Case Report

Achalasia Cardia Masquadring as Mediastinal Tuberculosis in a 12-year-old Girl

Kamal Nain Rattan[1], Jasbir Singh[2] and Poonam Dalal[2]

Departments of Pediatric Surgery[1] and Pediatrics[2], Pt. B.D. Sharma Postgraduate Institute of Medical Sciences, Rohtak (Haryana), India

Abstract

Achalasia is a rare neurodegenerative disorder with deranged oesophageal peristalsis and lower oesophageal sphincter function. We report a case of achalasia cardia in 12-year-old child, who presented with respiratory tract infections. This patient was misdiagnosed as a case of mediastinal tuberculosis and anti-tuberculosis drugs were given for three months. However, she was diagnosed having achalasia cardia later with barium contrast studies and managed successfully with surgical intervention. **[Indian J Chest Dis Allied Sci 2018;60:39-41]**

Key words: Achalasia, Dysphagia, Respiratory disease, Tuberculosis.

Introduction

Achalasia was described first almost 300 years ago by Thomas Willis in a patient who required oesophageal dilation with the whalebone.[1] The term achalasia means "failure to relax" and it is associated with impaired relaxation of lower oesophageal sphincter (LES) along with the loss of peristalsis in body of the oesophagus. Typical presenting features include gradual progressive dysphagia, regurgitation of undigested food with pooled up saliva and chest pain.[2] Esophagogram is the preferred initial screening investigation but high-resolution manometry remains the gold standard tool for establishing the diagnosis.[3,4] Various pharmacological agents, like calcium channel blockers, nitrates and sildenafil are being used in its management with variable success. Conservative treatment with pneumatic balloon dilatation of LES and surgery (Heller's myotomy) are the other available interventions.

Case Report

A 12-year-old, female child (weight: 13Kg) presented with cough since five months, regurgitation of food with saliva and difficulty in swallowing food for the last two months. Mild fever was present at the start of illness which got settled in the preceding two months. The patient had weight loss during last four months. She was treated at a peripheral hospital with conventional antibiotics for respiratory tract infections but did not respond. The grandfather of the patient had received anti-tuberculosis treatment (ATT) for sputum-positive pulmonary TB one year ago. Chest radiograph showed bilateral hilar prominence associated with opacities in the right lung field (Figure 1). Mantoux test was reactive (14 mm at 72 hours). Investigations revealed: haemoglobin 8.2 g/dL, erythrocyte sedimentation rate (ESR) 45 mm in first hour. Liver function test and C-reactive protein (CRP) levels were within normal limits. Based on history and basic investigations, a presumptive diagnosis of mediastinal lymph node TB was made and ATT was started at the peripheral hospital. But despite good treatment compliance, the patient did not improve. The patient was referred to our center.

Clinical examination revealed no abnormality on chest auscultation; the cardiovascular, abdominal and nervous system examinations were normal. Repeat chest radiograph showed hilar prominence

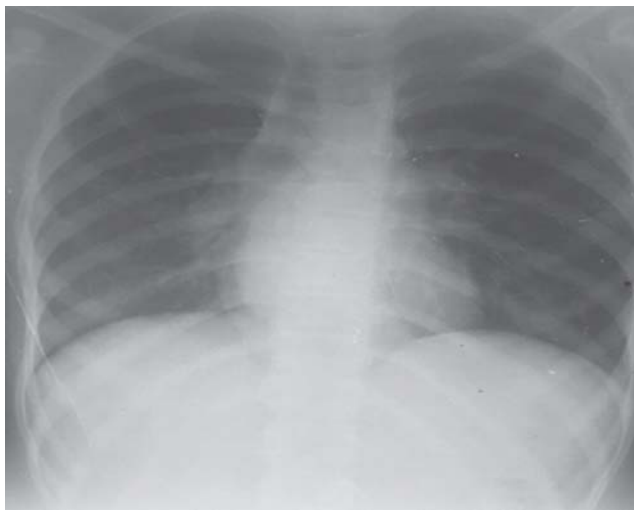

Figure 1. Chest radiograph (postero-anterior view) showing bilateral hilar prominence and infiltrates in the right lung field.

