JOURNAL OF COMMUNITY HOSPITAL INTERNAL MEDICINE PERSPECTIVES, 2017
VOL. 7, NO. 5, 303–306
https://doi.org/10.1080/20009666.2017.1383119

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REVIEW ARTICLE

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Recurrent dyspnea and wheezing- pulmonary function test and dynamic computed tomography may unfold the diagnosis of tracheobronchomalacia

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ABSTRACT

Tracheomalacia patients often present with nonspecific symptoms like cough, wheezing and dyspnea. Tracheomalacia diagnosis is usually attributed to alternative common conditions such as asthma or chronic obstructive lung disease. Certain maneuvers, like forced expiration, or recumbent position may elicit subtle signs of tracheomalacia. Ordering novel pulmonary function testing in sitting upright and supine positions may provide additional clues to suspect tracheomalacia, which can be confirmed by either dynamic chest tomography or bronchoscopy.

ARTICLE HISTORY
Received 14 July 2017
Accepted 13 September 2017

KEYWORDS
Asthma; chronic obstructive lung disease; positional dyspnea; tracheomalacia; pulmonary function test; dynamic chest tomography; bronchoscopy

1. Introduction

Asthma and chronic obstructive lung disease (COPD) are common respiratory conditions characterized by episodes of coughing, shortness of breath, and wheezing. Environmental triggers and smoking are commonly associated with their exacerbation. Orthopnea (dyspnea in the recumbent position), though not a physical finding in obstructive lung diseases, is seen with congestive heart failure, obstructive sleep apnea (OSA), morbid obesity, goiter, bronchogenic carcinoma, and tracheobronchomalacia (TBM) [1,2]. Structural obstruction of airway in conditions such as TBM can elicit a symptom of positional dyspnea, more pronounced when lying flat, in association with other findings consistent with obstructive lung diseases, making the diagnosis difficult. TBM is underdiagnosed, and is often missed clinically [3,4]. Certain maneuvers like forced expiration, or certain positions like recumbency, can sometimes elicit signs of TM. Ordering appropriate testing such as pulmonary function tests (PFT), dynamic chest tomography (CT), or bronchoscopy can be rewarding to identify undiagnosed tracheobronchomalacia cases.

2. Case report

A 60-year-old woman with a 45 pack-year history of smoking presented with positional shortness of breath and wheezing for more than a month when lying flat. The patient had had multiple admissions in the last year for documented COPD exacerbations. She had never fully recovered from those symptoms despite several courses of antibiotics, corticosteroids and breathing treatments. Her significant past medical history included essential hypertension, type 2 diabetes mellitus, and anxiety disorder. Medications on admission included montelukast, budesonide-formoterol fumarate aerosol, ipratropium- albuterol by nebulization, roflumilast tablet, tiotropium aerosol solution, prednisone, lansoprazole, amlodipine, insulin glargine, lorazepam, fluoxetine, and nicotine patch. Surgical history was significant for subtotal colectomy 3 years ago for colon cancer currently in remission. No history of prior prolonged intubation, OSA, or acid reflux disease was reported. She continued to smoke half a pack per day. The patient had worked as a janitor, but stopped working 5 years ago due to dyspnea.

On physical examination, the patient's vital signs revealed no fever, blood pressure of 132/74 mmHg, heart rate 90 bpm, respiration rate of 18/min, pulse oximetry showed spO_2 of 95% with 2L nasal cannula, and body mass index was 25 kg/m^2. Nose and ear examination were normal, and there was no goiter or lymphadenopathy. Chest examination on inspection showed midline trachea, bilateral equal chest expansion, and no increased antero-posterior diameter. On auscultation, diffuse expiratory wheezes were heard in all lung fields and stridor was heard over the anterior chest wall. No crackles or rales were noted;

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Patient consent obtained- Yes

there was no jugular venous distension and no cardiac murmurs. No clubbing, cyanosis, or pedal edema was noted on extremity examination.

