

# Dilated Esophagus from Achalasia Presenting as a Neck Mass

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## Abstract

**Objective:** Review management of esophageal achalasia and the workup of adult neck masses in the context of a case report describing severe esophageal achalasia. **Method:** Retrospective chart review case report. **Results:** We report a case of a 64 year old female with chronic progressive achalasia over a 15 year period who presented with a new left sided neck mass. Imaging workup revealed significant esophageal dilation extending to the thoracic and cervical esophagus expanding toward the skin through the infrahyoid strap muscles. **Conclusion:** Management of severe achalasia requires a multidisciplinary approach utilizing multiple medical and surgical management options. Additionally a systematic approach is necessary in evaluating adult neck masses.

## Background

Esophageal achalasia is a degenerative neuromuscular process resulting in failure of the lower esophageal sphincter (LES) to relax resulting in functional obstruction of the esophagus. Clinically this is classically associated with the development of esophageal distention, intermittent dysphagia, cough, regurgitation, halitosis, weight loss and aspiration sensation.

In rare, very severe cases of achalasia there can be significant dilation to the point of cervical esophageal involvement. Such cases have been reported to cause dyspnea and even acute compressive airway compromise<sup>1,2</sup>. On exam these patients have also been noted to have pharyngeal swelling, glottic edema and lateral neck swelling consistent with a cervical mass in some instances<sup>3,4</sup>.

Achalasia is traditionally and definitely diagnosed with esophageal manometry. Those with achalasia will be noted to have impeded LES relaxation and aperistalsis. Radiographic work-up on CT or barium esophagram revealing a tapering esophageal “birds beak” pattern can be used in diagnosing achalasia. Endoscopy can also be useful in patient assessment to identify retained food contents or abnormalities of the gastroesophageal junction<sup>5</sup>. Most importantly endoscopy can be used to rule out mechanical obstruction or pseudoachalasia<sup>5</sup>.

Treatment strategies for achalasia all serve to relax the LES and allow for emptying of the esophagus. Conservative treatment methods involve oral medications to relax smooth muscle such as nitrates, calcium channel blockers and phosphodiesterase-5-inhibitors<sup>5</sup>. Procedures such as endoscopic botox injection or balloon dilation of the LES also serve to relieve symptoms of achalasia. More aggressive and definitive treatments include myotomy or esophagectomy can also be employed in refractory cases.

## Works cited

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## Case Report

- Initial presentation:** A 64 year old female with a history of gastroesophageal reflux disease initially presented in 2009 for otolaryngology evaluation with complaints of a food “catching” sensation in her mid-thoracic area that had been present for the previous 6 years prior to presentation. This sensation was associated with a pressure pain that was alleviated with eructation.
- Initial work-up:** barium swallow study at this time noted a moderately sized hiatal hernia with reflux esophagitis, mild esophageal dysmotility and mild cricopharyngeal dysfunction. Subsequent manometry noted esophageal dysmotility notable for only one peristaltic contraction and non-propagating contractions.
- Initial treatment:** The patient’s symptoms continued to progress and in January 2014 progressed to occasional emesis containing undigested food. At this point patient underwent upper endoscopy with the department of gastroenterology. Findings were consistent with achalasia: endoscopy noted a dilated proximal esophagus with a distal bird-beak appearance along with a lower esophageal resting pressure of 50 (figure 1) with failure of the sphincter to relax. The patient then underwent botox injection of the gastroesophageal junction in January 2014. Post-procedure patient noted significant alleviation of her dysphagia and cessation of regurgitation and episodes of emesis.
- Refractory symptoms:** Patient continued to complain of refractory symptoms over the course of the next several months. Patient underwent multiple botox injection procedures which temporized her symptoms. Patient was counseled on and recommended Heller myotomy which she decided to forgo.
- Presentation of the neck mass:** In June 2018, 2 years after her last botox injection, patient presented in otolaryngology clinic complaining of left sided neck swelling (figure 2) associated with eating along with associated sore throat and cough. Nasopharyngeal laryngoscopy revealed what appeared to be a left sided submucosal mass of the hypopharynx (figure 3). Patient was also noted to have a indistinct soft tissue swelling the in left anterior neck in the region of level III.
- Work-up and management of the neck mass:** Patient subsequently underwent a CT neck which revealed thickening of the left posterior wall of the oropharynx and severe dilation of the cervical and thoracic esophagus which expanded between the strap muscles and toward the skin (figure 4). There were no masses or pathologic appearing lymph nodes appreciated on this study. Surgical evaluation for Heller Myotomy was offered; however, patient continues to elect for conservative management and observation.

## Manometry

PATIENT	NORMAL VALUES
<b>LOWER ESOPHAGEAL SPHINCTER</b>	
(LES Location: 38 cm)	
Resting Pressure: 50 mmHg	10-45 mmHg
Relaxation: Complete	Complete
Incomplete_x_	
Residual Pressure: 24.0 mmHg	≤ 8 mmHg
Length of LES: 3 cm	3-5 cm
<b>ESOPHAGEAL BODY</b>	
Distal (10 Wet Swallows)	
#Peristaltic Waves: 0	8-10 Peristaltic
Average Amplitude of peristaltic waves: mmHg	30-180 mmHg
Average Duration of peristaltic waves: seconds	3-6 seconds
#Simultaneous: 0	< 2 simultaneous
#Not Transmitted (< 30 mmHg): 10	< 3 not transmitted
#Triple Peaks: 0	*Never seen in normals
Proximal (1 Wet Swallows)	
Average Amplitude: mmHg	No normal values
Peristalsis (Distally) Yes/No	

Fig 1. Patient manometry

## Discussion

The above described case is a severe and rare presentation of an uncommon condition. Achalasia can typically be treated conservatively with medications and endoscopic injections; however, this patient’s condition progressed despite these interventions. This case highlights the importance of a broad differential and systematic approach to patient evaluation.

Upon evaluation in otolaryngology clinic the patient’s clinical vignette of a progressive anterolateral neck mass with hypopharyngeal swelling would alone suggest pathologies such as an infectious or neoplastic process. In accordance with the American Academy of Otolaryngology clinical practice guidelines for evaluation of an adult neck mass, it is crucial to obtain a thorough history, physical and perform imaging studies to prevent unnecessary procedures. This is highlighted in this case as the morbidity associated with biopsying the neck swelling and causing a potential esophageal perforation was avoided.

This case also points to how achalasia refractory to conservative measures can progress when not addressed surgically. By forgoing myotomy the patient’s achalasia continued to evolve to the cervical esophagus. Despite these recommendations the patient elected for less invasive measures and follows under close observation of the otolaryngology and gastroenterology services. This highlights the importance of a multiple team approach and the principle of patient autonomy.

## Clinical and radiographic findings



Fig 2. Left anterolateral neck swelling



Fig 3. Stroboscopy revealing left posterior hypopharyngeal wall prominence



Fig 4a. Axial CT demonstrating left esophageal dilation

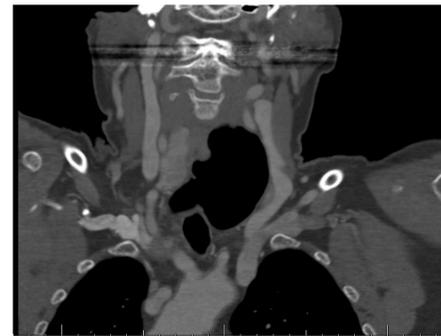


Fig 4b. Coronal CT demonstrating left esophageal dilation



Fig 4c. Sagittal CT demonstrating left esophageal dilation