[Received: March 20, 2017; accepted after revision: December 6, 2017]

Correspondence and reprint requests: Dr Jasbir Singh, Senior Resident, Department of Pediatrics, Pt. B.D. Sharma Postgraduate Institute of Medical Sciences, Rohtak-124 001 (Haryana), India; E-mail: jasbir2001@gmail.com

along with mediastinal widening. Gastric aspirate was non-reactive for acid-fast bacilli (AFB). Due to progressive nature of dysphagia, alternative diagnosis of obstructive upper gastrointestinal lesion was considered and the patient was investigated further.

An esophagogram revealed a typical bird's beak appearance (Figure 2), which confirmed the diagnosis of achalasia cardia. The diagnosis was confirmed on computed tomography of the chest that showed grossly dilated oesophagus with fluid levels and compression of the adjacent airways (Figure 3). Upper gastrointestinal endoscopy revealed massively dilated oesophagus with residual food particles and secretions. ATT was stopped immediately and surgery was planned. Heller's myotomy under general anaesthesia was performed through the left transpleural thoracotomy along with intercoastal tube drainage. Post-operative period was uneventful and the patient was started on oral feed on 6th post-operative day and discharged on 10th day in stable condition.

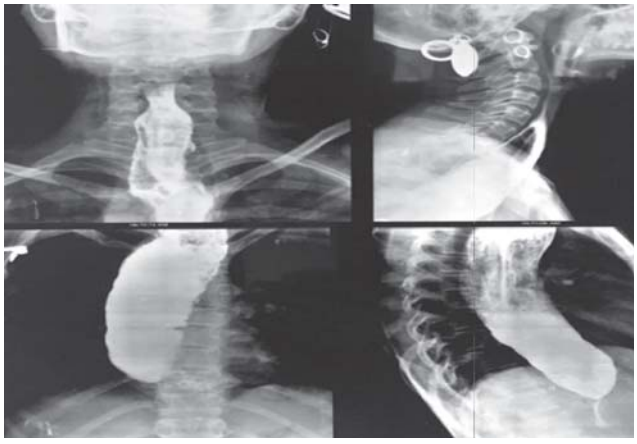

Figure 2. Barium contrast studies showing air fluid levels in dilated proximal oesophagus and narrowed distal part as typical "Bird's beak appearance".

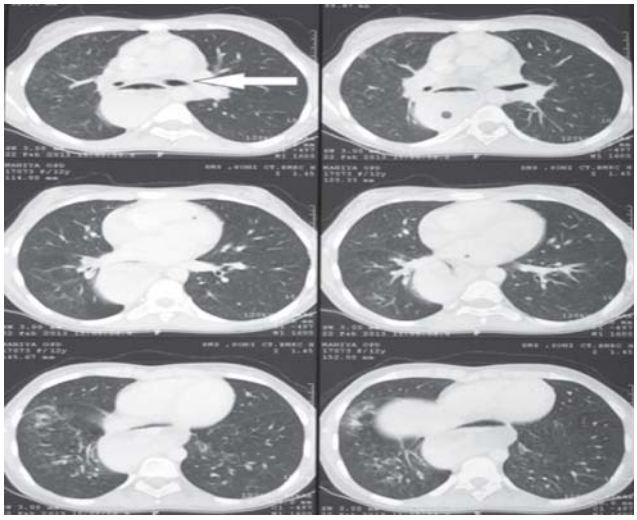

Figure 3. Computed tomography of the chest showing dilated oesophagus compressing surrounding airways (arrow).

Discussion

Achalasia cardia is a primary oesophageal motor disorder in which there is loss of peristalsis in the body of oesophagus and lack of coordinated LES relaxation while swallowing. The reported incidence is 0.11 cases per 100,000 children with the peak between 30-60 years and with no gender preference.[5] Although the precise aetiopathogenesis remains still unknown; available research suggests that hereditary, degenerative, autoimmune and infectious factors may be the possible causes.[3]

The classic presentation of achalasia consists of dysphagia to both solids and liquids in association with regurgitation of undigested food, chest pain, and belching. These symptoms can lead to confusion with reflux disease, but regurgitation in achalasia does not respond to proton pump inhibitors. Recurrent chest pain is also seen as presenting feature in about 17% to 95% of patients. Other less frequently encountered symptoms are cough, hoarseness, wheezing, shortness of breath and sore throat. Achalasia may have atypical presentations sometimes, and hence, can pose a diagnostic dilemma. It can be misdiagnosed with other prevalent diseases, like tuberculosis as in the index case. Mehdi *et al*[6] had reported two cases of achalasia which presented as chronic nocturnal cough and were treated for a long time as respiratory diseases before being diagnosed correctly.[6] Dabritz *et al*[7] had also found two cases of achalasia, where dysphagia, chest pain, and regurgitation were not predominant symptoms and patients were being treated as eating disorder.[7] Wani *et al*[8] had reported a achalasia patient who presented with massive haemoptysis and was initially treated as pulmonary tuberculosis but diagnosed as achalasia associated with *Actinomycosis* later on.[8]

In the present case also, there was history of recurrent cough and fever, so she was treated for chest infection initially.

