanding therapeutic options to allow for enteral

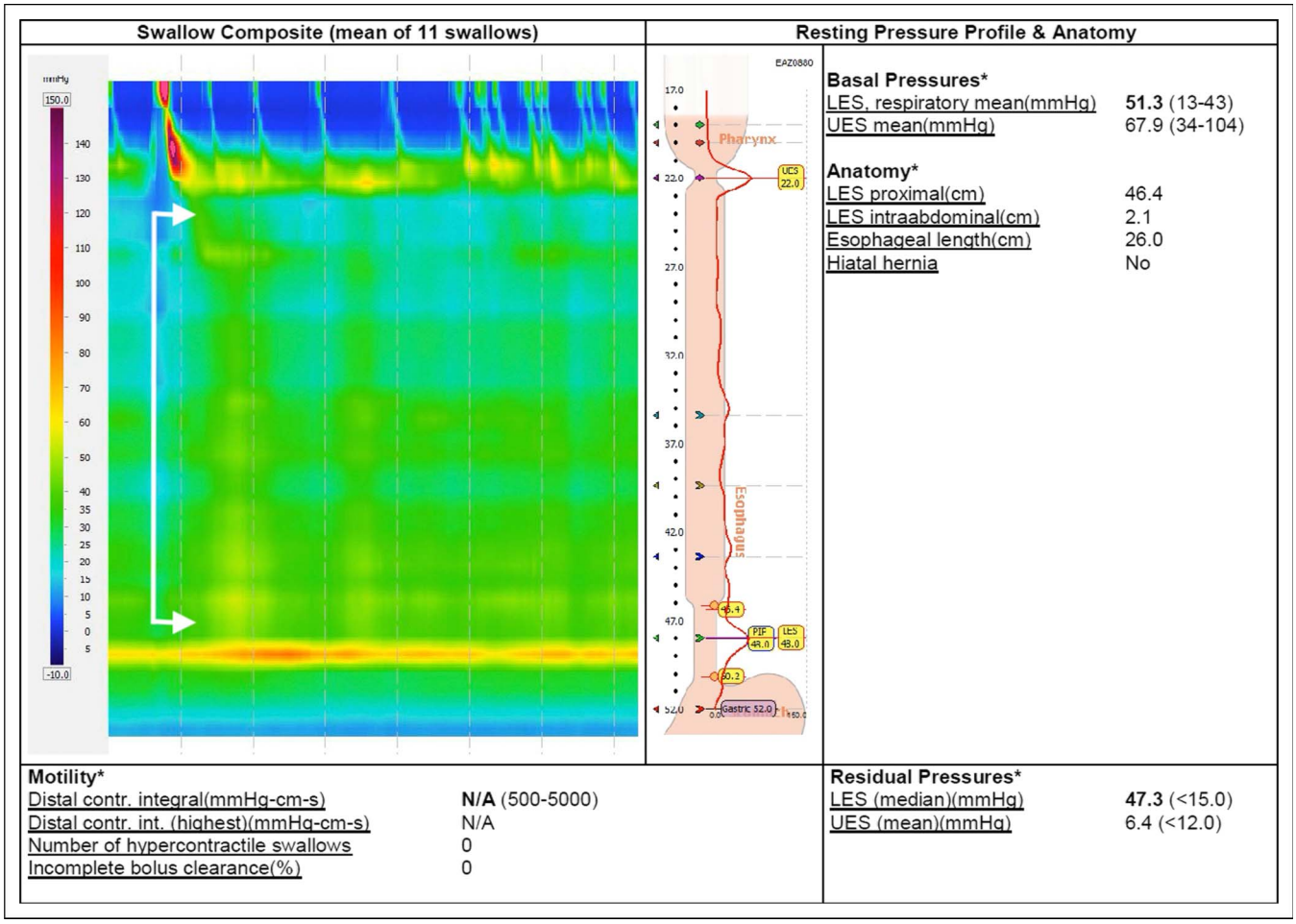

Figure 3. Results of high-resolution esophageal manometry demonstrating elevated integrated relaxation pressure, failed peristalsis, and panesophageal pressurization (white bracketed arrows) consistent with type II achalasia.

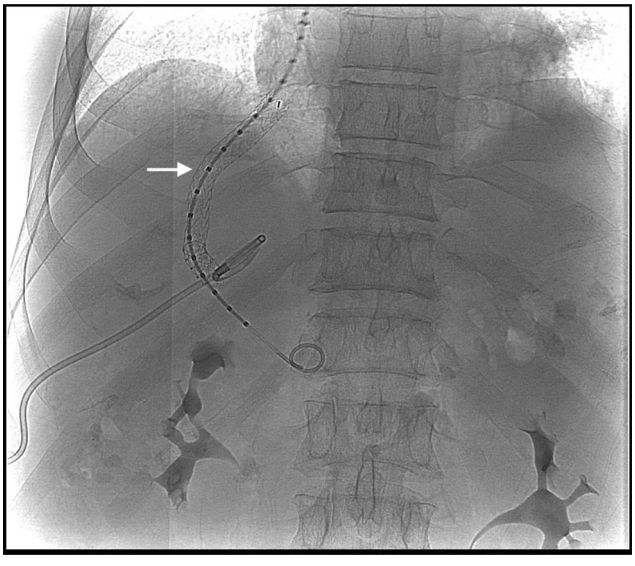

Figure 4. Fluoroscopic images demonstrating successful placement of the transjugular intrahepatic portosystemic shunt (white arrow).

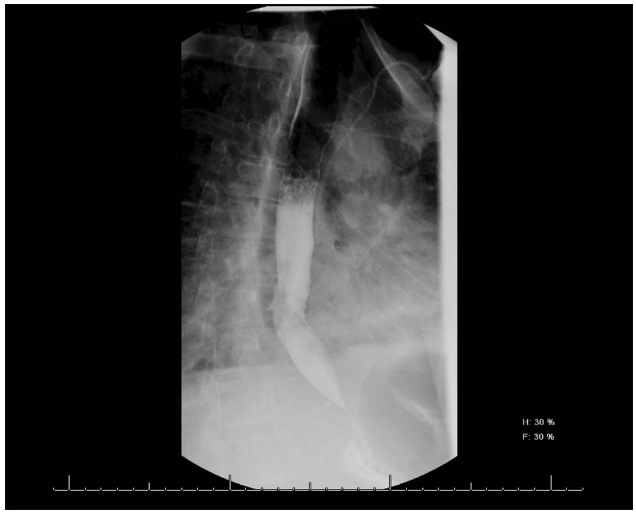

Figure 5. Esophagram, 1 day after peroral endoscopic myotomy.

feeding in those patients with concomitant decompensated cirrhosis and gastrointestinal disease such as achalasia should be a focus for future investigations because adequate nutrition may both improve quality of life and prevent adverse outcomes in these patient populations.

DISCLOSURES

Author contributions: All authors made substantial contributions to the conception of this report as well as drafting/revising of the content of the report and provided final approval for publication. R. Loh is the article guarantor.

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