

Case Report

Achalasia during Pregnancy Treated with Botulinum Toxin A Injection at the Lower Esophageal Sphincter

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Abstract

A 30-year-old African American woman, gravida 8 para 6016, was admitted at 34 weeks of gestation because of shortness of air, dysphagia for solids and liquids, and loss of weight. A chest radiograph demonstrated a mediastinal mass that was compressing the trachea. Findings of chest and neck tomography were suggestive of esophageal achalasia with massively enlarged esophagus with tracheal compression. Endoscopic treatment with evacuation of enlarged esophagus and submucosal injection of botulinum toxin A in the lower esophageal sphincter was successful in relieving the obstruction and allowed for outpatient management with soft mechanical diet for remainder of pregnancy. A healthy male baby was born by spontaneous vaginal delivery at 38 weeks. The patient underwent laparoscopic esophageal myotomy and fundoplication at seven months postpartum.

INTRODUCTION

Achalasia is a rare motor disorder of the esophageal smooth muscle that occurs in 1 in 100,000 individuals. The disorder occurs at the level of the lower esophageal sphincter which does not relax normally with swallowing [1]. This leads to progressive dysphagia, chest pain and regurgitation of undigested foods and liquids. The underlying abnormality is loss of intramural neurons and is usually idiopathic. In rare cases, achalasia can be associated with gastric carcinoma infiltration of the esophagus, Chagas' disease, lymphoma, and neurodegenerative disorders [1]. Achalasia occurs in both males and females and at all ages and therefore, can be an uncommon cause of emesis and poor weight gain during pregnancy. While nausea and vomiting are common conditions of pregnancy, achalasia and other rare gastrointestinal disorders should be considered when vomiting and symptoms during pregnancy are persistent, not associated with the first trimester of pregnancy, and associated with concerning symptoms such as dysphagia, regurgitation of undigested food, and weight loss in particular [2]. Our case describes a severe case of achalasia during the third trimester of pregnancy that was able to be treated non-surgically with endoscopic injection of botulinum toxin A into the submucosal region of the lower esophagus without maternal or infant complications and provides additional evidence for a treatment option in the pregnant patient with achalasia that can be managed in the outpatient setting.

CASE PRESENTATION

A 30-year-old African American gravida 8, para 6016 was admitted to the hospital at 34 weeks due to worsening shortness of air not responsive to albuterol and inhaled steroid as an outpatient. The patient did not have a formal diagnosis of asthma prior to this pregnancy, but had been seen in the emergency room on multiple occasions for shortness of air with symptoms improved after nebulized albuterol treatments in this pregnancy. Patient had a past medical history of depression, gastroesophageal reflux disease, late prenatal care starting at 23 weeks, and Wolff-Parkinson-White syndrome for which she had seen cardiology as an outpatient and was taking pindolol 10 mg twice daily until she could receive an ablation procedure for ultimate resolution, which cardiology recommended to be performed in the postpartum period. The patient's previous pregnancies were 6 uncomplicated term vaginal deliveries. She reported one spontaneous miscarriage previously at 6 weeks with no complications.

During patient's hospitalization for worsening shortness of air, she also reported multiple episodes of vomiting partially digested foods and feeling very hungry but unable to eat due to persistent emesis. It was noted that the patient had lost 10 kilograms since her first prenatal visit at 23 weeks. The patient had a chest radiograph that demonstrated a mediastinal mass compressing the trachea. The patient was moved to the intensive

care unit due to concerns for acute airway obstruction from the mediastinal mass. Chest computed tomography revealed a markedly dilated esophagus with smooth tapered narrowing to the gastroesophageal junction, which is most suggestive of severe achalasia (Figures 1 and 2). There was also marked narrowing of the trachea due to compression between the right innominate artery and dilated esophagus. Pulmonary critical care and gastroenterology were consulted. Gastroenterology recommended acute decompression of the mega-esophagus and submucosal injection of the lower esophageal sphincter with botulinum toxin A. Due to her large gravid uterus at 34 weeks, the gastroenterologist was concerned about increased risks of severe reflux esophagitis and hiatal hernia from myotomy of the lower esophageal sphincter. The patient was counseled regarding the limited use of the botulinum toxin A in pregnancy. The patient

agreed to the esophago-gastro-duodenoscopy with botulinum injection for treatment of her severe achalasia. Careful attention was taken to avoid intra-arterial injection of any toxin during the procedure. A total of 100 IU of botulinum toxin A was injected into the submucosal region subdivided into 4 quadrants.

