Rare Cause for Common Symptoms; Adult Tracheomalacia- A Case Report

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Abstract
Tracheomalacia characterized by weakness of the tracheal wall is encountered uncommonly in adult clinical practice. However, many cases are under-recognized. Here we report a case of tracheomalacia in an adult, managed as poorly controlled asthma for a long duration prior to diagnosis.

A 44 year old male with a past history of mild intermittent bronchial asthma, diabetes mellitus and hypertension presented with cough and shortens of breath for 2 years duration with significant progressive symptoms associated with a change in voice over 5 months. Physical examination was unremarkable. Chest radiograph and pulmonary functions were within normal limits. However, high resolutions computerized tomography (HRCT) revealed crescent shape collapse of the trachea. Near complete obliteration of tracheal lumen due to approximation of anterior and posterior tracheal walls was noted in bronchoscopy, confirming the diagnosis of tracheomalacia. He underwent successful implantation of endotracheal silicon stent with significant resolution of symptoms.

In conclusion, clinical features of tracheomalacia are non-specific. Therefore it is likely to be under-recognized and managed as for other common respiratory pathologies. The high degree of suspicion on this uncommon condition is required to avoid misdiagnosis.

Key words: *Tracheomalacia, Bronchial asthma, intratracheal stenting*

Introduction

Tracheomalacia is characterized by weakness of the tracheal wall due to the softening of tracheal cartilages and hypotonia of the myo-elasitc elements leading to an excessive approximation of anterior and posterior tracheal walls resulting of narrowing of the lumen during expiration(1). The primary form of tracheomalacia is seen in infants as a congenital defect in tracheal cartilages. Secondary or acquired tracheomalacia is increasingly being recognized among adults though exact incidence is yet undetermined (2). Adult tracheomalacia can be idiopathic or secondary to many aetiologies such as tracheobronchitis, smoking, chronic obstructive pulmonary disease (COPD), asthma, and connective tissue disorders, or mechanical factors such as extrinsic compression, trauma, and intubation (3). Signs and symptoms of tracheomalacia are non-specific; hence it is increasingly recognized as an imitator of other common respiratory pathologies like asthma and COPD (3).

Case presentation

A 44 year old Sri Lankan non-smoking male presented to us with chronic cough for 2 years with significant worsening over the last 5 months. He was suffering from mild intermittent type bronchial asthma since childhood, however had good disease control prior to the current presentation. The cough was intermittent at the outset, but worsened in severity and frequency during the last 5 months to cause persistent daily symptoms. His cough was predominantly non-productive and never associated with haemoptysis. He also noticed shortness of breath, especially at exercise, with the severity of the modified research council (mMRC) grade II at presentation. There was no wheezing or stridor, however, he complained of alteration of the quality of voice. Past medical history revealed diabetes mellitus diagnosed for 3 years, well controlled on oral hypoglycemic agents and hypertension for 1 year, managed with losartan (angiotensin receptor blocker) only. Systemic inquiry failed to recognize fever, constitutional symptoms, recurrent chest infections, features of connective tissue diseases, gastro-esophageal reflux disease or past history of endotracheal

intubations. He was receiving treatment for poorly responding bronchial asthma consisting of inhaled steroids, beta-agonist, long acting muscarinic antagonists, theophyllin, montelukast and oral steroids while presenting to our unit.

Physical examination revealed averagely built male without dyspnoea, pallor, clubbing or lymphadenopathy. Respiratory and otolaryngeal examinations were clinically unremarkable as were cardiovascular, abdominal and neurological systems. Initial investigations found white cell count 8000/mm3 with normal differentials, haemoglobin 14.2 g/dl, platelets 273000/mm3, sedimentation rate 10mm/hour, C-reactive protein 4.0 mg/dl (normal <10), normal renal and liver profile. Pulmonary function tests including spirometry and diffusion capacity were within normal limits. Chest radiograph showed no abnormality, but high resolution computered tomography revealed crescent shape collapse of the trachea during expiration suggestive of tracheomalacia (Figure 1). Fiber-optic bronchoscopy demonstrated near total obliteration of tracheal lumen involving the whole length by approximation of anterior and posterior tracheal walls at forced expiratory techniques confirming tracheomalacia (Figure 2).

