

# Journal Of Clinical Practice and Medical Case Report

Genesis-JCPMCR-1(1)-09  
Volume 1 | Issue 1  
Open Access

## A Case Report on Achalasia: A Comprehensive Overview

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**Citation:** Jain V. A Case Report on Achalasia: a Comprehensive Overview. J Clin Pract Med Case Rep. 1(1):1-4.

**Received:** April 29, 2024 | **Published:** May 13, 2024.

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### Abstract

The objective is to describe two uncommon cases of Achalasia, primarily esophageal motor disorder of unknown etiology. Pathologically inflammation surrounds the ganglionic cells. Two teen age children presented with complains of chronic cough, regurgitation after meals and significant weight loss. With the help of possible investigations like HRCT, Endoscopy & Barium swallow, diagnosis of Achalasia confirmed and treated appropriately through pneumatic dilatation of LES.

### Keywords

LES (lower esophageal sphincter); Auerbach plexus; Post ganglionic cholinergic neurons

### Introduction

The primary esophageal motor condition known as achalasia, which has an unclear etiology, is characterized by a lack of LES (lower esophageal sphincter) relaxation and esophageal peristalsis, which contributes to the functional blockage of the distal esophagus [1]. Achalasia is a rare condition in children. Even less prone to have this illness are kids under the age of five. An yearly incidence of 0.11/100000

children are affected by achalasia [2, 3]. Less than 5% of symptomatic patients are typically under the age of 15, on average. Males are more commonly affected by the illness, which is generally idiopathic. Trisomy 21, glucocorticoid insufficiency, eosinophilic esophagitis, familial dysautonomia, Chaga's disease, achalasia, alacrimia, and ACTH insensitivity (AAA) syndrome are only a few of the disorders that have been related to achalasia [4]. Trypanosoma cruzi infection and degenerative autoimmune response (antibodies to Auerbach plexus) are thought to be potential causes. The ganglion cells, whose number is reduced in this disease, are encircled by pathological inflammation. Postganglionic inhibitory neurons, which ordinarily cause sphincter relaxation, are selectively lost, leaving postganglionic cholinergic neurons unopposed. High basal LES pressure and proximal peristalsis loss are brought on by this imbalance (a secondary LES constriction phenomena) [4,5].

Regurgitation and dysphagia for foods and liquids, together with a persistent cough and slow weight loss, are the predominant clinical manifestations. In females, anorexia nervosa may be misdiagnosed as asthma, TB, or persistent cough with regurgitation/vomiting [6]. According to a barium research, the lower esophagus gently tapers down to a nearly closed LES, giving it a "bird beak" or "rat tail" look. In order to distinguish between the three different types of achalasia, manometry is useful [7, 8]. Through better esophageal emptying and mega-esophagus avoidance, therapy aims to relieve symptoms. Pneumatic (balloon) dilatation and surgical 'Heller's myotomy' are the two most efficient therapeutic techniques [9, 10]. We present two achalasia instances with typical and identical.

### Case-1

13-year-old Hindu male child product of non-consanguineous marriage admitted at our center with complaints of chest pain, difficulty in swallowing, cough (more at night) and marked weight loss for last 2 years. Pain in chest was dull, constant with heaviness on right side. He took treatment from multiple centers with no/negligible response. This all symptomology started with high grade fever of long duration (2 weeks) diagnosed as typhoid fever for which patient received full course of treatment. Later fever was never present. Family history, development history including birth history were not contributory.

On examination, the child looked undernourished and mild anemic, beside that there was no abnormality was detected even on repeated thorough examination. Systemic examination of all systems was also normal. Since history of weight loss was significant (23kg in 2 years from 55 to 32kg.) and chronic cough was there initially, differential diagnosis kept were Koch's chest, RAD (reactive airway disease) and esophagitis with GERD. He was investigated for that and all tests for Koch's chest and RAD were negative including CBNAAT, sputum & gastric aspirate for AFB, ESR, MT test, chest X-Ray & spirometry. Following this HRCT (THORAX), endoscopy and manometry were planned. HRCT (chest) clinched the diagnosis of Achalasia.



Patient 2

Child was then sent for Endoscopy and balloon dilation was done.



Patient 1

Child became asymptomatic from the next day and was allowed full diet. In next 15-16 days he gained 4 kg weight (became 37kg). He is now completely asymptomatic i.e., no cough dysphagia, chest pain or heaviness of left side chest.

## Case-2

12-year-old Muslim male child product of non-consanguineous marriage admitted in our center with complaints of cough, chest pain, and difficulty in swallowing and gradual but significant weight loss for last 2 years (documented loss of 13kg). Cough was more during sleep with regurgitation of meals.

The symptomatology started with long duration fever diagnosed as typhoid fever, for which he received full course of treatment (akin to case 1). Family history, birth history including developmental history was not contributory. On examination the child looked under nourished and mild anemic. Systemic examination of respiratory, cardiovascular or abdomen revealed no significant abnormality. Since history of significant weight loss for last 2 years and chronic cough was there, initially we thought Koch's chest or

RAD or gastritis with GERD. Now we had already diagnosed a case of ACHALASIA, so for this case we planned barium swallow as well.

Like first case, tests for tuberculosis, asthma were negative and barium swallow confirmed the diagnosis of achalasia, showing typical bird's beak appearance. (Picture 3& 4) Patient underwent esophageal pneumatic dilatation and on follow up found to have gained 3 kg in 2 weeks (32 to 35kg) with complete disappearance of symptoms i.e., difficulty in swallowing, cough, chest pain etc.

## Discussion

Both these boys showed striking similarities like same age group, product of non-consanguineous marriage, same gender, and symptomatology started after typhoid fever. Can it be a complication of enteric fever? No fixed pattern of inheritance? Male preponderance? Although rare disease in childhood, but one should keep in mind, while dealing with case of chronic cough and chest pain with significant wt. loss. Important to note fever was missing in both cases.

## Lesson learnt

- Achalasia is uncommon in pediatric population.
- Chronic cough with significant weight loss over long duration without fever in teenage can be achalasia.
- It is easily treatable without extensive surgery.

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