The patient was intubated for the procedure and remained intubated overnight for monitoring. Fetal heart monitoring was simultaneously performed at the bedside in the intensive care unit with supplies necessary for emergent cesarean available if indicated during the procedure. The patient and fetus tolerated the procedure well. She was subsequently extubated the following morning and moved back to the labor and delivery floor. However, she had a repeat bout of emesis that converted her into supraventricular tachycardia at a rate of 200-210s for

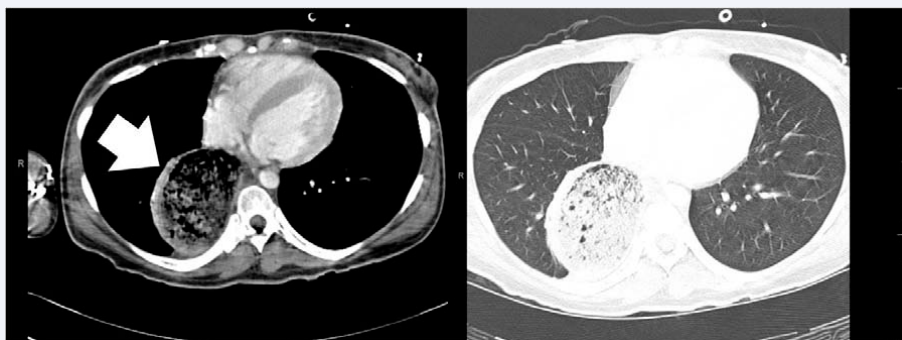


Figure 1 Axial chest tomography demonstrating enlarged esophagus (arrow) filled with debris in the mediastinum compressing the trachea and the ipsilateral lung.

