

End Stage Achalasia Cardia Masked by Chest Trauma: A Case of Diagnostic Mimicry

J.I. Ahmad¹, M.M.Tetty²

¹Consultant Cardiothoracic Surgeon, Division of Cardiothoracic Surgery, Department of Surgery, Aminu Kano Teaching Hospital, PMB 3452, Kano, Nigeria

²Consultant Cardiothoracic Surgeon, National Cardiothoracic Centre, Korle-Bu Teaching Hospital, Accra, Ghana,

Correspondence to: Dr. Jameel Ismail Ahmad, E-mail: ijameel@yahoo.com

Background: Achalasia is the commonest primary esophageal motility disorder. Long standing achalasia reaches the end-stage form when the oesophagus becomes dilated and tortuous and often amenable only to esophagectomy. At this stage the diagnosis may not be straight forward. Our objective is to emphasise the need for detailed evaluation of chest trauma patients who may have coincidental background chest diseases such as end-stage Achalasia.

Case Report: We report a 52 year old lady with end-stage achalasia who presented elsewhere with blunt chest trauma and the radiographic features were misdiagnosed as the sequelae of the chest trauma or a chest tumour and then referred to us. Thorough evaluation led to the diagnosis of the end-stage Achalasia and she successfully underwent transhiatal esophagectomy.

Conclusion: There is need for detailed evaluation of chest trauma patients and high index of suspicion for concomitant conditions should be demonstrated to prevent complications and improve the patients' outcome.

Key words: End-stage Achalasia, Misdiagnosis, Chest trauma

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Introduction

Achalasia cardia is a primary oesophageal motility disorder characterized by failure of relaxation of the lower oesophageal sphincter (LES) during swallowing and aperistalsis of the oesophageal body. Achalasia has an incidence of 1 in 100,000 individuals and a prevalence of 10 in 100,000 with equal gender and racial preponderance¹. The peak incidence is in 20-50 years age group². The aetiology is unknown but autoimmune, viral and neurodegenerative processes are thought to be responsible.

Achalasia when left untreated can progress to an end-stage form and lead to complications which can make the diagnosis difficult. Concomitant other conditions may also mask the diagnosis. This can lead to inappropriate treatment and further complications. The gold standard investigation for the diagnosis of Achalasia is oesophageal manometry although it is rarely available in our setting. When the diagnosis is made early, modified Heller's esophagocardiomyotomy is the treatment of choice and has good outcome. Transhiatal esophagectomy with cervical oesophagostomy is the treatment of choice when the oesophagus becomes markedly dilated and tortuous assuming a sigmoid shape.

Case Report

We report a 57 year old woman referred to our centre with 2 months history of breathlessness, easy fatigability and right sided chest pain. She was involved in a road traffic accident 9 months earlier when she sustained right clavicular fracture which was managed conservatively. Chest

radiographs showed opacity occupying most of the right lung zones which persisted on the follow up chest X-rays (Figure 1). She was referred to us with differential diagnoses of right lung collapse, fibrothorax from clotted Haemothorax and right lung tumour.

Further review revealed a prolonged history of intermittent dysphagia for liquids and solids for the past 20 years. She had an episode of haematemesis and passage of malaena 5 months prior to presentation. No odynophagia or significant weight loss. She is a known hypertensive on medications and had hysterectomy 10 years earlier.

On examination, she was not pale, afebrile and haemodynamically stable. Respiratory rate was 18cpm and there was decreased right chest movement with dullness and decreased air entry over the right middle and lower chest zones.

A Chest CT Scan requested before obtaining the history of dysphagia suggested a massive dilatation of the oesophagus. The anteroposterior diameter of the esophagus at the upper, middle and lower chest zones are 6.8cm, 7.4cm and 10.07cm respectively with an average diameter of 8.09cm(Figure 2). Barium swallow was done which showed features of achalasia cardia with grossly dilated and tortuous oesophagus (sigmoid oesophagus) and an upper gastrointestinal endoscopy showed a dilated and roomy oesophagus, stenosed gastroesophageal junction and small superficial gastric ulcers. Other baseline haematological investigations were normal

The patient was counseled and prepared for oesophagectomy. Transhiatal oesophagectomy with cervical oesophagogastrostomy was done. The postoperative recovery was stable and she was commenced on graded oral sips 7 days postoperatively. The chest tube was removed 2 days later and she was discharged on 12th day postoperatively. When seen 2 weeks at the out-patient department clinic she was stable. She was noticed to develop keloid on the scar 6 months later but she has remained stable and satisfied 18 months after the oesophagectomy.



