

## Case Report

# Achalasia cardia presenting with bilateral broncheactasis in a child

Bhavana Venkata Nagabhushana Rao<sup>1\*</sup>, Venkata Satya Raman Bhavana<sup>2</sup>

<sup>1</sup>Department of Medicine, Queens NRI Hospital, Visakhapatnam, India

<sup>2</sup>Department of Surgery, Queens NRI Hospital, Visakhapatnam, India

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### \*Correspondence:

Dr. Bhavana Venkata Nagabhushana Rao,  
E-mail: bhavanavnrao@gmail.com

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### ABSTRACT

Achalasia cardia usually presents in the age group of 25 to 60 and rare in children. Dysphagia, regurgitation, vomiting and chest pain are the cardinal symptoms. If they present mainly with respiratory symptoms diagnosis may be difficult and delayed. Bilateral bronchiectasis is rarely reported in patients with achalasia both in adults and children. Sometimes children may be erroneously diagnosed as eating disorder as both may present with similar symptoms hence information that achalasia can occur in children reduces such risk. In those children presenting with chronic respiratory symptoms it is prudent to look for esophageal motility disorders. CT scan, endoscopy, manometry are useful diagnostic tools but gold standard is barium esophgogram to diagnose Achalasia. Chest postural drainage may be delayed in bronchiectatic patients with achalasia. Heller myotomy is standard surgical treatment for children. Untreated Patients may end up with megaesophagus, a progressively dilated esophagus.

**Keywords:** Achalasia, Respiratory disease, Bilateral bronchiectasis, Chronic cough, Child

## INTRODUCTION

Achalasia is a primary motor disorder of the esophagus characterized by insufficient lower esophageal sphincter relaxation and loss of esophageal peristalsis. This results in patients' complaints of dysphagia to solids and liquids, regurgitation, and occasional chest pain with or without weight loss. Endoscopic finding of retained saliva with puckered gastroesophageal junction or barium swallow showing dilated esophagus with birds' beaking in a symptomatic patient should prompt appropriate diagnostic and therapeutic strategies.<sup>1</sup>

Bilateral bronchiectasis in achalasia as presenting feature not reported in literature many a times.<sup>2</sup>

Here we present a case of 13 year boy presenting with bilateral bronchiectasis and achalasia cardia.

## CASE REPORT

Thirteen year old boy was brought to our hospital with history of recurrent cold, cough, fever and purulent sputum of two year duration. Cough and fever were getting controlled with a course of antibiotic and recurring again. Patient has been vomiting the food after repeated coughing. Refusing to eat adequate amounts in fear of vomiting and regurgitation. He also gets chest pain anteriorly and increased by coughing. His mother says he takes long time to eat a little food. He gets disturbed sleep as violent bouts of cough wakes him from sleep. Frequent belching in the classroom annoying his classmates and teachers as well. Many a times he received antibiotic, cough medicines and proton pump inhibitors.

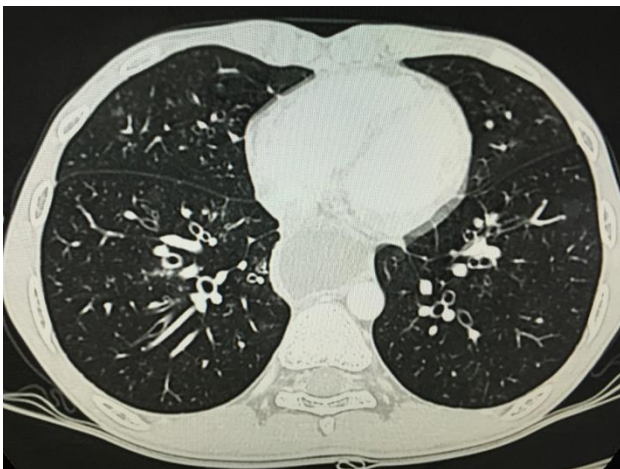
Six months ago he consulted a pulmonologist who did chest X-ray and mantoux test. Chest x ray was normal and mantoux was positive 12 mm. In the view of chronic

cough, fever and a positive mantoux test he started him on trial of anti tuberculous drugs. In spite of antibiotics and anti tubercular treatment his problem persisted. The boy was losing weight, uninterested to play with peers and parents very much worried that he has to abstain from the school frequently due to ill health.

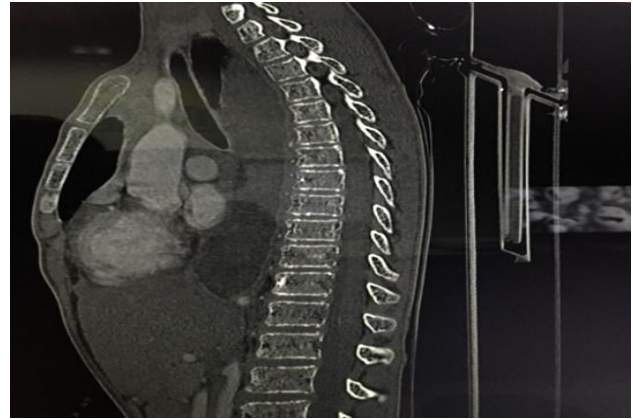
Thin built, under nourished boy, has been coughing frequently and has early clubbing of fingers. Auscultation of lungs revealed bilateral rhonchi, and coarse crackles at the base and mammary regions. TWBC: 13,400, ESR: 40, X ray PNS: Bilateral maxillary sinusitis, Chest X ray: Normal

