Abstract: Achalasia is an idiopathic disorder of esophagus involving lower esophageal sphincter (LES), characterized by insufficient relaxation of LES and absence of esophageal peristalsis. We present a case report of a 44 year old female presenting with complaints of difficulty in swallowing for ten years. She underwent laparoscopic Heller's myotomy and anterior fundoplication for the disease but continued to have symptoms of dysphagia. On evaluation upper GI endoscopy showed features of dilated esophagus suggestive of achalasia. Esophageal manometry, barium esophagogram and CT chest with abdomen revealed dilated esophagus suggestive of achalasia. Endoscopy showed end stage achalasia and biopsy revealed severe dysplasia. Laparoscopic assisted transhiatal esophagectomy was done for the patient. Histopathology report showed features of dysplasia and carcinoma in situ. Definitive treatment was done for this unique condition of end stage achalasia after failed myotomy and histological features of severe dysplasia in the esophagus.

Keyword: Achalasia, Heller's myotomy, Severe dysplasia, Transhiatal esophagectomy, Failed myotomy

INTRODUCTION
Achalasia is an idiopathic disorder of esophagus involving lower esophageal sphincter (LES), characterized by insufficient relaxation of LES and absence of esophageal peristalsis. Pneumatic dilation or Heller's cardiomyotomy with fundoplication are the preferred therapies for achalasia. Long standing achalasia can lead to squamous cell carcinoma of esophagus and adenocarcinoma of esophagus. We present the case report of a patient with achalasia with previous failed myotomy with symptoms and endoscopic biopsy showing dysplasia.

CASE REPORT
44 Year female presented with complaints of difficulty in swallowing for 10 years, initially for solids and then for liquids. She also presented with history of food regurgitation and retrosternal discomfort. She complained of loss of weight and foul smelling vomitus on and off. She was evaluated and diagnosed to have achalasia cardia 8 months back.

She underwent laparoscopic Heller's myotomy and anterior fundoplication for the above condition. Patient continued to have dysphagia in the post operative period. Patient was evaluated in another hospital and endoscopy showed end stage achalasia with severe dysplasia in the mid esophagus. She was then referred to our medical gastroenterology department for dilatation. They decided not to dilate because of the presence of dysplasia and referred the patient to our department for definitive management.

The patient was evaluated. Upper gastrointestinal endoscopy showed grossly dilated esophagus and achalasia cardia. OG junction at 38cm. Biopsy from mid esophagus done showed severe dysplasia.

Esophageal manometry done showed features of achalasia cardiotype 1. CT Chest and Abdomen revealed dilated esophagus, diameter 6.7cm. Barium swallow showed dilated esophagus suggestive of end stage disease.
The patient is diagnosed to have end stage achalasia cardia post Heller’s myotomy with anterior fundoplication with severe dysplasia in mid esophagus. Laparoscopic assisted transhiatal esophagectomy was done for the patient.

**Intra operative finding are:**

**Discussion**
Achalasia is characterized manometrically by insufficient relaxation of the lower esophageal sphincter (LES) and loss of esophageal peristalsis; radiographically by aperistalsis, esophageal dilation, with minimal LES opening, “bird-beak” appearance, poor emptying of barium; and endoscopically by dilated esophagus with retained saliva, liquid, and undigested food particles in the absence of mucosal strictureing or tumor. Achalasia must be suspected in those with dysphagia to solids and liquids and in those with regurgitation unresponsive to an adequate trial of proton pump inhibitor (PPI) therapy. The peak incidence occurs between 30 and 60 years of age.

The pathology is degeneration of ganglion cells in the myenteric plexus of the esophageal body and the LES leads to loss of inhibitory neurotransmitters nitrous oxide and vasoactive intestinal peptide and consequently imbalance between the excitatory and inhibitory neurons.

