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Case report

# Achalasia with massive oesophageal dilation causing tracheomalacia and asthma symptoms

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A B S T R A C T

Achalasia is an uncommon oesophageal motor disorder characterized by failure of relaxation of the lower oesophageal sphincter and muscle hypertrophy, resulting in a loss of peristalsis and a dilated oesophagus. Gastrointestinal symptoms are invariably present in all cases of achalasia observed in adults. We report a case of a 34 year-old female patient with long standing history of asthma-like symptoms, labelled as uncontrolled and steroid resistant asthma with no gastrointestinal manifestations. Thoracic CT scan revealed a massive oesophagus due to achalasia, which caused severe tracheomalacia as a result of tracheal compression. Her symptoms regressed completely after a laparoscopic Heller myotomy surgery intervention.

## 1. Introduction

Tracheomalacia (TM) can be caused by weakness of the supporting cartilage of the trachea and main bronchi, or by dynamic closure from the posterior membranous wall. The aetiology of TM is varied with most cases being idiopathic and external tracheal compression is rare. This case illustrates an unusual presentation of late stage achalasia with mega-oesophagus causing tracheal compression and asthma-like symptoms with an insidious onset during many years. The case highlights the need for a diagnostic review when classical symptoms of dyspnoea, cough and wheeze do not respond to conventional asthma treatment.

## 2. Case report

We report the case of a 34 year-old female who had been labelled asthmatic at the age of three and described a history of two to three chest infections per year in her childhood. She had been a full term delivery, never smoker, who reported no allergic or atopic symptoms and had a family history of asthma. After being asymptomatic in early adult life, she re-presented over the last three years with recurrent chest infections requiring courses of corticosteroids and antibiotics every month, which limited her professional career. In between exacerbations, she reported episodic breathlessness, cough and wheeze. She had been a professional classical singer and had to stop singing due to her respiratory symptoms. She was taking combined inhaled corticosteroids and long acting beta-agonists along with short action beta-agonists when needed, although she found these therapies ineffective.

In February 2015 she was referred from her general practitioner direct to the regional “Severe Asthma Clinic” reporting increased wheeze and recurrent respiratory tract infections.

Her predominant symptoms were shortness of breath and fatigue, non-responsive to standard asthma treatment, along with daily sputum production. Triggers to her symptoms included cold food, cold drinks, dust, perfumes, cold weather, animal hair and stress. She had a peak-flow diary showing peak-flows ranging from 110 to 210 L/min. She reported frequent nocturnal wakening with cough and breathlessness. On physical examination she had an audible expiratory wheeze and there was also an inspiratory wheeze component, which was considered almost “stridulous”.

The “Severe Asthma Clinic” performs a standardised assessment on all referrals considered appropriate, including full blood count, total IgE, allergy antibodies, laryngoscopy [in order to discount vocal cord dysfunction (VCD)] and a thoracic CT scan (to identify bronchiectasis). In this patients case a bronchoscopy was also planned due to the stridulous wheeze, but the patient declined the latter investigation.

Total and specific IgE were in the normal range. Blood eosinophilia was $0.44 \times 10^9$/L. Laryngoscopy showed no evidence of VCD but laryngeal sensitivity was evident.

Thoracic CT demonstrated a gross dilatation of the oesophagus, which was distended with food debris and fluid from the gastro-oesophageal junction right up to her larynx, compatible with achalasia (Fig. 1). The dilated oesophagus was displacing the trachea and central airways causing important compression of the tracheal lumen. The long

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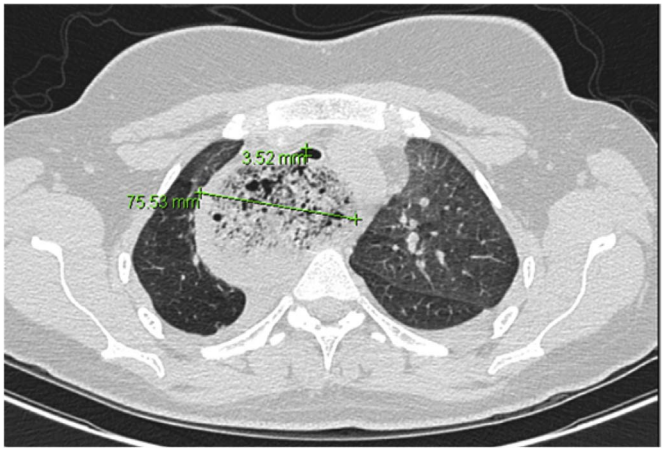


