

An unusual case of ‘exacerbation of COPD’

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Teaching Points

- Consider tracheomalacia when patients with suspected COPD present atypically, particularly where breathlessness is eased in the left lateral position.
- Use of CTPA as a diagnostic test for pulmonary embolism (PE) may identify an alternative diagnosis to account for the patient's symptoms when PE is excluded.
- CT scanning, bronchoscopy and pulmonary function testing with flow-volume loops will help to confirm the diagnosis of tracheomalacia.
- Surgery or tracheal stenting may be feasible in some cases of tracheomalacia.
- Delayed diagnosis of tracheomalacia in patients with COPD may prevent definitive treatment from being undertaken if underlying COPD deteriorates during this period.

Abstract

A 66 year old man who had been diagnosed with COPD 18 months earlier was readmitted with breathlessness unresponsive to bronchodilators and steroids. His symptoms were noticeably improved by lying on his left side. CTPA showed no evidence of pulmonary embolism but raised the possibility of tracheomalacia, which was later confirmed with bronchoscopy and lung function testing. The classification and treatment options for tracheomalacia are discussed.

Keywords

Tracheomalacia; Chronic obstructive airways disease.

Case History

A 66-year old man was admitted to hospital complaining of worsening dyspnoea at rest. He gave an 18 month history of chronic obstructive pulmonary disease (COPD), and had been admitted to hospital 7 times in this period with similar symptoms. There was no significant cough or wheeze, and there was no sputum or pyrexia to suggest any infective component to his current illness. However he commented that his dyspnoea was significantly improved when lying on his left side. He was a recent ex-smoker with a thirty pack-year history. His past medical history included renal cancer for which he had undergone a nephrectomy 6 years ago, and prostate cancer, which was being managed medically. His COPD was being treated with regular inhaled combivent (two puffs qds), seretide (one puff bd), tiotropium bromide (18mcg inhaled od), aminophylline 225mg orally od, and nebulised salbutamol as required. He had also received a number of courses of oral prednisolone during the past 18 months, in which period he had gained around 20Kg in weight.

Admission blood gas analysis while breathing 28% oxygen revealed $p\text{CO}_2$ 5.46, $p\text{O}_2$ 11.22, HCO_3 27.3 and BE 2.9. This was similar to blood gas analysis undertaken while breathing air 3 months earlier.

On examination, he was noted to have an irregular tachycardia at 110/min, with blood pressure 110/70. He was using accessory muscles of respiration with a respiratory rate of 22/min. He was obese, and appeared cushingoid, with gynecomastia and bilateral pitting oedema to his knee level. Respiratory examination revealed prominent expiratory wheeze, with equal but reduced air entry bilaterally. There was no inspiratory wheeze or stridor. His jugular venous pressure was not elevated and there were no cardiac murmurs.

His ECG showed fast atrial fibrillation but was otherwise normal. Chest x-ray showed features consistent with chronic obstructive airways disease without focal consolidation. His C-reactive protein was elevated at 94mg/l; Full blood count, liver and renal function were unchanged from previous admissions.

His symptoms were initially attributed to an exacerbation of COPD with cor pulmonale. However it was noted that there were some atypical features, particularly the lack of response to bronchodilators and steroids, the absence of cough and the apparent improvement in symptoms when lying on his left side. It was decided that pulmonary embolism required exclusion and CT Pulmonary Angiography was arranged (see Figure 1). Although there was no evidence of pulmonary embolism it was noted that the bronchi were small, and both the trachea and the bronchi had marked variability of diameter during respiration. This was felt to be consistent with a diagnosis of tracheomalacia.

