

Achalasia cardia with esophageal varix managed with endoscopic ultrasound-guided botulinum toxin injection

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Abstract Achalasia cardia is a motility disorder of the esophagus characterized by failure of relaxation of the lower esophageal sphincter. Nitrates and calcium channel blockers, pneumatic dilatation, botulinum toxin injection and surgical myotomy have been described in literature as possible management options. We present a patient who presented with achalasia and was co-incidentally diagnosed to have cryptogenic cirrhosis with portal hypertension and had esophageal varices. This clinical combination precluded the use of pneumatic dilatation and surgical myotomy. We injected botulinum toxin into the lower esophageal sphincter using a celiac plexus neurolysis needle under endoscopic ultrasound guidance; the clinical response was good.

Keywords Achalasia · Endoscopic ultrasound · Esophageal varices

Introduction

Achalasia cardia is a primary esophageal motility disorder characterized by failure of relaxation of the lower esophageal sphincter (LES), and aperistalsis of the distal esophagus. Oral medications (nitrates, calcium channel blockers), endoscopic pneumatic dilation, botulinum toxin injection into the LES and surgical myotomy have all been used in its management with variable success rates. However, co-existence of

achalasia with gastroesophageal varices presents a unique clinical situation with limited management options.

We present such a patient who underwent endoscopic ultrasound (EUS)-guided botulinum toxin injection using a celiac plexus neurolysis (CPN) needle into the LES with symptomatic relief.

Case report

A 58-year-old lady presented with dysphagia and heartburn for 2 years. She had hypertension and type 2 diabetes mellitus for 10 years, managed with calcium channel blockers and sulphonylureas.

Gastroscopy showed a dilated esophagus, tight gastroesophageal junction (GEJ) and a single column of grade 2 esophageal varix (Fig. 1). The gastric fundus was free of any mucosal lesion or varices. Esophageal manometry (water perfusion, Kangaroo JEF, RMH Limited, Australia) confirmed achalasia (high resting LES pressure, failure of LES relaxation on wet swallows, and aperistalsis of lower esophagus). Evaluation of varices led to a diagnosis of Child B cryptogenic cirrhosis with portal hypertension.

Pneumatic dilatation of the LES and surgical myotomy could not be done in view of high risk of complications, especially bleeding from varices. A trial of nitrates failed to yield any symptomatic improvement. In view of limited options, treatment with EUS-guided botulinum toxin was considered. Figure 2 shows endoscopic ultrasound views of large peri-esophageal collaterals which we planned to avoid during the injection.

After obtaining an informed consent, botulinum A toxin (Botox, Allergan Inc., Irvine, California, USA) was injected

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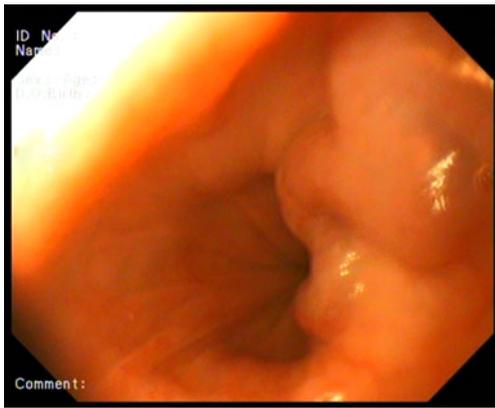


Fig. 1 Gastroscopy showing single column of tortuous Grade 2 esophageal varix in dilated esophagus

under EUS guidance (Olympus GF-UCT180, EU-ME1) at 4 sites into the LES (20 U per site) using a CPN needle (Echo-20-CPN, Wilson Cook, USA) (Figs. 3 and 4). There were no procedure-related complications. The patient had symptomatic relief the following day; at 3 months follow up, she did not have recurrence of symptoms.

