Case Report

Achalasia cardia: A Case Report and a Brief Review

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Achalasia cardia (AC) is one of the rarely seen disorders of esophagus motility which happens as a result of degeneration of neurons which is irreversible. Treatment is mainly of palliative nature, and no complete curative treatment is available as of now. The diagnostics and therapeutics in this direction has strengthened mainly due to high-resolution manometry and per-oral endoscopic myotomy, referred to as Peroral endoscopic myotomy (POEM).The characterization of type of achalasia cardia is made easier by high-resolution manometry as we have mentioned, which also has an important therapeutic role. Achalasia has to be managed for each patient individually, and the role of pneumatic balloon dilatation, POEM, or Heller's myotomy needs to be reconsidered. In our case report and brief review, we discussed the main aspects of diagnosing an OPD-based patient and a brief review of achalasia cardia.[1]

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Introduction

Thomas Williams of England firstly described achalasia cardia in the year 1672.[2] As we know Achalasia cardia is a rare disorder of motility of the esophagus which results due to autoimmune neurodegeneration of the myenteric plexus of the esophagus. Although it is rare, but the most common and it is the best-characterized motility disorder of the esophagus.The primary etiology of this disease differs from other motility disorders (e.g., Jackhammer esophagus and distal oesophageal spasm) in the failure of lower oesophageal sphincter (LES)to relax in case of achalasia. Achalasia cardia is mainly divided into three subtypes manometrically that aid treatment and its management and hence have a good prognostic significance.[3]

Epidemiology

As per a study based in Canada the incidence and prevalence of achalasia is 1.63/100,000 and 10.82/100,000. It was 0.5/100,000 and 8/100,000 in the United Kingdom.[4] Both genders equally affected. Common between ages and 25 and 60 years but it can be seen at any age. In a Lucknow-based study on the numerous causes for motor dysphagia that was seen, 77% of the subjects in this study were diagnosed to have achalasia.[5]

Case Report

A 29 years old patient presented to our Ent OPD with complaint of difficulty in deglutition since 1 year.

The medical history of our case shows that the patient was apparently alright then at 28 years of age when he complained for the first time symptoms of dysphagia which disturbed him atleast once in around every 2 months and these symptoms continued until he was 2w9 years of age when dysphagia became more frequent and occurred once or twice in every 15 days. The patient also had a history of weight loss but there was no history of anorexia as such.

Clinical Examination And Investigations

Physical examination of the patient revealed no abnormality. Blood works that we got done like complete blood count and other routine investigations, were normal. Barium swallow Esophagus x-ray was done and dilated distal esophagus was seen in it along with a rat tail appearance. Computed Tomography of chest revealed dilatation (Fig 1 and 2) at the level of the distal esophagus. On Upper GI Endoscopy dilatation on the lower third of the esophagus was noted and features of achalasia cardia

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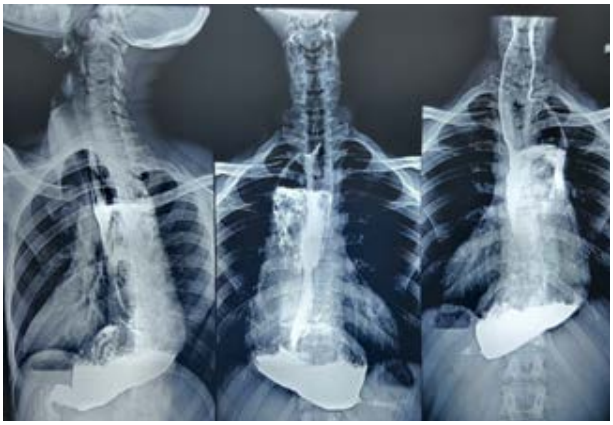

Fig 1: Barium swallow oesophagogram showing Rat tail appearance.