**Treatment Algorithm**
Recommendations for achalasia management:

Either graded pneumatic dilation (PD) of laparoscopic surgical myotomy with a partial myotomy or fundoplication are recommended as initial therapy for the treatment of a achalasia in those fir and willing to undergo surgery. PD and surgical myotomy should be performed in high-volume centers of excellence. The choice of initial therapy should be guided by patients’ age, gender, preference, and local institutional expertise. The therapeutic choice is between pneumatic dilation and laparoscopic Heller myotomy as the primary therapy for achalasia. Achalasia management differ according to manometry subtypes. Achalasia with esophageal compression responded favorably to any therapy (botulinum toxin, PPI therapy may be indicated in those who complain of heartburn. (2) Studies support that a postdilation LES pressure of 10 mm Hg in manometry associated with a higher rate of remission. Surveillance endoscopy for esophageal cancer is not recommended. In our case endoscopy was done because patient presented with symptoms after myotomy. Some experts are in favor of some form of endoscopic or radiographic surveillance in patients with achalasia if the disease has been present for more than 10–15 years with an interval of every 3 years. (3)

End Stage Achalasia

Severe achalasia is defined by a diameter >6 cm, whereas others consider the presence of a distal angulation and a sigmoid-like configuration as markers of end-stage achalasia and a doomed myotomy. Our patient had the features of end-stage achalasia. (6,7)

Hallmarks of End-Stage Achalasia

Clinical: Severe dysphagia and/or regurgitation Radiographic: Massive esophageal dilation (megaesophagus) and/or tortuosity (sigmoid esophagus) Pathologic: Reduction or absence of ganglion cells with fibrous replacement of the myenteric plexus(2)

Approximately 10–15% of patients who have undergone treatment for achalasia will have progressive dilation of the esophageal diameter and will fulfill criteria for megaesophagus or end-stage achalasia and up to 5% of patients may require esophagectomy. (7,8)

Management of treatment failures

In those failed myotomy, the rates of symptom response after PD and repeat myotomy were only 67% and 57%, respectively, with 8 patients eventually requiring esophagectomy. For these patients, the best chance of success is a multimodality approach that may include botox, PD, re-do myotomy, and, as a last resort, esophagectomy. Our patient needed sophagectomy because of the unique clinical presentation. (4,5)

Achalasia and malignancy

Bolivar and Herendeen observed 0.7-7.7% of clinically determined malignancy but a higher incidence of 20-29% malignancy at autopsy. Chronic stasis of food and bacterial overgrowth seen in long standing achalasia is responsible for dysplasia and eventual esophageal carcinoma. Although squamous cell carcinoma is the most commonly associated malignancy, adenocarcinoma in Barrett’s metaplasia has been reported in achalasia patients developing chronic gastroesophageal reflux after pneumatic dilation. The treatment of choice for early stage cancer is esophagectomy.

Our patient presented with endstage disease with severe dysphagia and hence esophagectomy was done. (2) Indications for esophagectomy in achalasia:

1. Ulceration, bleeding, fistulization, or perforation
2. Posttreatment reflux esophagitis/trictrure
3. Development of carcinoma
4. Inadequate nonoperative therapy (botulinum toxin [Botox], pneumatic dilation)
5. Inadequate surgical therapy

Incomplete myotomy

Healing of myotomy

Recurrent hiatal/paraesophageal herniation

Technically flawed fundoplication

Other technical problems (excessive hiatal closure, angulation)

CONCLUSION

Patients of achalasia with malignancy are usually diagnosed late and multimodality therapy may prolong the survival. Regular pneumatic dilation or cardomyotomy does not protect from the occurrence of cancer in achalasia. Though endoscopic pneumatic dilation is a universally accepted therapy, there is no consensus for endoscopic screening in achalasia. Endoscopic surveillance in patients with longstanding achalasia may detect cancer in early stage so that patient will be benefited with curative therapy. In our case since the patient presented with symptoms of dysphagia and features of end stage disease after previous myotomy. Early endoscopy was done and severe dysplasia of esophagus was the indication for esophagectomy in our patient.

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