Discussion

Achalasia cardia is associated with degeneration of ganglion cells in the myenteric plexus of esophagus which results in unopposed excitation of LES by the cholinergic system. Treatment is aimed at decreasing the LES pressure. Medical treatment with nitrates and calcium channel blockers has variable efficacy of 50% to 90%, with adverse effects being

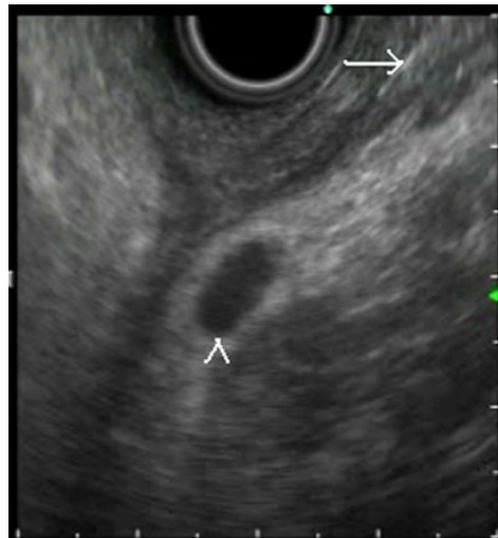


Fig. 3 Celiac plexus neurolysis needle in situ before botulinum toxin injection; a large collateral vessel is seen near it. *Arrow*: needle; *arrow head*: large collateral vessel

reported in up to 30% patients [1]. Pneumatic dilatation of LES provides significant symptomatic relief initially in more than 85% patients. Surgical myotomy carries high initial cost, protracted recovery but provides higher success rates. Botulinum toxin injection decreases LES pressure by inhibiting the release of acetylcholine. Initial success rate is close to 80% which usually weans off with time (70% at 3 months and 41% at 1 year) [2].

The co-existence of varices in a patient with achalasia is extremely infrequent and has been sparingly reported in literature [3–5]. Pinillos et al. reported a similar patient

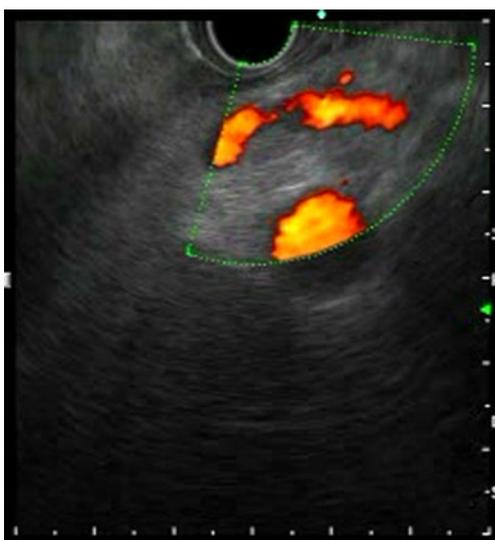


Fig. 2 Endoscopic ultrasound Doppler image showing multiple peri-esophageal collaterals



Fig. 4 Endoscopic ultrasound image after botulinum toxin injection; the injectate has resulted in compression of collateral vessel

who initially responded well to endoscopic botulinum toxin injection but later underwent transjugular intrahepatic portosystemic shunt (TIPSS) followed by pneumatic dilatation [3]. This remains an option for our patient should she fail to respond to future injections of botulinum toxin.

Conventionally, endoscopic botulinum toxin injection is carried out using a sclerotherapy needle. This may inadvertently go deeper than the muscularis propria since it is a “blind” technique and may result in a sub-optimal response. Accurate injection is likely to provide better relief of dysphagia [6–8]. The CPN needle has the additional advantage of “spray effect” which enables circumferential diffusion of botulinum toxin in the muscularis propria. The benefit is analogous to EUS-guided celiac plexus neurolysis, when alcohol is injected directly into the visualized celiac ganglia. EUS guidance also helps avoid sub-mucosal and peri-esophageal collaterals even though inadvertent intra-variceal injection is unlikely to cause any systemic side effects [9].

In conclusion, therapeutic options are limited when achalasia and varices co-exist. EUS-guided botulinum toxin injection using a CPN needle appears to be a safe and effective option for such patients.

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