He was treated with bronchodilators, physiotherapy and breathing exercise, which provided little benefit. Therefore, Y shape tracheobronchial silicone stent was inserted, which alleviated his symptoms (figure 3).

Discussion

Tracheomalacia, and its extension into a bronchial tree known as tracheobronchomalacia are uncommon conditions of adults characterized by loss of integrity of airway wall leading to expiratory airflow limitation (1,2). The true incidence of tracheomalacia in adults is yet unknown (2). However, it has been reported in 1-4.5% of all patients undergoing bronchoscoies, and in as many as 23% of patients with a diagnosis of chronic bronchitis (2). This wide variation is due to a lack of uniformly applied criteria for diagnosis and interchangeable use of the term for similar, yet distinctive entities like excessive dynamic airway collapse (EDAC) (2, 3). EDAC is a pathologically different condition than tracheomalacia where the luminal obstruction occurs due to inward bulging of the atrophic muscular fibers in the posterior airway membrane during exhalation in contrast to cartilaginous weakness in latter(2,3).

Currently accepted criteria for the diagnosis of tracheomalacia require a reduction of tracheal lumen >50% during expiration (3,4). However, it has been recently shown that many healthy volunteers without respiratory symptoms frequently exceed this diagnostic threshold (5). Therefore, image guided assessment of tracheal cross sectional area alone should not be taken as diagnostic of tracheomalacia, without considering the clinical features (2). Direct observation by bronchoscopy of the tracheal lumen during tidal and forced expiratory cycles remains the gold standard for evaluating airway collapse (2,6). However, dynamic expiratory CT has emerged as a highly sensitive, non-invasive method of evaluation, which has been shown to provide concordant results with bronchoscopy (2,6). Lung function tests may demonstrate reduced expiratory flow, dynamic airway compression, biphasic flow-volume loop or flow oscillations (3). However, it may show normal results in up to 21% of moderate to severe tracheomalacia as in our case (2).

Clinical features of tracheomalacia are non-specific and include cough, wheezing, dyspnoea, difficulty in clearing secretions, and recurrent bronchitis or pneumonia which are often similar to those of patients with chronic lung disease such as COPD or asthma (3). Therefore it has a high tendency for being misdiagnosed unless clinically suspected. Moreover, tracheomalacia often co-exists with other common respiratory pathologies such as asthma and COPD. Aetiologically, adult tracheomalacia can be idiopathic, genetic or secondary to chronic airway irritation as seen in smokers, malignancy, chronic infections, long term ventilation, trauma, chronic compression of the tracheobonchial tree and inflammatory conditions like relapsing polychondrits (3, 7). However, we were unable to recognize any possible aetiology in our patients.

Treatment depends on the degree of airway collapse and severity of clinical symptoms determined by thorough objective assessment. It is recommended that all symptomatic patients should receive medical management before considered for minimally invasive procedures and surgical options (3). A wide array of therapeutic options including bronchodilators, expectorants, flutter valve and such devices, rehabilitation, purse lip breathing and continuous positive airway pressure are utilized for medical management (2). Moreover, underlying conditions, if present, should be optimized according to practice guidelines (3).

Intratracheal stent placement has been used for the treatment of adult tracheomalacia since 1965, especially in patients where medical management alone has failed (7). Intratracheal stents are broad of two types- metal and silicon. Metal stents are less likely to migrate or be obstructed by mucous plugging. However, the main disadvantages of metal stents are the formation of granulation tissue with the risk of re-stenosis and difficulty in removal and propensity for collapse or fracture (3). In contrast, silicon stents are easier to insert, reposition and remove. However, they are more likely to migrate than metal stents (7).

Conclusion

Tracheomalacia is an uncommon condition in adults. It often produces non-specific symptoms and tends to co-exist with other common respiratory pathologies like asthma and COPD, and therefore easily misdiagnosed. Hence, tracheomalacia should be considered as a possibility for unusual presentations or inadequate control of common respiratory diseases.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal

Competing interest

The authors declare that they have no competing interests.

