CASE REPORT

Flat trachea syndrome: a rare condition with symptoms similar to obstructive airway disease

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SUMMARY

Flat trachea syndrome, commonly known as 'tracheobronchomalacia', is a central airway disease characterised by excessive expiratory collapse of the tracheobronchial posterior membrane due to weakness in the airway walls. Patients present with symptoms such as chronic cough, dyspnoea and recurrent respiratory tract infections, which are often attributed to more common conditions such as asthma and chronic obstructive pulmonary disease (COPD). The term 'Flat Trachea Syndrome' was first proposed by Niranjan and Marzouk in 2010 following a retrospective study of 28 patients with the condition who underwent surgery for it. The authors advocated the term due to the primary abnormality being collapse of the posterior membranous wall of the central airways as opposed to softening of the tracheal cartilage (tracheobronchomalacia), which they proposed is a misnomer. We present a rare case of a patient with flat trachea syndrome on a history of COPD who initially presented with recurrent respiratory tract infections.

BACKGROUND

Flat trachea syndrome is an undertreated condition that can be fatal if not diagnosed, as it can eventually lead to respiratory failure and death.[1] It can be a difficult condition to recognise due to its symptoms being confused with obstructive airway diseases such as chronic obstructive pulmonary disease (COPD) and asthma. Familiarity with the condition can lead to earlier recognition and appropriate management of patients.

CASE PRESENTATION

A 68-year-old man presented with a 4-year history of a persistent productive cough with a seal-like barking quality. The cough was worse at night and during winter months. It was becoming increasingly debilitating for the patient. He also reported occasional spells of dyspnoea and an episode of cough syncope in the past and had been diagnosed with COPD 7 years previously. The severity of cough and its troubling nature led to numerous presentations to the acute medical clinic over a 4-year period with the diagnosis of recurrent respiratory tract infections being made and antibiotics being prescribed. One such presentation resulted in admission for suspected community acquired pneumonia. The patient is an ex-smoker who stopped 25 years ago with a 20-year pack history. He has been prescribed steroids during periods of infection and a regular bronchodilator (tiotropium bromide).

INVESTIGATIONS

Pulmonary function testing revealed an obstructive pattern with forced expiratory volume in 1 s (FEV1) of 1.96 L (66%), a forced vital capacity (FVC) of 3.42 L (89%) and FEV1/FVC of 57%. Chest X-ray did not show any significant changes. Owing to the long-term persistence of his symptoms, an inspiratory/expiratory CT was performed, which showed a collapsed trachea. Figure 1

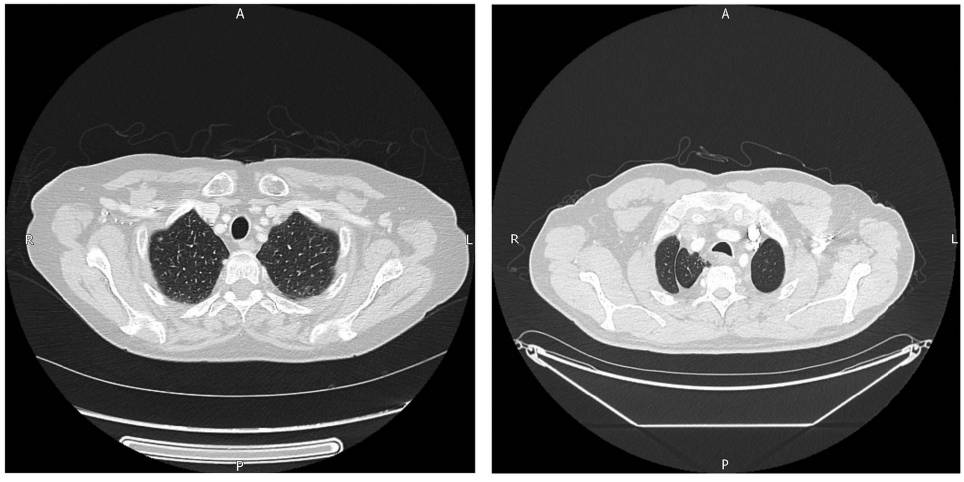

Figure 1 Left CT scan showing normal trachea. Right CT shows abnormal trachea of the patient discussed. Permission gained from trust with consent.

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compares this patient's CT of the thorax (expiratory) with that of a patient with a normal trachea (expiratory). Flexible bronchoscopy showed expiratory prolapse of the entire posterior tracheobronchial membranous wall.

DIFFERENTIAL DIAGNOSIS

- COPD
- Upper respiratory tract infection
- Chronic bronchitis

TREATMENT

A tracheal stent was inserted temporarily and subsequently the patient underwent a Modified Nissen Herzog Tracheal plication (tracheobronchoplasty) through a right thoracotomy incision. The material used in the plication was a polytetrafluoroethylene (PTFE) (Teflon) patch. The goal of surgery was to stabilise the posterior membranous wall of the trachea by suturing it to the PTFE patch. This increased rigidity resulted in the airway taking on a more appropriate 'C shape', as opposed to its original flat state.