Laboratory studies showed no peripheral blood eosinophilia, hemoglobin (Hb) 12 gm/dL, HbA_{1C} 8.2%, creatinine 0.74 mg/dL, and normal liver chemistry, cardiac enzymes, and B-type natriuretic peptide. Chest radiograph showed no infiltrate or mass. Electrocardiography had shown normal sinus rhythm, and left ventricular hypertrophy. Echocardiography had normal left ventricular function, right ventricular systolic pressure and normal valves.

The patient had no prior PFT test in the chart and with findings of positional dyspnea, a diagnosis other than COPD was suspected. Positional flow volume spirometry was performed without a bronchodilator. Forced expiratory volume in 1 second (FEV_1) was 55% of predicted and decreased to 41% in sitting position. Forced vital capacity (FVC) was 53% of predicted in sitting position, decreasing in supine position to 42% of predicted. Interestingly, the FEV_1/FVC ratio was >75% in both sitting and standing positions, questioning the initial presumed diagnosis of COPD. Dynamic inspiratory and expiratory CT was done the next day and showed a 'frown sign' of the intrathoracic trachea with >50% expiratory collapse in airway lumen, suggestive of TBM (Figure 1). No evidence of pulmonary infiltrates, pleural effusions, or bronchiectasis was noted on CT. A tracheobronchial stent was placed by a cardiothoracic surgeon.

The patient was referred to pulmonary rehabilitation and advised to quit smoking. She had significant improvement of symptoms after placement of the stent, but unfortunately continued to smoke tobacco. At 3-month follow-up, she was admitted to hospital with atypical pneumonia and hemoptysis. The tracheobronchial stent was therefore removed at that time. She did have a follow-up bronchoscopy that showed mild airway malacia, thought to be an underestimation of disease severity as the patient was unable to perform forced inspiratory and expiratory maneuvers secondary to sedation.

3. Discussion

Expiratory wheezing in a smoker usually suggests asthma or COPD. Narrowing the differential diagnosis can miss alternative treatable conditions and lost opportunity [3,4,5]. Positional dyspnea and wheezing are commonly seen with congestive heart failure, goiter, OSA, and lung cancer [1,2], but this patient's history and our findings on examination did not suggest those conditions. Abnormal PFT during sitting upright and supine positions led to a suspicion of

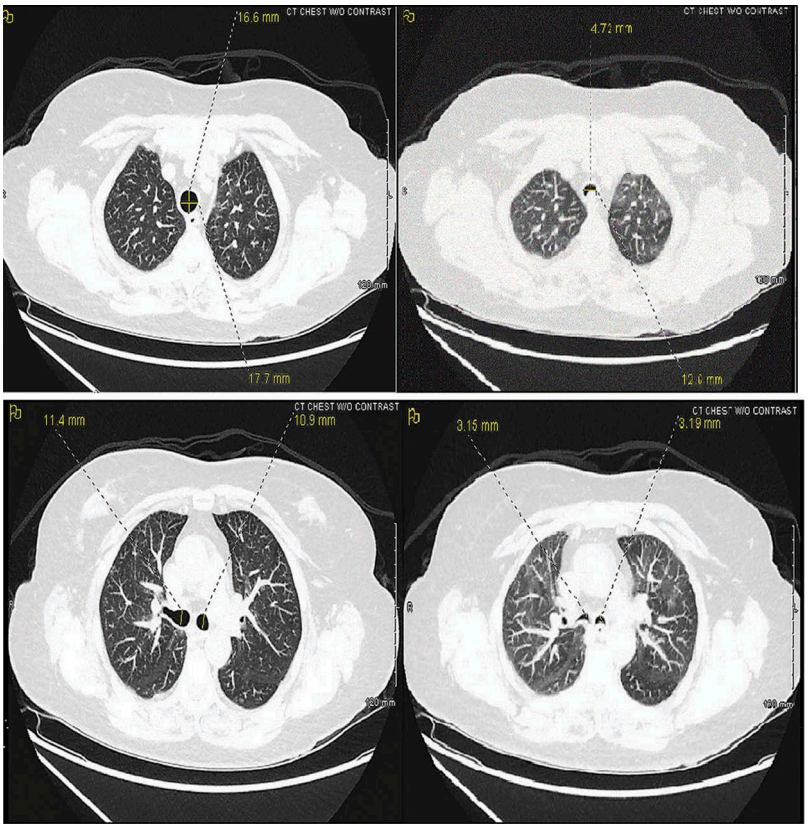

Figure 1. Axial noncontrast computed tomography (CT) during inspiration (left panels) showing a normal tracheal and both main bronchi lumen. During expiration, axial noncontrast CT (right panels) showing thin crescent-shaped ('frown sign') tracheal lumen and both mainstem bronchi.

a significant airway disease such as tracheobronchomalacia, which was confirmed by dynamic chest CT.

