

# CASE REPORT

## ROLE OF IMAGING IN THE DIAGNOSIS OF ACHALASIA CARDIA

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### ABSTRACT

**Background:** Achalasia of the cardia is a condition of oesophageal motility disorder that starts at the oesophago-gastric junction and may spread upwards to involve portions of the mid-oesophagus. It is characterized by inability of the cardiac sphincter to relax fully in response to swallowing, hypertrophy and dilatation of the rest of the oesophagus, and absence or diminution of oesophageal peristalsis. It is commonly seen between the 2nd and 5th decades of life. The case is presented because of its rarity in our environment and its classical presentation

**Methods:** We reviewed the case of a 36 year old man who presented to the clinic with a 4 year history of progressive chest pain, difficulty in swallowing, difficulty in breathing, and weight loss.

**Results:** Plain radiography showed features that are suggestive of achalasia of the cardia and was confirmed on Barium swallow which showed grossly dilated mid and lower oesophagus with barium seen collected at the lower oesophagus with smooth narrowed tapering distally and absent peristaltic movement noted.computed tomographic features were in keeping with achalasia cardia.

**Keywords:** Imaging, diagnosis, achalasia cardia

### INTRODUCTION

Achalasia is an oesophageal motility disorder involving the smooth muscle layer of the oesophagus and the lower oesophageal sphincter (LES).<sup>1</sup> It is characterized by incomplete LES relaxation, increased LES tone, and lack of peristalsis of the oesophagus in the absence of other explanations like cancer or fibrosis.<sup>2</sup>

Achalasia, is in two forms, the primary type if the pathology is due to abnormality of oesophageal Aurbach plexus, which is the commonest and secondary type or pseudoachalasia which is seen in small proportions. The secondary is seen in oesophageal cancer, chagas disease, diabetes mellitus and scleroderma.<sup>3</sup>

The prevalence of the disease is 1 per 100,000 of the populace with no racial or sex predilection.<sup>3,4</sup> The peak incidence of the disease is between 20-40, but can be diagnosed in all age group.<sup>4</sup>

The radiological evaluation of a patient with achalasia include: Plain chest radiographs, barium oesophagogram and possibly trans-abdominal ultrasound.

This case is presented because of its rarity and its classical presentation.

### CASE REPORT

S. A. is a 36year old man that was admitted in Jos University Teaching Hospital (JUTH) with a 4 year history of progressive chest pain, difficulty in breathing, difficulty in swallowing, and weight loss. Difficulty in swallowing started with liquid and later to solid food intake. No history of fever or cough, but had upper abdomen burning sensation that was relieved by taking antacid. The patient does not smoke but drinks alcohol. No previous history of surgery.

Physical examination revealed a chronically ill-looking (wasted) young man not in any obvious respiratory distress. He was afebrile to touch, not pale, acyanosed, nil peripheral lymphadenopathy. Chest, central nervous system, cardiovascular and abdominal examinations were essentially normal.

A provisional diagnosis of Dysphagia ?Achalasia cardia with peptic ulceration disease was made.

The blood parameters, urea and electrolyte and were all within normal limits.

Plain chest radiograph (postero-anterior) showed a widened superior mediastinum with soft tissue mass extending from the root of the neck through the right thorax to the abdomen. Mottled opacity was seen in most part of the mass. The ipsilateral cardiac margin was not obscured. The lung fields were clear.

Barium swallow under fluoroscopy guidance showed grossly dilated oesophagus with barium seen collected at the lower oesophagus with smooth narrowed tapering distally. No peristaltic movement seen.

Computed tomography of the chest on mediastinal window(Fig 3) showed grossly dilated oesophagus with air-fluid levels. Coronal reformatted computed tomography(Fig 4) showed grossly dilated oesophagus filled with food particles. Multiple hypodense areas are seen within the dilated portion of the oesophagus. There is uniform thickening of the wall of the oesophagus.

Abdominal ultrasound was essentially normal.

Upper GI endoscopy with analysis of biopsy specimens from multiple sites showed areas of inflammatory mucosa.

