Extreme Esophageal Spasm Treated with Botulinum Toxin

Brijesh Patel1*, Kimberly Kolkhorst2, Joel Richter2 and Jay Mamel2

1Department of Internal Medicine, University of South Florida, USA
2Department of Gastroenterology, University of South Florida, USA

Abstract

Diffuse esophageal spasm is a rare motility disorder. The primary symptoms include dysphagia, and chest pain. There is array of treatments available from medical therapy with nitrates to surgical therapy; Heller myotomy. We present a case of severe Diffuse Esophageal Spasm (DES) that showed one of the highest Distal Contractile Integral (DCI) per manometry. Patient was treated with Botulinum Toxin (BTX) with complete resolution of symptoms. The use of botulinum toxin for treatment of motility disorders; achalasia and diffuse esophageal spasm is well reported. We present a case of a severe DES that responded well to endoscopic botulinum toxin injections. This is a novel approach to DES who has failed conservative medical therapy.

INTRODUCTION

Primary esophageal motility disorders are a spectrum of diseases, including achalasia, Diffuse Esophageal Spasm (DES), and nutcracker esophagus. DES is rare, making up ~ 3% of all motility disorders [1]. The work-up for DES involves barium studies, Esophago Gastro Duodeneoscopy (EGD) and esophageal manometry. Manometry is regarded as the gold standard for assessing esophageal motor function. Simultaneous waves in >20% of wet swallows with alternating normal peristalsis and high amplitude contractions are required for diagnosis of DES. The high resolution manometry allows for measurement of intraluminal impedance monitoring, transient Lower Esophageal Sphincter (LES) relaxations and diffuses Distal Contractile Integral (DCI). The DCI integrates the length, contractile vigor, and duration of contraction of the first two sub-segments of the distal esophageal segment contraction, expressed as mmHg*sec*cm with > 5,000 mm Hg*sec*cm is considered as high [2]. Multiple treatments have been used to treat severe esophageal spasms including; Calcium-Channel Blockers (CCB’s), nitrates and surgical approaches, including Heller myotomy. More recently, botulinum toxin (BTX) has been introduced as a treatment option for patients with DES [1,3-5].

CASE PRESENTATION

A 51 year old male with no significant past medical history was referred to our center for intermittent dysphagia to solids associated with severe chest pain for approximately 6 months. Previous cardiac work-up was negative. His chest pain was described as a “strong, squeezing sensation” that woke him from sleep. He denied dysphagia, odynophagia and acid reflux. Symptoms were associated with 15-lb weight loss. Previous treatment with calcium-channel blockers, nitrates and proton pump inhibitor did not provide symptom relief.

He subsequently underwent a high resolution esophageal motility study which revealed high amplitude, prolonged simultaneous contractions with mean diffuse Distal Contractile Integral (DCI) of 25,901 mmHg*sec*cm (normal < 5,000 mmHg*sec*cm). The DCI was highest with swallow # 4 and #9, peaking at 90,000 mmHg*sec*cm in a series of 10 swallows (Figure 1). Barium swallow was negative for primary achalasia. EGD showed a non-dilated, narrowed esophagus with no evidence of rings or furrows (Figure 2). The lower esophageal sphincter was widely patent (Figure 3). Active spasms were noted during endoscopy along a 2-3cm long segment just above the Z line with resistance to passing the endoscope into the stomach. We injected 200 units of BTX in 10, 1cc aliquots: first 3 injections just above the Z line at ~ 42cm, and another 7 injections ~ 10 cm above distal esophagus. Esophageal biopsies revealed changes of chronic GERD, but negative for dysplasia or eosinophils. Two month follow-up after BTX injections revealed 80% symptomatic improvement and DCI of only 659 mmHg*sec*cm on repeat manometry. By six months all his symptoms have resolved.

DISCUSSION

This case highlights an extreme case of DES and symptomatic relief after treatment with botulinum toxin injections. Studies

ABBREVIATIONS

EGD: Esophago Gastro Duodeneoscopy; BTX: Botulinum Toxin; DES: Diffuse Esophageal Spasm; CCB: Calcium Channel Blocker; DCI: Distal Contractile Integral; LES: Lower Esophageal Sphincter; SSRI: Selective Serotonin Reuptake Inhibitor

Keywords

• Dysphagia
• Botulinum toxin injections
• Dysmotility
• Esophagus
• Endoscopy
• Manometry
by Storr et al. have shown the use of BTX in DES with reduction of total symptom score (chest pain and dysphasia) of 8 prior to treatment to 2 post treatment with maintenance score of 2 one month later. Vanuytsel et al. showed symptomatic improvement with botulinum toxin vs. placebo in a 22 patient sham controlled study on patient with DES or nutcracker esophagus. Our patient experienced 80% symptomatic relief after treatment with BTX and continued symptomatic relief at 2-month follow-up and complete resolution of all symptoms by 6 months. In addition; repeat manometry showed a significant reduction in the DCI from previous study.

Historically calcium-channel blockers, antimuscarinic agents, and SSRI’s have been used to treat DES, but our case supports a novel approach using botulinum toxin injections for effective, symptomatic relief of diffuse esophageal spasm, even in rather severe case. Use of BTX can be a safer alternative for patients who are unfit for surgeries and or have failed medical therapies. This case highlights the severe disease with very high DCI and complete resolution with BTX treatment. Future prospective studies involving large patient populations to assess BTX doses, need for repeat treatment and timing of doses are warranted.

REFERENCES