TBM is an underdiagnosed condition that presents with nonspecific symptoms such as dyspnea, cough, and recurrent infections. Certain maneuvers can sometimes elicit signs of TM including forced expiratory, cough, a Valsalva maneuver or by recumbent position. The symptoms and signs of TM can be nonspecific and are often attributed to alternative conditions, and patients are frequently misdiagnosed with more prevalent diseases such as asthma or COPD [3].

The prevalence of TBM in adults is uncertain. In a series of 2150 patients subjected to bronchoscopy, 94 (4.5%) were found to have TBM [6]. In another study, TBM was diagnosed in 542 (12.7%) of 4283 patients with pulmonary disease who underwent bronchoscopy [7].

In adults, the most common cause of tracheomalacia (TM) is prolonged mechanical ventilation. Other causes of segmental tracheomalacia include prolonged external pressure on the tracheal wall by a large substernal goiter or a congenital vascular ring [1,8]. More diffuse TM can be seen with relapsing polychondritis. Reported risk factors of TM pathogenesis are cigarette smoking and long-term use of glucocorticoids [3]. Up to 5% of patients with severe emphysema and smokers have TM, suggesting that chronic inflammation due to inhalation of irritants – e.g. cigarette smoking and recurrent infections – may cause TM [9].

Tracheomalacia is characterized by the flaccidity of the supporting cartilage and hypotonic myo-elastic elements. These factors cause tracheal collapse, especially in recumbent position, and during times of increased airflow associated with increased airway dynamic pressure changes such as coughing. Tracheomalacia is also associated with intermittent or continuous dyspnea, difficulty in clearing secretions, wheezing, cough, and recurrent bronchitis and pneumonia [10].

The utility of PFT in diagnosis of TM is supportive only. One study demonstrated airway obstruction being proportional to the severity of TM, as 44% of patients with TM had an obstructive ventilatory defect, 18% had restrictive ventilatory defect, 17% had a mixed defect, and 21% were within normal limits. The abnormal findings of PFT included low maximum forced expiratory flow (81%), notched expiratory loop (9%), and expiratory oscillations (2.6%) [11]. Typically, PFT is performed in a sitting position, which can easily miss the positional change in flow volumes. To our knowledge, no studies have been performed to compare PFT during supine and seated positions. Also in this prospective case series, normal PFTs were found in a substantial number of patients with moderate to severe TM, showing that PFT alone should not be used to make the diagnosis.

Tracheomalacia is a form of dynamic airway collapse, and diagnosis can be missed with routine lung imaging. Once TM is suspected, dynamic expiratory chest CT is currently used and many radiologists use a luminal narrowing of 50% on exhalation as a benchmark for diagnosis. Anterior bulging of the posterior tracheal wall gives the appearance of a crescent shape, as seen in Figure 1. To date, only small case series have been published to detect the sensitivity and specificity of dynamic chest CT in comparison to bronchoscopy. In a study of 29 patients, to access the accuracy of CT for detecting malacia, chest CT correctly diagnosed malacia in 97%. In this study bronchoscopy was used as the initial diagnostic method and CT was performed within a week of the procedure to compare the results [12]. Although bronchoscopy is a useful tool for diagnosis, it is invasive procedure and may underestimate the severity of TM because sedation during the procedure may prevent maximal effort during forced exhalation maneuver, as noted in our patient. A study of 10 patients where 23 pulmonologists at different levels of training were involved to discuss bronchoscopy results found high intra- and inter-observer agreement in evaluation of degree of collapse to diagnose TBM, but did not specify the sensitivity and specificity of the test in diagnosing the condition [13].