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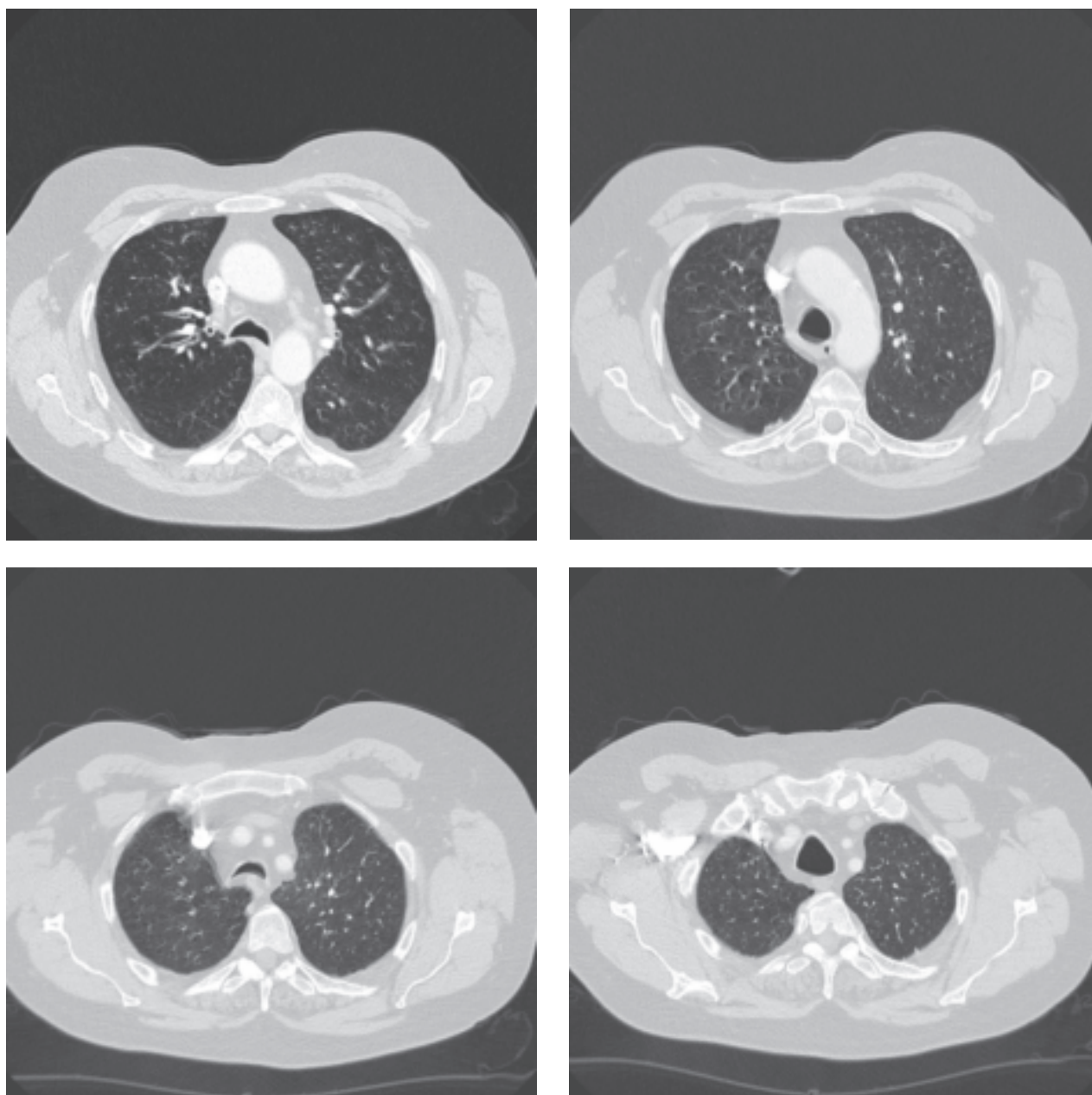


Figure 1. CT scan images at different thoracic levels. Note the varying diameters of the trachea as the CT cuts through ascending slices of his thorax.

These unusual CTPA findings were followed up by spirometry with flow volume loop tests and fibreoptic bronchoscopy. Spirometry revealed an obstructive defect (see table). The flow loop volume tests showed variable intrathoracic obstruction, typical of tracheomalacia. Bronchoscopy showed a collapsible trachea, particularly on expiration and coughing, confirming the diagnosis of tracheomalacia.