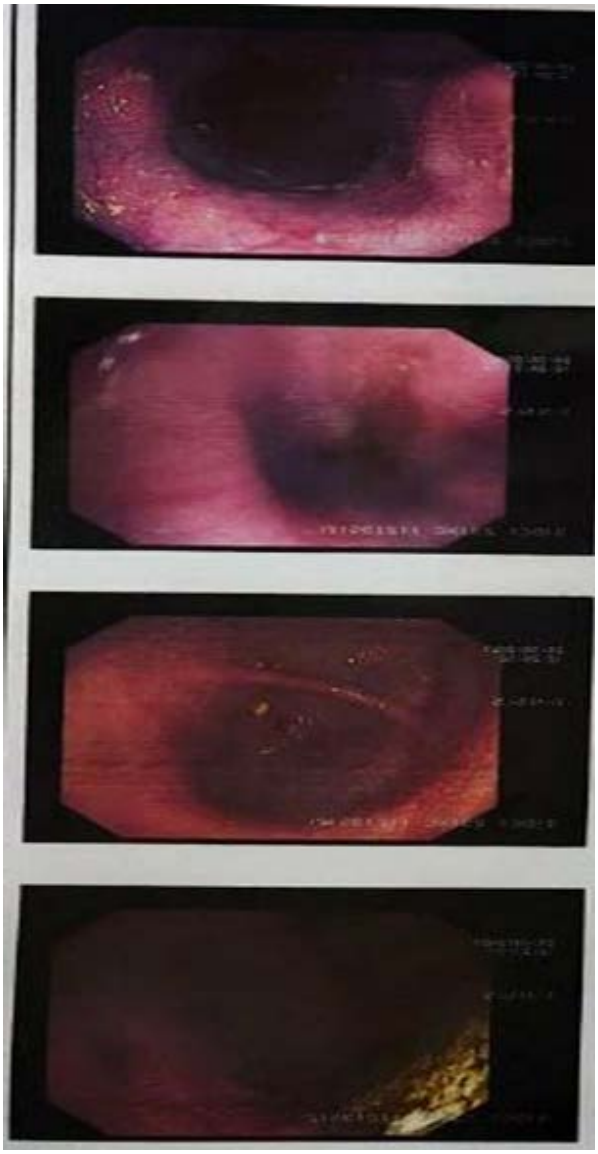

Fig 3: Upper GI Endoscopy revealing features of Achlasia cardia.

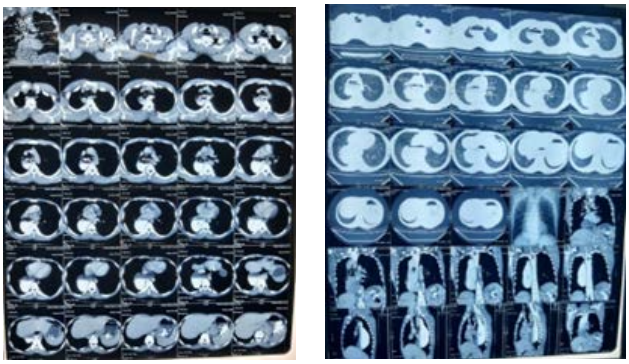

Fig 2 (a,b): CT scan of neck and chest suggestive of a distally dilated esophagus

Table : ECKARDT SCORE:6

Score	Dysphagia	Regurgitation	Retrosternal pain	Weight loss (kg)
0	None	None	None	None
1	Occasional	Occasional	Occasional	<5
2	Daily	Daily	Daily	5-10
3	Each meal	Each meal	Each meal	>10

and based on these examinations and investigations, we made the diagnosis of achalasia cardia.

Discussion

The excitatory (eg, acetylcholine, substance P) and inhibitory (eg, nitricoxide, vasoactive intestinal peptide) neurotransmitters are responsible for regulating the pressure and relaxation of lower esophageal sphincter (LES) in normal physiological state.But in patients who suffer from achalasia, lack nonadrenergic, noncholinergic, inhibitory ganglion cells,as a result of which an imbalance is caused in the excitatory and inhibitory neurotransmissionof signals,due to which esophageal sphincter is non relaxed.We can say that the primary cause to this is still not known.Myenteric neurons coordinate and mediate the peristalsis of the esophagus and LOS tone but in this condition, the neurons are either less or not present, due to which there is impaired relaxation of the LOS. Also observed were the antibodies against these neurons in serum samples.