Figure 1. Chest X-ray for end-stage Achalasia showing right chest opacity due to the dilated esophagus

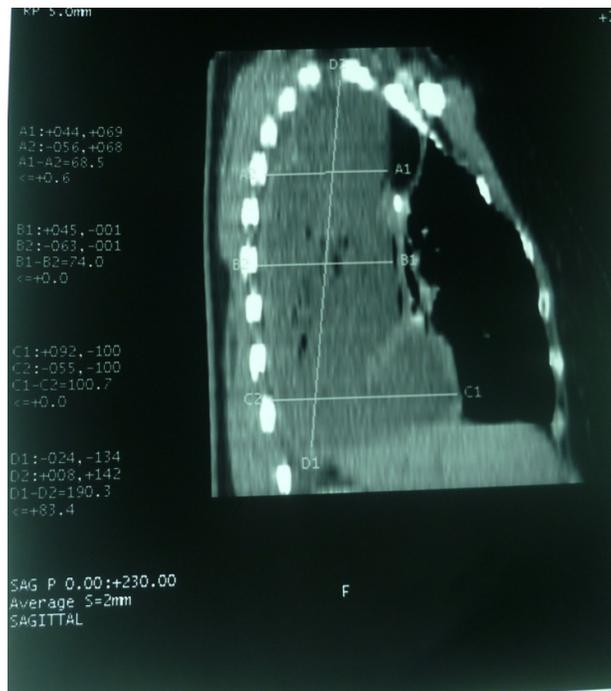


Figure 2. CT-Scan of the chest with dilated esophagus with the diameter at different levels

Discussion

Achalasia cardia is a primary oesophageal motility disorder characterized by failure of relaxation of the lower esophageal sphincter (LES) during swallowing and aperistalsis of the oesophageal body. Its features were first described by Sir Thomas William's in 1674 who also suggested the use of fish whalebone for dilatation but the term achalasia was first introduced by Arthur Hurst in 1927³.

The aetiology is unknown but autoimmune, viral and neurodegenerative processes are thought to be responsible with resultant damage to the myenteric plexus, paucity of inhibitory neurons and selective stimulation at the plexus. This leads to unopposed cholinergic stimulation and contraction of the lower oesophageal sphincter and consequent hypertension at the LES. The normal diameter of the Esophagus is 2.5-3cm.

The clinical features of achalasia include presence of prolonged intermittent dysphagia to liquids and solid, odynophagia and regurgitation. Weight loss may be a feature especially in prolonged cases. Oesophageal manometry is the gold standard for diagnosis in achalasia¹ but it is not widely available in our setting and plain chest X-ray, barium swallow and upper gastrointestinal endoscopy are the most commonly used diagnostic modalities which our patient did.

Achalasia is classified into four based on the severity and oesophageal dilatation.⁴ In minimal achalasia there is retention of food but no dilatation while mild achalasia is associated with vigorous disorganized motor activity and a dilated diameter of less than 4cm. Moderate Achalasia has a uniform dilatation of 4-6cm and severe achalasia has an esophageal diameter of more than 6cm.

End-stage Achalasia is the advanced form of achalasia where the oesophagus is dilated (diameter >6cm), tortuous and assume a sigmoid configuration. The oesophagus is termed megaesophagus. This is similar to the finding in our patient who had a dilated oesophagus with average diameter of 8.09cm. The dependent part within the chest allow for retention and stasis of food. The associated regurgitation results in recurrent chest infections and lung abscesses while the prolonged inflammation can lead to oesophageal squamous cell cancer.⁵The diagnosis may be difficult because of these associations.