The patient had heller's operation and Dor's fundoplication. He did well and was subsequently discharged home after 12 days post surgery.



Fig 2: Barium swallow examination showing a dilated contrast filled oesophagus with a narrowed lower end.



Fig 3: Axial computed tomography of the chest on mediastinal window showing grossly dilated oesophagus with air-food particles levels.



Fig 1: Plain chest radiograph (frontal projection) showing widened mediastinum with mass lesion in the right hemithorax with mottled opacities.



Fig 4: Coronal reformatted computed tomography of the chest on mediastinal window showing grossly dilated oesophagus that has soft tissue dense mass with multiple interspersed hypodensities.

## DISCUSSION

The exact aetiology of primary achalasia cardia is not known, though the most widely accepted theories implicate auto-immune disorder. Infectious diseases or both. In all cases of achalasia, the most consistent finding is the degeneration of myenteric plexus of Aurbach in the oesophagus.<sup>5</sup> This degeneration results in impaired oesophageal emptying with resistance to ante grade flow. As a result of this, the oesophagus becomes dilated and elongated.<sup>5</sup>

The clinical symptoms and signs of patients with achalasia cardia depend on the severity of the lesion and the associated complications. But the most common presenting symptoms and signs include: dysphagia, regurgitation, weight loss, chest pain and cough. The index case presented with retrosternal chest pain, difficulty in breathing and swallowing. The retrosternal pain was likely due to the peptic ulcer. However, the difficulty in breathing and swallowing may be due to the dilated oesophagus which was evidenced from the plain chest radiographs and barium swallow.

The radiological evaluation of a patient with achalasia include: Plain chest radiographs, barium oesophagogram and possibly trans-abdominal ultrasound. Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) have no useful role in the management of primary achalasia.<sup>8-12</sup>

Plain chest x-ray may be normal or in most cases show widened mediasternum with air fluid levels. Features of chest infection may be seen in the lung fields. The fundal gas may be absent or partially reduced.<sup>9,12</sup> The plain chest radiographs of this patient showed features of widened mediasternum with a mass that has mottled lucencies.

Barium oesophagogram in a patient with achalasia will show a decrease or absent primary peristalsis, dilated thoracic oesophagus with a tapered distal part.<sup>7</sup> But it must be stated that a normal finding on barium swallow examination does not completely exclude achalasia cardia in its early stage.<sup>7</sup> This patient's barium study was able to demonstrate a grossly enlarged thoracic oesophagus with tapering at its distal end and absent peristaltic movement.

Ultrasonography is not recommended for the primary diagnosis of Achalasia. Though, some workers are of the opinion that this method may be useful as a screening test to discern achalasia from

pseudoachalasia.<sup>12</sup> This patient had a trans-abdominal ultrasonography which was normal.

Computed tomography can be used in differentiating achalasia from pseudoachalasia in a patient with oesophageal dilatation. The wall thickness said to be uniform in achalasia as against pseudoachalasia that is uneven.<sup>11</sup> The wall of the oesophagus appears uniformly thickened (Figs 3 and 4) as seen in achalasia cardia. It may show mass lesions in oesophagus, which was not seen in this case.<sup>11</sup>

A diagnosis of achalasia supported by the results of radiological studies must always be confirmed by performing upper gastrointestinal endoscopy and oesophageal manometry. These tests allow the direct evaluation and inspection of the oesophageal mucosa and objective measurement of oesophageal contractility.<sup>13</sup> Endoscopy supplemented by biopsy when necessary, helps in excluding gastrointestinal malignancies, fungal or bacterial infections and other disease processes that can mimic achalasia.<sup>5,13</sup> However, endoscopy and biopsy results showed associated complications i.e. epithelia metaplasia and oesophagitis in this patient.

Manometric study was not done on this patient because of lack of facility in our centre.

The management of achalasia could either be medical or surgical depending on the stage of the disease and the age of the patient.<sup>14,15</sup> This patient had a successful surgery after his clinical condition was stabilized and was discharged home 12 days after.

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