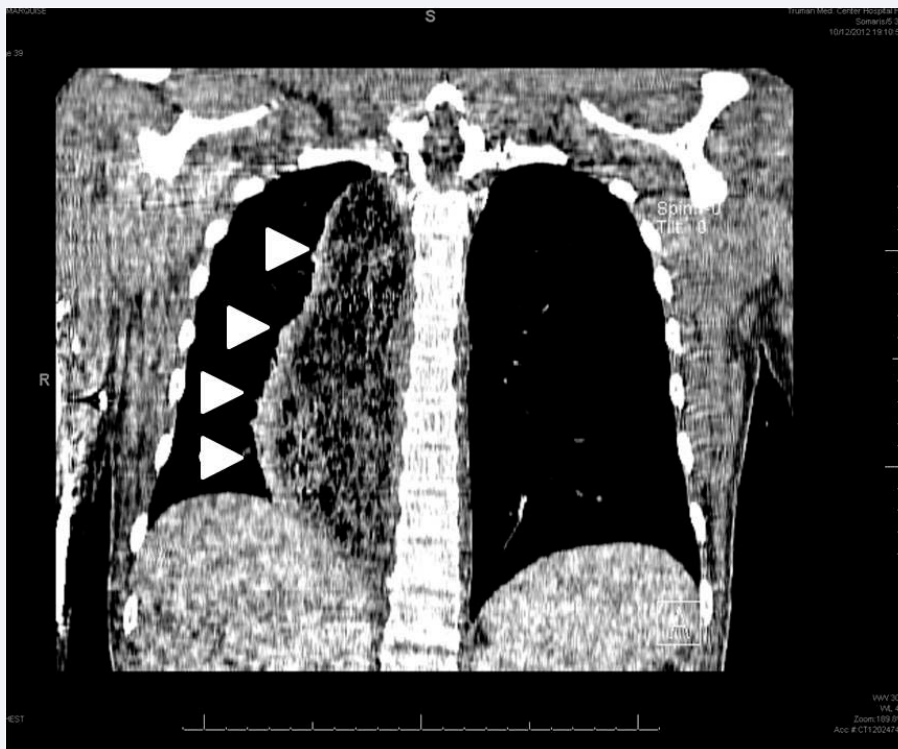


Figure 2 Coronal reconstruction chest tomography demonstrating full length of enlarged esophagus in the right chest cavity (arrowheads).

which she was transferred back to the intensive care unit, where she again had another bout of emesis which terminated the supraventricular tachycardia. This was felt to be due to her Wolff-Parkinson-White syndrome and she was followed an additional day in the intensive care unit without any further episodes of tachycardia and no need for additional treatment. She was restarted on her pindolol and discharged from the hospital 4 days after her esophagoduodenoscopy with botulinum injection.

At the time of discharge, she was tolerating a mechanical soft diet. She was able to maintain outpatient hospital visits while eating a soft mechanical diet and no further episodes of shortness of air. She did have an occasional episode of emesis, but never more than once a day and usually she attributed the episode to overeating. She gained 4 kilograms in 2 weeks. The patient missed her last prenatal visit and subsequently delivered at 38 weeks and 0 days at an outside hospital. She denied any complications at her 6 week postpartum visit at our institution. She chose to bottle feed her infant and they were discharged to home on hospital day two after vaginal delivery. At 7 months postpartum, the patient underwent laparoscopic Heller myotomy with Dor fundoplication without apparent complication, although she did miss her post operative follow up visits at our institution.

DISCUSSION

Conservative management of achalasia during pregnancy can lead to prolonged hospitalization of patients on total parenteral nutrition and increase the risk of esophageal perforation and lower quality of life due to prolonged inability for oral nutrition [3]. Pharmacologic management of achalasia with calcium channel blockers and long acting nitrates is the least effective therapy with significant side effects including headache, hypotension, and pedal edema [4].

Balloon dilation of the lower esophageal sphincter leads to tearing of the muscle fibers and is about 85% effective in relieving achalasia symptoms, which is the most effective of the non-surgical techniques [4]. However, balloon dilation is associated with the highest risk of immediate complications of the non-surgical therapies [5] and is considered less effective in the setting of mega-esophagus [4]. Esophageal perforation occurs in 3% of patients undergoing balloon dilation, which can necessitate emergency surgery to repair the viscous and perform the myotomy [6,7]. Balloon dilation during pregnancy has previously been described for treatment of achalasia, with promising results in the first, second and third trimesters of pregnancy, but evidence is limited to a handful of case reports [8-10].

Surgical management of achalasia can be performed via open laparotomy or laparoscopically, with similar effectiveness [5]. Therefore, definitive surgery is usually performed laparoscopically using Heller's extramucosal myotomy of the lower esophageal sphincter, in which the circular muscle layer is incised and is considered effective in relieving achalasia symptoms in more than 90% of patients [11]. Laparoscopic myotomy is usually the procedure of choice for definitive treatment of achalasia outside of pregnancy [5]. Reflux esophagitis is a common complication after laparoscopic myotomy and can be severe. Pregnancy-specific conditions including increased abdominal protuberance,

increased intra-abdominal pressures and increased risks of reflux esophagitis make the laparoscopic myotomy procedure less-preferred in pregnant women. Acute diaphragmatic hernia during pregnancy has been described after surgical myotomy was performed for achalasia [12].