Few misdiagnoses were reported in the literature. Kapadia et al⁴ reported a case of end-stage achalasia clinically misdiagnosed as pulmonary Tuberculosis which was resolved on chest x-ray evaluation. Khan et al⁶ also reported a patient who presented with acute airway obstruction with unclear diagnosis which was also resolved with chest x-ray evaluation and immediate improvement noted with rapid oesophageal decompression. Our literature search did not yield a prior report of end-stage Achalasia masked by chest trauma leading to its misdiagnosis similar to our patient but the diagnosis of the end-stage achalasia was resolved by the radiological evaluation done as in the previous reports although chest CT-scan gave the first hint instead of chest x-ray as in earlier reported misdiagnoses.

The Chest X-ray features of our patient were thought to be the sequelae of the chest trauma. This could be interpreted as a pneumothorax with some lung collapse and inserting a chest tube would have lead to oesophageal perforation and mediastinitis with the attendant worsened morbidity and mortality. The misdiagnosis of a right chest tumour was also entertained subsequently and the requested CT-Scan was the modality that gave the first hint to an oesophageal disease.

Thus it is pertinent for physicians to thoroughly evaluate patients with chest trauma and have an index of suspicion for other concomitant conditions. This will prevent complications and offer the appropriate treatment to the patient.

The treatment for end-stage achalasia is surgical. Laparoscopic Heller’s cardiomyotomy with antireflux procedure has been proposed by some surgeons as the initial treatment option with good outcome reserving oesophagectomy for failed cardiomyotomy ⁷. Feccani and co-workers ⁸ described a laparoscopic Heller’s cardiomyotomy, Dor fundoplication and a pull down procedure where 1-2 intramuscular U-stitches are applied on the right side of the lower oesophagus to restore the verticality of the oesophagus (Figure 3 a-d).

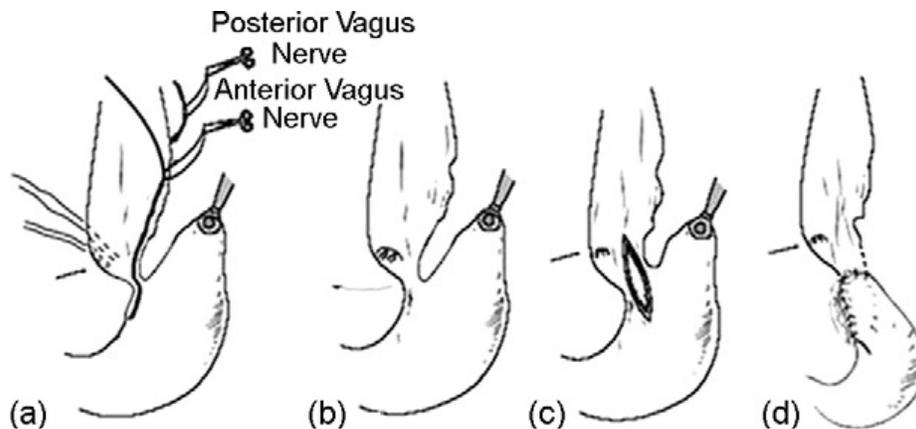


Figure 3. Heller’s cardiomyotomy, Dor fundoplication and a pull down procedure. Adapted from Feccani and co-workers

Oesophagectomy is the commonest treatment for end-stage achalasia and Transhiatal esophagectomy with cervical oesophagostomy is the preferred option. Devaney et al⁹ reported good outcome of patients who underwent oesophagectomy for end-stage Achalasia with 94% done through the Transhiatal approach. They stated their patient selection criteria for those to undergo esophagectomy. Cardiomyotomy is chosen for those with oesophageal diameter of >6cm when the oesophageal axis is straight. On the other hand, Oesophagectomy is done once the oesophagus becomes tortuous and sigmoid which renders it functionless. Colon interpositioning is another option especially if the stomach is not suitable as a graft¹⁰.

The index patient underwent successful transhiatal oesophagectomy and cervical oesophagostomy with good outcome.

Conclusion

End-stage achalasia occurs in neglected prolonged achalasia and its diagnosis may be shrouded by its complications or other clinical conditions such as chest trauma and the misdiagnosis may lead to wrong treatment with fatal outcome. Thus Physicians should consider the possibility of concomitant clinical conditions such as end-stage achalasia in evaluating chest trauma patients.

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