Case Report

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A case report on achalasia cardia type - II

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ABSTRACT

Achalasia cardia is the rare chronic neurodegenerative disorder of the esophagus which causes progressive delay in contractility of lower esophageal muscles during swallowing leading to backup of food contents and fluids in later stages within the region of esophagus. It is considered as the common leading cause of motor dysphagia. The underlying etiology include autoimmune disorder, nervous degeneration due to loss of inhibitory ganglion in myenteric plexus of esophagus, presence of inhibitory neurotransmitters such as nitric oxide and its receptors in lower esophagus. At initial presentation drugs are prescribed as the symptoms mimic other GI disorders which often lead to delayed diagnosis. The symptoms mainly include severe pain or discomfort of chest after eating, weight loss, regurgitation of bland undigested food or saliva, severe heartburn, dysphagia, aspiration. Initial treatment includes the pharmacotherapeutic management to alleviate the symptoms and improve the esophageal outflow. Surgical treatment involves pneumatic dilatation, Heller's myotomy, preoral endoscopic myotomy, esophagectomy. Heller's myotomy may be performed by open or laparoscopic procedure. It is done along with Dor's fundoplication. Case report discussed below is of a 55 years old female patient presented with dysphagia for one year. The symptoms of which worsened for 2 months along with vomiting of undigested foul-smelling food.

Keywords: Achalasia cardia, Motor dysphagia, Neurodegenerative disorder, Pharmacotherapeutic management, Esophageal motor disorders

INTRODUCTION

Achalasia remains a disorder of idiopathic aetiology. It occurs from progressive degeneration of ganglion cells in the myenteric plexus in the esophageal wall, leading to failure of relaxation of the lower esophageal sphincter which is accompanied by a loss of peristalsis in the distal esophagus. The disease is insidious in onset and the progression is gradual. Patients classically experience symptoms for years before seeking any medical consult. In one study patients were prospectively interviewed and found 87 patients with freshly diagnosed achalasia wherein the mean duration of symptoms was 4.7 years before the diagnosis. In this disease, there is inhibitory

nerves degeneration in the esophagus which causes an unrestricted action of the excitatory neurotransmitter such as acetylcholine and this leads to contractions of high amplitude. This then results in the progressive damage to the neurons of cholinergic system and hence subsequent to dilation accompanied by concurrent contractions of low amplitude in the esophagus.⁴

The pharmacological management is directed towards alleviating symptoms by decreasing the resting pressure of the nonrelaxing LES. Drugs such as antacid, proton pump inhibitors, calcium channel blockers, long acting nitrates, phosphodiesterase 5 inhibitors, anticholinergics and beta-adrenergic agonists are used for the management. Antacids are mainly given to alleviate the

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symptoms of Achalasia. They decrease the amount of acid reaching the duodenum by neutralizing the acid present in the stomach and thus reduce pain.⁵ Proton pump inhibitors (PPIs) inhibits gastric H+/K+-ATPase by inhibiting proton pump which leads to inhibition of gastric aid production which then reduces heartburn and regurgitation.⁶ Calcium channel blockers (CCBs) include Nifedipine, Verapamil, diltiazem etc. which reduce the lower esophageal sphincter (LES) pressure and thus improves the symptoms of dysphagia.7 Sublingual isosorbide dinitrate 2.5 to 5 mg or oral 10 to 20 mg given 20 to 30 minutes before meals improve symptoms of dysphagia. It is a long acting oral nitrate which acts by relaxing the vascular smooth muscle.⁸ Sildenafil, a phosphodiesterase type 5 inhibitor degrades nitric oxide (NO) stimulated by cyclic monophosphate (cGMP), thus relaxing smooth muscle cells of esophagus. It lowers LES pressure and pressure in the body of the esophagus. Anticholinergics include atropine, dicyclomine, cimetropium bromide reduces LES pressure and emptying.¹⁰ Beta-adrenergic improves esophageal agonists mediated by receptors 1 and 2 also reduce the mean LES pressure and thus relaxation.11

Pharmacological treatment is given in initial stages and in patients intolerable to surgery. Endoscopic botulinum toxin injection is given in the lining of lower esophagus which inhibits the release of acetylcholine at presynaptic terminal. This blocks the unrestricted excitatory cholinergic stimulation that causes paralysis of the sphincter muscle due to neuronal inhibition. The nonsurgical treatment includes pneumatic dilatation. It is done by using graded, sized polyethylene balloons that are intra-luminally dilated which leads to disruption of the LES circular muscle fibers due to air pressure. Surgical treatment includes Heller's myotomy, preoral endoscopic myotomy, esophagectomy. In Heller's myotomy, the muscles of the lower esophageal sphincter are cut allowing food and liquids to permit to the stomach.12

