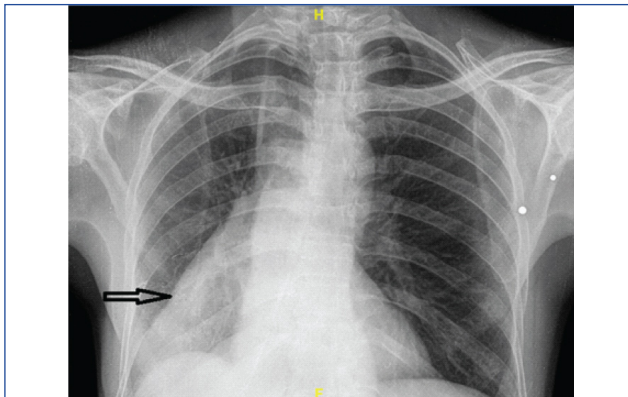


Sigmoid Oesophagus as an Unusual Cause for Cough in a 40-Year-Old Man: Interesting Radiology

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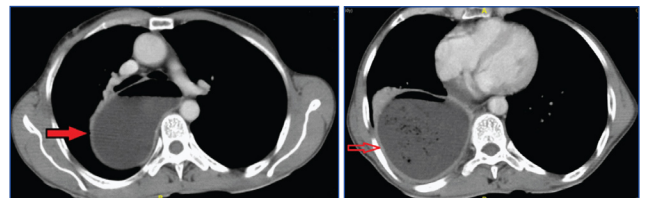
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A 40-year-old agricultural labourer was referred to Pulmonary Medicine Department with complaints of cough with minimal expectoration and right sided chest pain for past two months. He also had symptoms of recurrent belching and heartburn. No relevant history of trauma, smoking, alcohol consumption or tuberculosis was reported by the patient. On physical examination, he was stable and his vital parameters were normal. Respiratory examination revealed few crepitations in the right interscapular region. Chest X-ray showed a homogenous opacity in the right mid zone and lower zone with no air bronchogram [Table/Fig-1]. A possibility of right



[Table/Fig-1]: Chest X-ray PA view showing a homogenous opacity in the right middle and lower zone not silhouetting the heart or diaphragm (arrow).

lower lobe collapse was considered and a Computed Tomography (CT) of the chest was planned. CT image revealed normal airways and lung parenchyma. However, the oesophagus was grossly dilated and a portion of it was seen to lie on the right side of chest [Table/Fig-2,3]. There was no diaphragmatic defect. Based on the CT image, a possibility of sigmoid oesophagus was considered and patient was referred to Surgical Gastroenterology Department. The final diagnosis was achalasia cardia-sigmoid oesophagus. After obtaining consent, he underwent a thoraco-laparoscopic oesophagectomy with gastric pull up and cervical oesophago



[Table/Fig-2]: CT-scan of chest at the level of main bronchi division showing a dilated oesophagus that is to the right of midline with fluid level (red arrow).

[Table/Fig-3]: CT-scan of thorax showing a well dilated oesophagus with axis rotation to the right (red arrow).

gastric anastomosis. He was symptomatically better till the last contact.

Achalasia cardia is a rare degenerative motility disorder of the oesophagus due to failure of the Lower Esophageal Sphincter (LES) to relax in response to food bolus with aperistalsis of the body of the oesophagus leading to dysphagia and progressive dilatation of the oesophagus. Sigmoid oesophagus represents end stage of achalasia cardia. The dilated oesophagus undergoes axis deviation resulting in a dilated and tortuous appearance [1].

The incidence of achalasia is about 0.5-1.2/100,000 per year and a prevalence of 8-10 per 100,000. Though, common in advancing age, it can occur in any age and in both sex [2].

Achalasia can be primary (idiopathic) or secondary. Primary achalasia has been associated with viral infections like measles, varicella and herpes simplex infections. A genetic predisposition and immunological causes are also suspected in the pathogenesis. Secondary achalasia also known as pseudoachalasia occurs in Chagas disease, autoimmune condition like scleroderma, previous oesophageal or vagal surgeries and gastric malignancy [2,3].

The pathology of achalasia is at the LES. In the initial stages there is degeneration of inhibitory neurotransmitters like nitric oxide and vasoactive intestinal peptide with unopposed action of excitatory neurotransmitters like acetylcholine and

substance P at the level of LES. This imbalance leads to high amplitude non-peristaltic contractions. This initial phase is known as vigorous achalasia. With further progression, there is dilation and low amplitude simultaneous contraction in the body of the oesophagus. This phase is known as the classical achalasia. In the end stage achalasia, the oesophagus is grossly dilated with axis deviation leading to a dilated and tortuous appearance known as sigmoid oesophagus [4].

Patients usually complain of dysphagia for both liquid and solid diet. Regurgitation of undigested food also occurs and may be mistaken for Gastroesophageal Reflux Disease (GERD). Regurgitation may occur even after hours after food intake particularly when patient assumes a recumbent position [5].

Confirmatory diagnosis is by manometric, endoscopic and radiographic investigations. Oesophageal manometry is widely considered as the gold standard in the diagnosis of achalasia. On manometry, there is aperistalsis and failure of relaxation of the LES (it should be mentioned as aperistalsis instead of a peristalsis [5]). Endoscopy is advised to rule out malignancy. Barium swallow may reveal the pathognomonic "bird's beak" appearance of the distal oesophagus with dilatation of the oesophagus proximally. A normal barium swallow however does not completely rule out achalasia. Radiographically, sigmoid oesophagus is seen as dilatation of the distal oesophagus of more than 10 cm with axis deviation. Sigmoid oesophagus can also be seen on CT as dilated tortuous oesophagus with single lumen (sigmoid Type 1) or with two lumen (sigmoid Type 2) [5].

Achalasia is a chronic condition without a significant cure. Current treatment modalities are directed at reducing the

hypertonicity of the LES by pharmacologic, endoscopic, or surgical means. However, no treatment significantly affects oesophageal peristalsis, and hypertonicity of LES recurs needing repeated interventions. The mainstay of treatment for achalasia is either pneumatic balloon dilatation or laparoscopic myotomy. A newer endoscopic oesophagomyotomy technique, peroral endoscopic myotomy has been suggested. For patients not fit or deny invasive procedures alternative treatment options include long acting nitrates and calcium channel blockers. These drugs are given sublingually due to delayed oesophageal emptying in achalasia. Botulinium injection into the LES is also performed. Oesophagectomy is advised for patients with recurrence despite pneumatic dilatation and myotomy [3-5].

Conditions like achalasia cardia have to be considered in patients with recurrent cough and regurgitation. A clinic radiological disagreement should raise suspicious of extra pulmonary cause and should be evaluated thoroughly.

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