**Fig. 1.** Thoracic HRCT showing dilatation of oesophagus with food residue near the larynx.

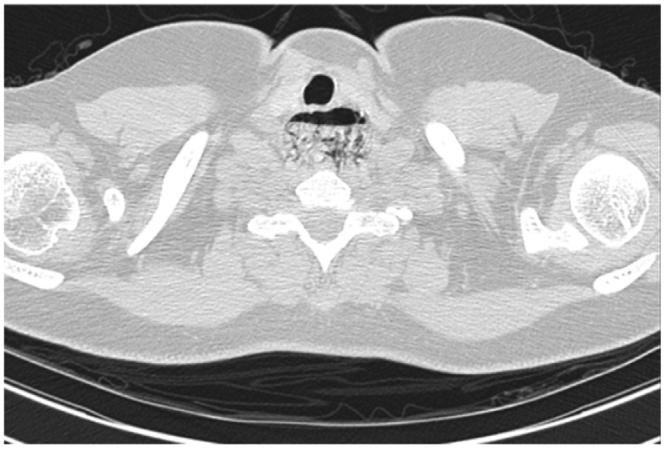

**Fig. 2.** Thoracic HRCT showing mega-oesophagus with the oesophagus causing severe compression to the trachea near the Aortic Arch.

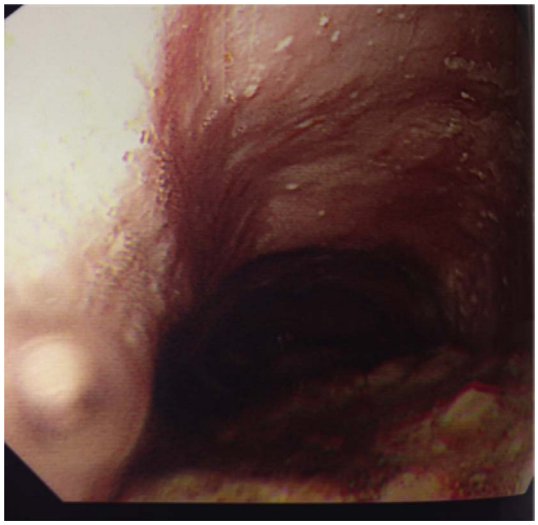

**Fig. 3.** Gastroscopy showing dilated oesophagus with food debris.

axis diameter of the trachea at the narrowest point was 3.5 mm near the Aortic Arch, where the oesophagus was most dilated (Fig. 2). Further investigations included a gastroscopy, which confirmed the presence of a very dilated oesophagus with significant food residue and normal mucosal appearances (Fig. 3). There was marked hypertrophy of the longitudinal and circular muscle layers of the oesophageal wall. A manometry study confirmed features consistent with achalasia.

Her stridor came from tracheal compression via the large oesophageal mass. It is possible that this degree of dilatation developed over many years. There were no preceding X-Rays for comparison.

The patient subsequently underwent a laparoscopic Heller's Myotomy with anterior fundoplication in September 2015. After Heller's myotomy her symptoms improved dramatically and she was asymptomatic from a respiratory perspective. She had no symptoms of asthma, and her exercise tolerance had improved dramatically. The patient did not require any inhalers following the surgery. Her peak flow prior to surgery was about 100 L/min, and it increased up to 350 L/min post surgery. Her current spirometry showed FEV1 3.4L and FVC 4L. Her respiratory infection rate reduced from monthly to none in 18 months since surgery. Moreover, gastroscopy one year after surgery showed normal appearance and her chest X-Ray was reported normal. Remarkably, she has been able to return to singing.