Treatment options include use of silicone Y-shaped stents, but variable efficacy has been reported. While >50% reduction of cross-sectional area on CT and bronchoscopy has been used as threshold for diagnosis, no clear indications for surgical intervention exist [14,15]. Tracheobronchoplasty remains the long-term treatment of choice and has been shown to improve symptoms [16]. Nonsurgical candidates can be managed with positive pressure therapy [17].

This case illustrates the need to have a broader differential diagnosis when symptoms are exacerbated in recumbent position and fail to improve with asthma or COPD standard treatments. Ordering PFT in both upright sitting and supine positions, followed by dynamic chest CT/bronchoscopy if abnormal, may identify undiagnosed cases of tracheomalacia.

Disclosure statement

No potential conflict of interest was reported by the authors.

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References

[1] Stang MT, Armstrong MJ, Ogilvie JB, et al. Positional dyspnea and tracheal compression as indications for goiter resection. Arch Surg. 2012;147(7):621–626.

[2] Mahler DA, Snyder PE, Virgulto JA, et al. Positional dyspnea and oxygen desaturation related to carcinoma of the lung. Up with the good lung. Chest. 1983;83 (5):826–827.
[3] Solomon DA, Fanta CH, Levy BD, et al. Clinical problem solving. Whistling in the dark. NEJM. 2012;366(18):1725–1730.
[4] Kerolus G, Ikladios O. Tracheomalacia and recurrent exacerbations of chronic obstructive pulmonary disease: a case report and review of the literature. J Community Hosp Intern Med Perspect. 2016 Dec 15;6(6):33540.
[5] Alici IO, Kar Kurt O, Dursun AB, et al. Two cases of tracheal disease misdiagnosed as difficult to treat asthma. Respir Care. 2013;58(11):e133–e137.
[6] Jokinen K, Palva T, Sutinen S, et al. Acquired tracheobronchomalacia. Ann of Clinic Research. 1977;9 (2):52–57.
[7] Ikeda S, Hanawa T, Konishi T, et al. Diagnosis, incidence, clinicopathology and surgical treatment of acquired tracheobronchomalacia. Nihon Kyobu Shikkan gakkai Zasshi. 1992;30:1028–1035.
[8] Odell DD, Gangadharan SP, Majid A. Pulmonary artery sling. A rare cause of tracheomalacia in the adult. J Bronchol Intervent Pulmonol. 2011;18:278–280.
[9] Bhatt SP, Terry NL, Nath H, et al. Association between expiratory central airway collapse and respiratory outcomes among smokers. JAMA. 2016;315(5):498–505.
[10] Kurnutala LN, Joshi M, Kamath H, et al. A surprising cause of wheezing in a morbidly obese patient: a case report. Int Med Case Rep J. 2014;7:143–145.
[11] Majid A, Sosa AF, Ernst A, et al. Pulmonary function and flow-volume loop patterns in patients with tracheobronchomalacia. Respir Care. 2013;58:1521–1526.
[12] Lee KS, Sun MRM, Ernst A, et al. Comparison of dynamic expiratory CT with bronchoscopy for diagnosing airway malacia: a Pilot evaluation. Chest. 2007;131(3):758–764.
[13] Majid A, Gaurav K, Sanchez JM, et al. Evaluation of tracheobronchomalacia by dyanamic flexible bronchoscopy. A pilot study. Ann Am Thoracic Society. 2014;11(6):951–955.
[14] Buetrago DH, Wilson JL, Parisk M, et al. Current concepts in severe adult tracheobronchomalacia: evaluation and treatment. J Thorac Dis. 2017 Jan;9(1): E57–E66.
[15] Ernst A, Majid A, Feller-Kopman D, et al. Airway stabilization with silicone stents for treating adult tracheobronchomalacia; a prospective observational study. Chest. 2007;132:609–616.
[16] Majid A, Guerrero J, Gangadharan S, et al. Tracheobronchoplasty for severe tracheobronchomalacia: a prospective outcome analysis. Chest. 2008;134:801–807.
[17] Patout M, Mylott L, Kent R, et al. Trial of portable positive airway pressure for the management of tracheobronchomalacia. Am J Respir Crit Care Med. 2016;193(10):e57.