Preoral endoscopic myotomy (POEM) is a minimally invasive surgical procedure for the treatment of achalasia in which the inner circular muscle layer of the lower esophageal sphincter is divided through a submucosal tunnel.¹³ The complication of esophageal myotomy include gastroesophageal reflux disorder (GERD), esophageal perforations, post-operative dysphagia. Antireflux surgery may be performed to prevent postoperative reflux.¹⁴ Posterior fundoplication may be performed for preventing post-operative dysphagia and it is more superior to anterior fundoplication.¹⁵ One of the studies concluded that Heller Myotomy plus Dor Fundoplication was superior to Heller myotomy alone in regard to the incidence of postoperative GER.¹⁶ Laparoscopic myotomy is the preferred treatment that durably relieve symptoms of achalasia as it can durably relieve symptoms of dysphagia.¹⁷ Esophagectomy is reserved for patients with end-stage achalasia in whom surgery isn't contraindicated or pneumatic dilatation with or without Heller myotomy has been unsuccessful. 18

CASE REPORT

A 55 years old lady, presented with progressive dysphagia for 1 year. Her symptoms worsened and increased for the past 2 months. She experienced progressive dysphagia (solids>>liquids), vomiting, and loss weight. She complained of progressive increase in difficulty swallowing for 2 months. She described that she was able to tolerate liquids with difficulty and also experienced vomiting's for past 1 month. Her vomitus consisted of undigested foul-smelling food which regurgitated immediately. No abdominal pain was complained. She also complained of unintentional weight loss which was about 8 kgs. On general examination no icterus, pallor, lymphadenopathy, or edema was observed.

Surgical history of tubectomy done 20 years ago. She is a housewife and non-smoker. No significant family history was observed. No addictions found.

Her vitals were recorded. Temperature was 98.2°F, heart rate was 78/ min, blood pressure 120/80 mmHg, and respiratory rate 17/ min. PFT performed one month back was not suggestive of any restrictive/ obstructive lung diseases. SpO₂ at room air was 99%. Lungs were clear. Spirometry was done and it was found to be normal. Other systemic examinations were also done which were also normal.

Complete blood picture (CBP) revealed hemoglobin 11 g/dl, WBC 7200 cells/ cmm, normal differential count, platelets adequate. Serum electrolyte test showed sodium (Na+) 141 mmol/l, potassium (k+) 3.6 mmol/l, chloride (Cl⁻) 116 mmol/l.

Initially when she complained of heart burn and difficulty swallowing, she was prescribed antacids such as magaldrate and simethicone. But that didn't show significant improvement.

When the symptoms worsened a list of diagnostic examinations were advised which included ECG which showed normal sinus rhythm, GI endoscopy revealed abnormally dilated esophagus with residual food even after 12 hours of fasting. Contrast enhanced X-ray with barium swallow (Figure 1) showed that the bilateral costophrenic angles are free, dilated esophagus, stricture detected at LES and bird's beak sign of esophagus. Esophageal manometry (Figure 2A and B) was performed by perfusion system. Wet swallow was done by 5ml water. Basal esophago-Gastric Junction (EGJ/JES) pressure was normal - 30 mmHg. EGJ relaxation to wet swallows was inadequate. Esophageal peristalsis was absent with wet swallows. Simultaneous contractions of low amplitude noted (Pan Esophageal compression). Maximal amplitude of contractions was 20 mmHg. The puckering at esophagogastric junction suggested achalasia cardia type-II.

After examining the symptoms and investigations of lab and radiology data, she was prescribed calcium channel blocker that is nifedipine as a plan for initial management prior to surgery to prepare the patient for surgery.

Laparoscopic Heller's myotomy along with Dor's partial fundoplication was performed. Cardiomyotomy was performed to a length of 6–7 cms by dividing longitudinal circular layers.

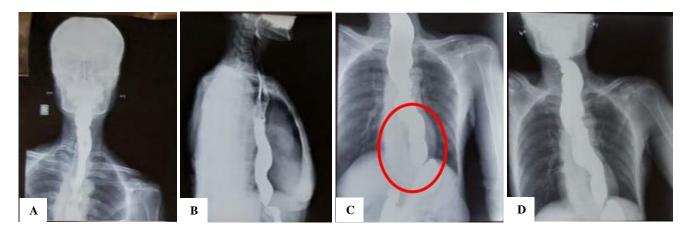


Figure 1: Barium swallow showing esophageal dilatation and stricture with images showing different views. (A) AP view, (B) lateral view; (C) dilatation seen; (D) esophageal dilatation and stricture.

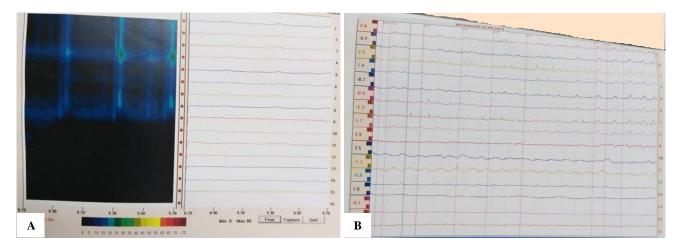


Figure 2: Esophageal manometry. (A) Esophageal manometry – I, (B) esophageal manometry – II.