## 3. Discussion

Achalasia is an uncommon oesophageal muscle disorder characterized by failure of relaxation of the lower oesophageal sphincter accompanied by a loss of peristalsis in the oesophagus. It is usually diagnosed in patients between the ages of 25 and 60 with an annual incidence of approximately 1.6 cases per 100,000 individuals and a prevalence of 10 cases per 100,000 individuals [1].

It results from inflammation and degeneration of neurons in the oesophageal wall [2] and the neurons that remain are often surrounded by lymphocytes and eosinophils [3,4]. This inflammatory degeneration involves the nitric oxide-producing, inhibitory neurons that affect the relaxation of oesophageal smooth muscle. Yet, the aetiology of the inflammatory degeneration of neurons in idiopathic achalasia is not known. In our patient, blood eosinophils were high representing inflammation.

The manifestations of achalasia depend on the degree and location of ganglion cell loss [5]. It usually has an insidious onset, and disease progression is gradual. Patients typically experience gastrointestinal manifestations. Dysphagia for solids (91%) and liquids (85%), regurgitation of undigested food or vomiting (76–91%), and substernal chest pain and heartburn (40–60%) are the most frequent presenting symptoms [6,7]. From a respiratory point of view achalasia can cause aspiration and subsequent chest infections and pneumonia.

Rarely, tracheal compression caused by a severely dilated oesophagus is observed. Life threatening acute dyspnoea and distress has rarely been reported previously in the literature as the presenting symptoms for achalasia in adulthood [8,9]. In our case, despite having an important narrowing of the tracheal diameter, the onset was not with acute airway distress. Chronic respiratory symptoms due to tracheal compression as a complication of known achalasia have also been previously described [10,11].

Spontaneous belch reflex occurs when the lower oesophageal sphincter relaxes to allow the food bolus to pass into the stomach. In patients with achalasia, the upper sphincter also relaxes allowing air to exit. Failure of this reflex, presumably secondary to achalasia, leads to progressive oesophageal dilatation and airway obstruction [12]. In early childhood, airway irritation and asthma symptoms might have occurred as a result of microaspirations and asymptomatic gastro-esophageal reflux secondary to achalasia.

Oesophageal manometry is required to confirm the diagnosis of achalasia and endoscopic evaluation with upper gastrointestinal endoscopy should also be performed.

Without treatment, patients with achalasia can develop progressive dilation of the oesophagus. Severe dilatation or mega-oesophagus is considered when the transverse width of the oesophagus is more than 70 mm [8].

In our case, the mega-oesophagus caused progressive tracheal

compression and simulated tracheomalacia (TM) and symptoms that were compatible with asthma. However, there was no response to asthma treatments, which should have led to an asthma diagnosis review [13].

Chronic compression of the trachea can cause secondary tracheomalacia. This is most commonly due to a benign mediastinal goitre [14] however; it can also be caused by malignancy, vascular compression, abscesses, cysts or other lesions.

The major symptoms and signs of TM are dyspnoea, cough and sputum retention. Wheezing or stridor may also exist, but almost exclusively in expiration. They are nonspecific symptoms and are often attributed to alternative diagnoses, mostly asthma, as in the present case.

Bronchoscopic visualization of dynamic airway collapse is considered to be the diagnostic gold standard [15] although inspiratory and expiratory CT scan may also be useful in the diagnosis. Lung function can show several features usually demonstrating obstruction, but this can be either a large or small airway pattern.

In our case, the patient had the diagnostic CT prior to performing detailed lung function. And once performed, the lung function was cancelled due to the degree of the observed tracheal compression. No preceding spirometry was available from primary care records. However, she had very low (and variable) peak-flow rate which may have delayed a review of the diagnosis.

This case report demonstrates the need of a correct diagnosis of asthma. This is most relevant if conventional treatment for asthma fails.

Our severe asthma service already has a large population of more classical cases of TM, who were receiving excess and inappropriate asthma therapy, before identifying TM as a cause of uncontrolled symptoms. This case broadens the differential diagnosis, with in this case a thankfully reversible cause.

## Conflict of interest

There are none financial or personal conflicts of interests.

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