Diagnosis of achalasia can be made based on the findings of physical examination, clinical presentation, and radiological investigations. Oesophageal manometry is the gold standard to make diagnosis irrespective of findings of barium studies. A peristalsis in the body of oesophagus and maintaining a high tone of LES during swallowing are the typical findings on manometry. High resolution manometry is preferred than the conventional, as it can portray the motor patterns with better treatment outcomes.[5] Timed barium oesophagogram is a simple, non-invasive and widely available contrast technique for evaluating oesophageal emptying in patients with achalasia. In advanced stages of the disease, there is the presence of smooth tapering of the lower oesophagus leading

to a closed LES, resembling a bird's beak. Other local lesions can mimick as achalasia, e.g., tumours of the oesophageal-gastric junction, lymph node compression, and pseudoachalasia. Upper gastrointestinal tract endoscopy is helpful in differentiating the above lesions.[9]

Achalasia can be treated with pharmacologic agents, pneumatic dilation and surgical myotomy with variable success rates.[10] The choice of initial therapy should be guided by patient's age, gender, preference, and local institutional expertise. In pharmacologic therapy calcium channel blockers, nitrates and sildenafil are commonly used. The short duration of relief and associated side effects are the main limiting factors in their use. Endoscopic injection of botulinum toxin is also used with limited success. Main disadvantages of botulinum injections are the high relapse rate and requirement of repeated injections. Pneumatic dilation of LES is also used with good success rate in experienced hands. The most dangerous complication is the oesophageal perforation necessitating immediate surgical intervention. Surgical (Heller) myotomy consisting of division of muscular fibers of LES without damaging mucosa has a high degree of safety and minimal recurrence rate.[2]

In conclusion, achalasia cardia is an uncommon disorder to be seen in children and usually presents with progressive dysphagia, retrosternal pain, vomiting, regurgitation of undigested food and weight loss. However, sometimes clinical presentation may be non-specific; mimicking commonly prevalent chest diseases. Therefore, a high index of suspicion should be kept in favour of achalasia in children presenting with progressive dysphagia and recurrent vomiting.

References

1. Willis T. The combination of medication and surgery of esophageal achalasia. In: Willis T, editor *Achalasia*. London: Has-comitis;1674:18–9.
2. Richter JE. Achalasia: an update. *J Neurogastroenterol Motil* 2010;16:232–42.
3. Patel DA, Kim HP, Zifodya JS, Vaezi MF. Idiopathic (primary) achalasia: a review. *Orphanet J Rare Dis* 2015;10:89.
4. Nabi Z, Reddy DN. Achalasia cardia: recent advances in diagnosis and endoscopic management. *J Gastrointest Dig Syst* 2016,6:1.
5. Zhang Y, Xu CD, Zaouche A, Cai W. Diagnosis and management of esophageal achalasia in children: analysis of 13 cases. *World J Pediatr* 2009;5:56–9.
6. Mehdi NF, Weinberger MM, N Abu-Hasan MN. Achalasia: unusual cause of chronic cough in children. *Cough* 2008;4:6.
7. Dabritz J, Domagk D, Monninger M, Foell D. Achalasia mistaken as eating disorders: report of two children and review of the literature. *Eur J Gastroenterol Hepatol* 2010;22:775–8.
8. Wani AW, Hussain WM, Banjar AA, Al Miamini WH, Khoujah AM, Bafaraj MG, *et al*. Haemoptysis in a patient of achalasia cardia: pulmonary actinomycosis, not tuberculosis. *BMJ Case Rep* 2010;bcr09.2009.2287.
9. Kahrilas PJ, Kishk SM, Helm JF, Dodds WJ, Harig JM, Hogan WJ. Comparison of pseudoachalasia and achalasia. *Am J Med* 1987;82:439–46.
10. Chuah SK, Chiu CH, Tai WC, Lee JH, Lu HI, Changchien CS, *et al*. Current status in the treatment options for esophageal achalasia. *World J Gastroenterol* 2013;19:5421–9.