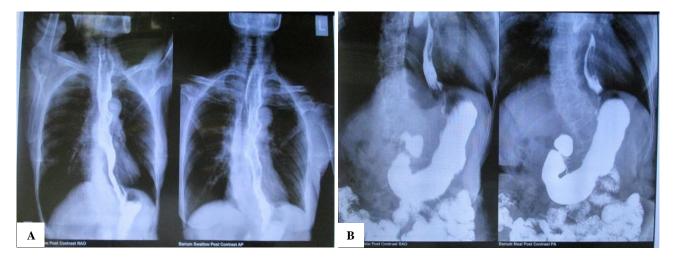


Figure 3: Post-operative Barium swallow post contrast; (A) AP view; (B) PA view.

Post-surgery

After performing the surgery, patient experienced postoperative surgical site pain for which analgesic was given. IV fluids dextrose normal saline and ringer lactate initiated. For the prevention of surgical site infection antibiotic combination cefoperazone and sulbactam was administered. Liquid diet was initiated on second day after surgery. Semisolid food was initiated on day three in the absence of presurgical symptoms. Post-op X-ray (Figure 3A and B) was done with barium contrast and it wasn't initially entering the stomach but entered only after delay.

Follow-up

The patient was reviewed after 7 days for dressing and post-surgery checkup. The patient was able to take foods and no difficulty swallowing was observed.

Post-operative X-ray (Figure 3A and B) was taken to review the condition.

DISCUSSION

Achalasia cardia is a disorder affecting the esophageal motility. Patients suffering from this disorder have imbalance of excitatory and inhibitory neurotransmission which results in non-relaxing lower esophagus. It is the rare disease and common cause of motor dysphagia. The incidence of achalasia has significantly increased from 1.07 to up to 2.8 in a population of 100,000 per year annually. It is mostly diagnosed in individuals of age 30 to 60 years.

In this case, the patient is a 55 years old female. Patients having achalasia usually present with difficulty swallowing food, chest discomfort, heartburn, loss of weight, vomiting, and difficulty swallowing liquids in severe cases. 1,21 However, heartburn and regurgitation are frequently observed in patients who have GERD. Hence the diagnosis of achalasia might be delayed and the patients are not responsive to the pharmacological therapy because these symptoms are often misinterpreted as gastroesophageal reflux. For such patients who do not respond to pharmacological treatment, esophageal manometry should be performed to exclude esophageal motility disorders including achalasia. 22 Similar condition was observed in this case that the diagnosis of achalasia was delayed and patient was on pharmacological therapy for GERD until the patient presented with progressive dysphagia followed by vomiting of undigested food. Upper GI endoscopy was then performed as initial step up investigations which revealed narrow lower esophageal sphincter. However, this doesn't confirm the diagnosis of achalasia.

Contrast enhanced X-ray with barium swallow is performed for patients with achalasia. In such patients, there is typically a dilated esophagus, absence of

peristalsis, and narrowing of the distal esophagus in a typical "bird's beak" appearance. ²¹ In addition to barium swallow, Oesophageal manometry is performed which stands as the gold standard in the diagnosis of achalasia¹. In this patient both barium swallow along with oesophageal manometry was performed that revealed typical bird's beak appearance and also confirmed the diagnosis of type II achalasia. This has also been observed in the previous studies. ^{24,25} Heller's myotomy along with Dor's fundoplication is the highly effective treatment option after the diagnosis of achalasia. 17,19,21,24 The same was performed as the patient was not willing for a trial of pneumatic dilatation when she was counselled for the risk of perforation of esophagus and chances of remission.²⁵ Dor's fundoplication was performed to prevent post-operative reflux. We are reporting a case of typical Achalasia cardia type-II and the key findings of this report is that the patient recovered completely with no postoperative symptoms. On subsequent follow-ups after 3 months and 6 months patient was completely without symptoms. We infer that the surgical myotomy is still the gold standard in the management of type 2 achalasia. Other non-surgical methods are reserved for patients who are not willing or are not fit for surgery. However, such conservative methods such as Botox injections, stenting, pneumatic dilatation either tend to fail or there is higher risk of symptom recurrence.¹⁹

CONCLUSION

This is being reported as it is the rare case and that patient was apparently well. Surgical therapy is a gold standard in the treatment of type II achalasia. All patients who do not respond to pharmacological therapy and who were diagnosed with GERD or gastritis should undergo GI endoscopy and differential diagnosis for achalasia must be done. If in case the symptoms worsen and progressive dysphagia occurs following which the therapy should be individualised and planned accordingly.

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