Achalasia cardia is mostly seen in adults most commonly in the age group of 25 to 60 years and the male to female ratio is 1:1. Here, the OPD patient is a male of 29 years.

The most common presentation of achalasia is dysphagia to solids as well as liquids from the start (in around 85–91% of patients)and the second most common presenting symptom is the regurgitation of undigested food particles (in about 75–91% of patients). Food regurgitates before reaching the stomach, which is uncommon in GERD. Also seen are the chest pain(mainly retrosternal) and retrosternal burning sensation in about 60% of patients,which ends up as achalasia misdiagnosed as GERD and hence delay in diagnosis.Heartburn results due to undigested and fermented carbohydrate which results in production of lactate .Weight loss can also be seen but it is not as common as in case of mechanical causes. The Eckardt score is given on the above mentioned presentation i.e, the dysphagia degree, regurgitation of contents,pain in chest, and weight loss, and treatment efficacy of achalasia is evaluated by it..Impaired belching is a rare but known symptom in achalasia. Few patients present with emaciation and ulcers in oral cavity which is mainly due to regurgitation of the acidic food material. The clinical diagnosis of AC should be made by a barium swallow oesophaagus xray, which is the best diagnostic modality.The characteristic of achalasia is a sharp wooden pencil tip appearance which is commonly known as **"Rat Tail Appearance"**. Endoscopic examination is ideal for all patients. Another study that is the manometry diagnoses AC. It shows that pressure at GE junction is twice the normal pressure (40 mm of Hg) and relaxation tone here is lost. In our case, x-ray showed a rat tail appearance. Then upper gastro intestinal endoscopy showed lower third esophagus dilatation. Computed Tomography showed distal esophagus dilatation.So, as per all of the above findings, the patient was diagnosed to be suffering from achalasia cardia (Fig 3).[7]

The Chicago classification released by HRM in 2009 for categorizing esophageal motility disorders has been revised multiple times since then.The Chicago classification categorizes achalasia into three manometric subtypes by hierarchical approach *viz.*, Type I AC, known as classic achalasia, in this there is absence of contractility of esophageal body smooth muscle along with compartmental pressurization absence.These changes are commonly seen at later stages achalasia.,The most commonest AC *i.e*, Type II AC shows periods of esophageal compression due to the build up pressure; the esophageal smooth muscle maintains its tone and the pan-esophageal isobaric pressure increases which can be seen on swallowing. The least common type of AC isType III of achalasia in this spastic contraction of the distal end of esophagus is noted.[8]

The management lies in relieving the symptoms by decreasing the pressure.Once the obstruction is taken care of relieved, the food passes by act of gravity through the peristalitic esophageal body. Akso given are Calcium channel blockers and nitrates which act by decreasing the LES pressure,this provides relief in around 10% cases.This treatment is mainstay and can be given to patients who are not fit for any sort of surgical intervention. Another modality is the Endoscopic treatment which coprise of botulinum toxin intrasphencteric injection which acts by inhibiting the acetylcholine release at LES, therefore it restores the balance between both the neurotransmitters i.e excitatory and inhibitory. Though this has not shown much effective results.. Repeated botulinum toxin injections are needed in most of the patients,also recommended is Pneumatic dilatation in the cases where surgery is not possible.Many surgeons consider a laparoscopic Heller myotomy the best primary modalities of patients with achalasia cardia.This along with a partial fundoplication performed thoracoscopically shows high incidence of GERD at the end.

Here, we prescribed the patient the patient with proton pump inhibitor and calcium channel blocker after which significant clinical improvement was seen. The patient showed clinical improvement on his fortnightly follow ups. The patient was then called for follow-up once a month in the OPD.

Conclusion

We are here reporting a case of a male who is 29 years old who suffering from achalasia cardia.after,initially treating him with above mentioned medication on OPD basis he was then planned for a fortnightly follow up and then planned for monthly OPD follow up.

Our patient is still on regular follow up and has shown tremendous improvements in his symptoms just by our conservative management.

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