We put him on Esomeprazole plus levosulpiride to be taken on empty stomach, salmeterol + fluticasone inhaler, Azelastine + fluticasone nasal spray at night and Cefuroxime 250 mg twice a day. He symptomatically improved for a week and returned again with cough, fever, large quantities of sputum. In the view of recurrent sinobronchial infections, purulent large quantities of sputum, varying cough and sputum with posture, coarse crackles on auscultation we asked for CECT Chest considering bronchiectasis a possibility. CT scan showed bilateral lower lobe, right middle lobe bronchiectasis (Figure 1). In addition to pulmonary pathology oesophagus was moderately dilated with no mural abnormality suggestive of Achalasia (Figure 2). We did a barium esophagogram to confirm the diagnosis which showed dilated, non peristaltic oesophagus with bird beak appearance of lower end of oesophagus. We did upper GI Endoscopy; oesophagus was dilated (Figure 3) withholding a lot of food debris and lower oesophageal sphincter was tight needing extra pressure to negotiate.

On further inquiry patient agreed that he has been suffering from dysphagia for solids and liquids and spontaneous chest pains were common.



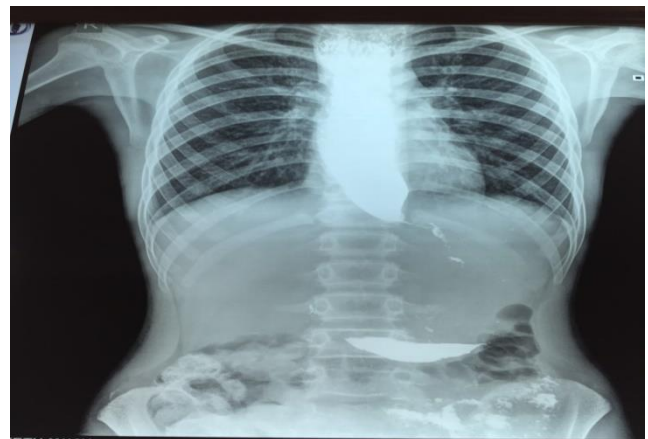
**Figure 1: CECT CHEST showing bilateral bronchiectasis.**



**Figure 2: CECT CHEST markedly dilated oesophagus without mural abnormality.**



**Figure 3: Upper GI endoscopy demonstrating dilated oesophagus.**



**Figure 4: Post dilatation barium oesophagogram demonstrating contrast entering stomach.**

## DISCUSSION

Achalasia is a rare disorder with prevalence of 10 cases per 100,000 population.<sup>3</sup> Men and women are equally affected, onset before adolescence is rare. Achalasia is insidious in onset and slow to progress. Mean duration of symptoms before diagnosis averages around 4.7 years.

The delay in diagnosis was due to failure to recognize typical symptoms rather than presenting with atypical features.<sup>4</sup> Dysphagia for solids (90%), liquids (85%), regurgitation of food or saliva (76-91), aspiration (8%) and chest pain are the common symptoms.<sup>5</sup>

Our patient presented with recurrent cough, fever, vomiting, and hiccups. Dysphagia though present relegated to the least by the patient and family as the respiratory symptoms dominated. Children suffering from hyper reactive airway disease may have recurrent exacerbation due micro aspiration. Children with achalasia suffering from recurrent vomiting and regurgitation may be mistaken for eating disorders, and at times both can coexist.<sup>6</sup> Achalasia patients may eat slow or adopt abnormal posturing to facilitate deglutition. Our patient adapted to eat slowly.

We did CT scan of chest to investigate for bronchiectasis found esophageal achalasia. CT scan is very useful investigation not only to diagnose primary achalasia but also to exclude secondary achalasia due to malignancy.<sup>7</sup> On manometric study a peristalsis of distal two third of esophagus and incomplete relaxation of lower esophageal sphincter are suggestive of achalasia. High resolution manometry is much more sensitive tool.

Chest X ray was normal in our patient but may show dilated mediastinum due to dilated esophagus and absent fundal gas shadow. Barium esophagogram is further confirmatory and easily available investigation in developing countries. Upper GI endoscopy is further informative and a must for elderly to exclude malignancy.

Without treatment esophagus may progressively dilate and end up in megaesophagus, up to 5% of patients may require oesophagectomy. Patients with achalasia are at increased risk malignancy often of squamous cell, but absolute risk is low, in general routine endoscopic surveillance is not warranted.<sup>8</sup>

Treatment options include mechanical disruption of the muscle fibers of the LES (e.g. pneumatic dilation or surgical myotomy) or biochemical reduction in LES pressure (e.g. injection of botulinum toxin, oral nitrates, or calcium channel blockers). Botulinum injection and medical therapy are not preferred methods of treatment in children. Heller myotomy is preferred and often performed laparoscopically.<sup>9</sup> Since LES disruption can cause reflux esophagitis; it is frequently combined with an antireflux procedure such as a fundoplication. Peroral endoscopic myotomy (POEM) is an endoscopic method for performing a myotomy of the LES and is being studied for the treatment of achalasia, needs expertise.

Our patient had undergone pneumatic dilatation; he is able to swallow normally. Barium esophagogram taken

after first dilatation shows contrast entering stomach (Figure 4). Cough, recurrent fever and shortness of breath are also subsided.

## CONCLUSION

Recurrent respiratory infections, with symptoms of gastroesophageal reflux even without classical dysphagia we have to consider achalasia as a possibility and investigate. Bronchiectasis can occur though rare in achalasia cardia.

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