Authors' contribution

DM made the clinical diagnosis and supervised the manuscript drafting. AB, LB, SD and SS drafted the first manuscript, reviewed the literature and involved in direct management of the patient. All authors read and approved the final manuscript.

Authors' information

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Figures of tracheomalacia case

Figure 01- HRCT showing narrowing of the tracheal lumen during expiration

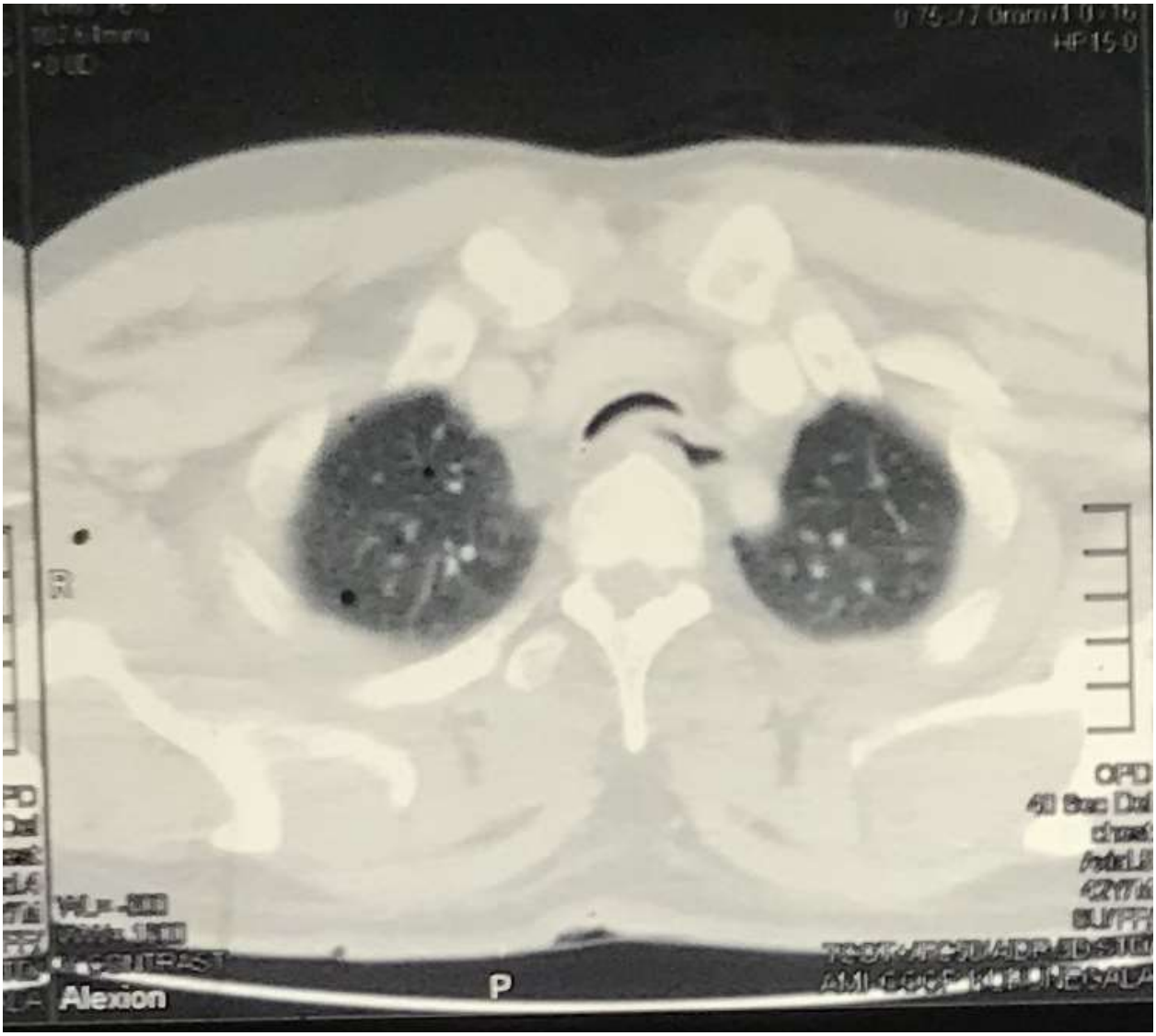

Figure 02- Bronchoscopic appearance of tracheal collapse

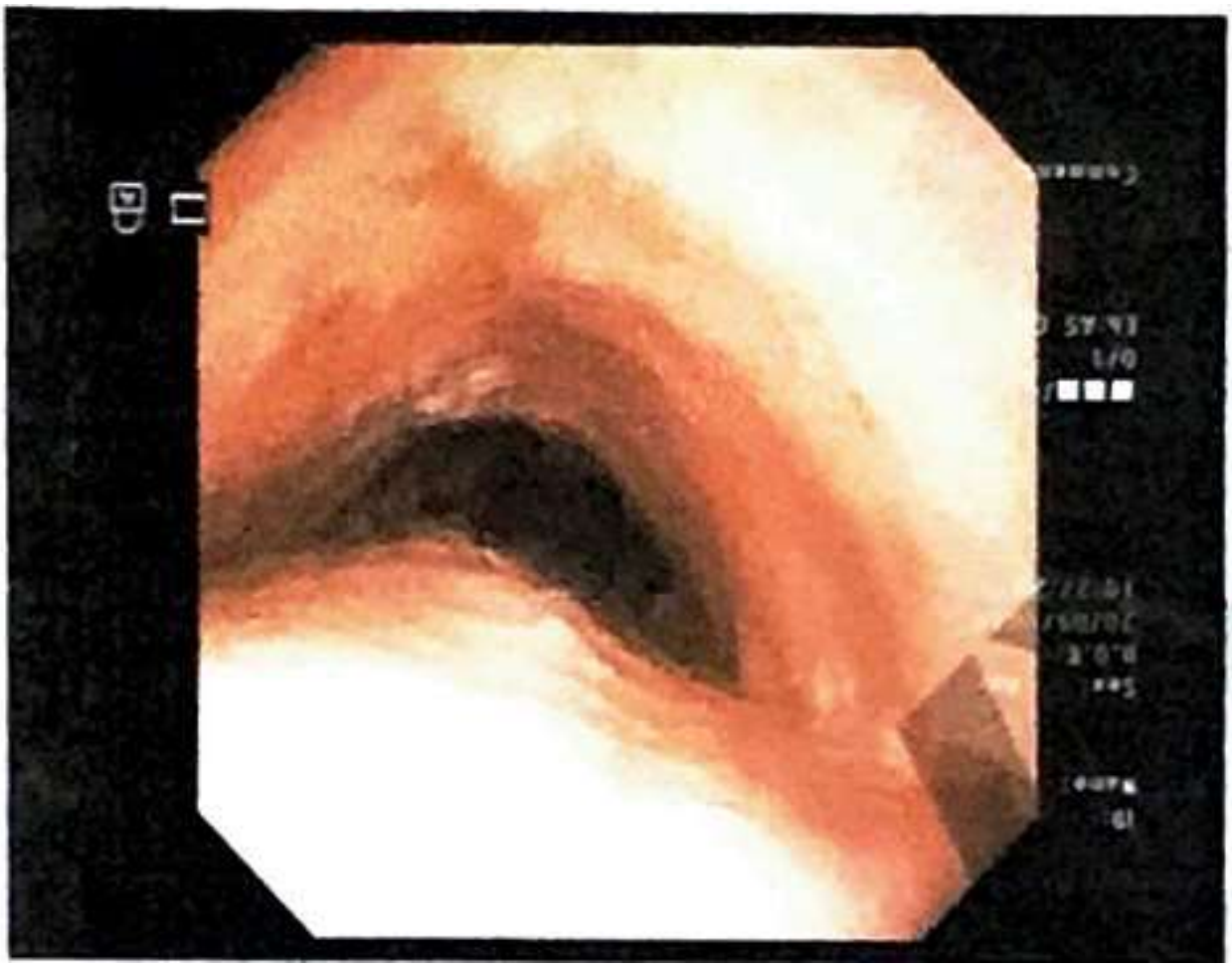

Figure 03- Following insertion of tracheal Y stent