Finally, endoscopic injection of botulinum toxin A injection in the lower esophageal sphincter is becoming more commonly used in the outpatient setting for achalasia and has the benefit of immediate effectiveness to relieve achalasia symptoms in more than 90% of patients [4,5]. The long term effectiveness of botulinum toxin is diminished compared to other non-surgical and surgical therapies, and symptom-free intervals are usually less than 1-2 years [4,5]. Serious complications associated with botulinum toxin injection are much less common than those of balloon dilation [4]. A meta-analysis demonstrated no serious complications in 151 patients treated with botulinum toxin injection [13]. The botulinum injection procedure is relatively easy to perform [4]. The risk of intra-arterial injection of botulinum toxin A injection are extremely small due to the submucosal direct visualization injection technique. Local injection of botulinum toxin A has rarely been associated with systemic weakness in patients, but typically occurs at doses exceeding 600 units (which are 6 times higher than doses used to treat achalasia) [14]. Additional uncommon risks include esophageal mucosal ulceration, pleural effusion, cardiac conduction defects, and mediastinitis [5].

Our cases demonstrate the safe administration of botulinum toxin A injection into the submucosal tissues of the lower esophageal sphincter in a pregnant women suffering from achalasia with a grossly enlarged mega-esophagus. Patient-specific criteria will help to assess the best possible treatment for achalasia during pregnancy. Clinicians are encouraged to share their experience of treatment of pregnant women with achalasia to provide further evidence for safe therapies as technology continues to improve.

REFERENCES

1. Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson JL, Loscalzo J (Editors). *Harrison's Internal Medicine*, 18th Edition. McGraw-Hill, 2012.
2. ACOG Practice Bulletin Number 52. Nausea and Vomiting of Pregnancy. Reaffirmed 2013.
3. Spiliopoulos D, Spiliopoulos M, Awala A. Esophageal Achalasia: An Uncommon Complication during Pregnancy Treated Conservatively. *Case Rep Obstet Gynecol*. 2013; 2013: 639698.
4. Vaezi MF, Pandolfino JE, Vela MF. ACG clinical guideline: diagnosis and management of achalasia. *Am J Gastroenterol*. 2013; 108: 1238-1249.
5. Stefanidis D, Richardson W, Farrell TM, Kohn GP, Augenstein V, Fanelli RD. Society of American Gastrointestinal and Endoscopic Surgeons. SAGES guidelines for the surgical treatment of esophageal achalasia. *Surg Endosc*. 2012; 26: 296-311.
6. Chuah SK, Wu KL, Hu TH, Tai WC, Changchien CS. Endoscope-guided pneumatic dilation for treatment of esophageal achalasia. *World J Gastroenterol*. 2010; 16: 411-417.
7. Khudyak V, Lysy J, Mankuta D. Achalasia in pregnancy. *Obstet Gynecol Surv*. 2006; 61: 207-211.
8. Clemendor A, Sall S, Harbilas E. Achalasia and nutritional deficiency

- during pregnancy. *Obstet Gynecol.* 1969; 33: 106-113.
9. Fiest TC, Foong A, Chokhavatia S. Successful balloon dilation of achalasia during pregnancy. *Gastrointest Endosc.* 1993; 39: 810-812.
 10. Satin AJ, Twickler D, Gilstrap LC. Esophageal achalasia in late pregnancy. *Obstet Gynecol.* 1992; 79: 812-814.
 11. Zaninotto G, Costantini M, Portale G, Battaglia G, Molena D, Carta A, et al. Etiology, diagnosis, and treatment of failures after laparoscopic Heller myotomy for achalasia. *Ann Surg.* 2002; 235: 186-192.
 12. Khandelwal M, Krueger C. Diaphragmatic hernia after laparoscopic esophagomyotomy for esophageal achalasia in pregnancy. *ISRN Gastroenterol.* 2011; 2011: 871958.
 13. Leyden JE, Moss AC, MacMathuna P. Endoscopic pneumatic dilation versus botulinum toxin injection in the management of primary achalasia. *Cochrane Database Syst Rev.* 2014; 12: CD005046.
 14. Crouner BE, Torres-Russotto D, Carter AR, Racette BA. Systemic weakness after therapeutic injections of botulinum toxin a: a case series and review of the literature. *Clin Neuropharmacol.* 2010; 33: 243-247.

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