Surgical intervention and stenting were not deemed feasible on account of his co-existent COPD. Despite continued treatment with corticosteroids and bronchodilators his condition worsened; he contracted a further infection during his hospital stay and died, 5 weeks after admission.

Discussion

Tracheomalacia can be divided into three types, as illustrated in Table 2.¹

During the normal respiratory cycle, the trachea dilates and lengthens during inspiration, and narrows and shortens during expiration. In tracheomalacia, this process is accentuated due to the increased flexibility of the trachea, and this deforms a large segment of the trachea causing airflow obstruction.¹ Tracheomalacia most commonly affects the distal third of the trachea. A prospective study of eighty patients with acquired tracheomalacia measured airway collapsibility and related it to expiratory flow limitation. It concluded that central

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	Result	Predicted	% Predicted
FEV1	0.71	2.69	26%
FVC	1.18	3.45	34%
FEV1/FVC %	61%	75%	81%

Table 1. Spirometry results.

Classification	Aetiology	Associations/features
Type I	Congenital or intrinsic abnormality	Associated with: Tracheooesophageal fistula Williams-Campbell syndrome Down’s syndrome Congenital heart defects
Type II	Due to extrinsic defects causing chronic external tracheal compression	Secondary to: Benign Tumours Cysts Abscesses Vascular Rings
Type III	Acquired; presents in adult life	Secondary to: Intubation Tracheostomy Empysema Chronic bronchitis Chronic inflammation

Table 2. Classification of tracheomalacia.

airway collapse is not related closely to airflow obstruction. Expiratory flow limitation at rest occurs mostly in the peripheral airways (without central airway collapse), indicating symptoms of dyspnoea are related to small, and not central airway change.² In a small percentage of cases of secondary tracheomalacia, external compression from various structures affects the integrity of the tracheal wall and increases the compliance over the adjacent tracheal tissue.³ This may explain why our patient’s symptoms were eased by lying on his left side, where the pressure of his heart on his airways was minimized.

Chest X-ray is usually normal, although there may be evidence of hyperinflation, abnormalities causing

tracheal compression or co-existent COPD as in our case. Dynamic expiratory CT, in which multiple images are taken during expiration, may reveal evidence of airway collapse in patients with tracheomalacia. However, a definitive diagnosis relies on bronchoscopy, in which there is a classic triad of findings: loss of normal semicircular shape of tracheal lumen, anteroposterior narrowing of the tracheal lumen, and forward ballooning of the posterior membranous wall on inspiration as well as expiration.¹

In adults with acquired tracheomalacia, care is initially supportive, treating any co-existent COPD. Non-invasive ventilation can be used to treat hypercapnoeic acidotic respiratory failure. In certain patients, surgery may be possible, such as a tracheostomy to bypass the malacic segment; however this is rarely feasible as the malacic segment is commonly in the distal airways.¹ Stenting with tracheal ceramic rings is an alternative treatment option in some cases. In one retrospective study of twelve patients who underwent this treatment for tracheomalacia, satisfactory results were achieved after stenting in over 80% of patients.⁴ A prospective observational study of adults with tracheomalacia showed that airway stabilization with silicone stents can improve respiratory symptoms, quality of life, and functional status.⁵

Conclusion

The above case illustrates the importance of maintaining an open mind when treating patients with recurrent exacerbations of COPD. In this case the lack of response to treatment led to investigation to exclude pulmonary embolism to account for his symptoms. The chance finding of variability in tracheal diameter led to further investigations which confirmed the diagnosis of tracheomalacia. This may have explained why the patient’s symptoms improved while lying on his left side, when pressure from the heart on the trachea was reduced. This patient had significant co-existent COPD and obesity which prevented any surgical intervention. However, his condition had deteriorated significantly during the past 18 months, with significant weight gain, which may in part have related to the recurrent courses of corticosteroids. An earlier diagnosis may have improved his outcome and quality of life.

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