OUTCOME AND FOLLOW-UP

Following surgery the patient's symptoms gradually resolved. The peculiar cough improved, becoming milder in character and less frequent in occurrence. The patient was regularly reviewed and was discharged at 1-year follow-up.

DISCUSSION

Flat trachea syndrome and tracheobronchomalacia (TBM) refer to the same condition. The term 'Flat Trachea Syndrome' was first proposed by Niranjan and Marzouk. They considered the term Trachebronchomalacia to be a misnomer. Flat trachea syndrome more appropriately describes the underlying pathophysiology. Flat trachea syndrome as well as TBM have the same clinical features, natural history and management, including investigations and treatment.

TBM is defined by softening of the elastic cartilage of the central airways, which leads to an expiratory collapse.[2] Niranjan and Marzouk[1] feel this is a misleading term as the main characteristic feature of this condition is expiratory prolapse of the posterior membranous wall of the central airways. Nonetheless, despite the term 'Tracheobronchomalacia' being a misnomer, it is commonly used in the literature and there is no other literature available on flat trachea syndrome aside from an Abstract by Niranjan and Marzouk;[1] thus we will continue to refer to the condition as 'Tracheobroncomalacia' for the rest of this discussion.

TBM can be classified into two categories: primary (congenital) and secondary (acquired).[3] The primary form of TBM is the most common congenital anomaly of the airways.[4] It usually improves due to the airway lumen increasing in diameter and cartilage becoming more rigid as a child ages.[5] Secondary TBM occurs mostly in middle-aged and elderly patients, and has many causes.[2] These include blunt trauma to the chest as well as post-traumatic causes such as after a tracheostomy and long-term intubation.[2] Other causes include extrinsic compression due to a tumour or goitre around the tracheobronchial tree.[6] Case studies have reported amyloidosis and rickets being associated with TBM.[7 8] COPD seems to be the most common risk factor for patients with the acquired form of the condition, as many studies have reported an association between the two conditions; however, it is not yet fully understood why this is the case.[2 9]

The true incidence of TBM is not very clear as studies have been based on selected populations rather than the population at large.[2] The most recent study on the incidence of airway collapse was reported in Japan,[10] where the rate of airway collapse of >50% was shown in 542 of 4283 patients (12.7%) with pulmonary disease who underwent bronchoscopy. Seventy-two per cent of these patients were aged between 50 and 80 years. Earliest studies in the 1970s by Jokinen (23%) also showed that the disease was common among middle-aged and elderly populations.[11]

Diagnosis can be made through dynamic biphasic inspiratory/expiratory CT and bronchoscopy. Bronchoscopy is currently the investigation of choice to diagnose TBM.[2] Several studies have used a threshold mark of more than or equal to 50% collapse of the airways as diagnostic of TBM when CT or bronchoscopy is used.[6] However, there have been studies that have suggested different thresholds, such as by Stern *et al*[12] (70%). Treatment options for the condition include conservative management, continuous positive airway pressure (CPAP), stent placement and surgery.[2 3 13]

Conservative/supportive management is employed in patients with mild symptoms. It includes helping the patient stop smoking cigarettes, breathing and relaxation techniques, and treating any respiratory infections.[1 5 14] CPAP can be used to maintain a free airway and help drainage of secretion.[15 16] It can improve obstruction of the airways during expiration and reduce pulmonary resistance.[15 16] Stents are effective at treating the symptoms temporarily. They are used to maintain the patency of the airway, with two types being the most common: metal and silicone. Nonetheless, stents seem to have many complications, which include their migration (silicone) or fracture (metal), mucus plug formation and higher risk of granuloma formation.[14] Hence stents seem to be a temporary measure to help stabilise the airway and relieve symptoms before surgery can be performed.[2 14] Surgery seems to be the most effective form of treatment for TBM.[1 5 13 14] The aim of surgery is to strengthen the walls of the trachea and prevent their excessive collapse.[1 5 13] The most common surgical approach currently is tracheobronchoplasty, a technique where the posterior membranous wall of the trachea is reinforced using different grafts such as Marlex or PTFE.[1 13 14] Tracheobronchoplasty was first described by Herzog and Nissen to improve airflow obstruction using bone grafts.[13] Since then, many materials have been used including polypropylene and high-density polyethylene mesh, among others, to plicate the trachea and provide rigidity.[1 13 14] Niranjan and Marzouk[1] used a PTFE (Teflon) patch with good outcomes in patients with the condition.[1]

Learning points

- Flat trachea syndrome is a rare disease characterised by excessive prolapse of the airway, typically occurring during expiration.
- It is increasingly being recognised as a potential cause of chronic cough, dyspnoea and recurrent infection. It can be fatal if not treated appropriately.
- Flat trachea syndrome can be a challenging condition to diagnose, hence patients presenting with refractory asthma or COPD that is worsening should alert physicians towards suspecting and excluding other causes of an obstructive airway, and it should include this condition as part of their clinical work up.
- Surgery for the condition has shown to be successful so far, hence it is important that these patients are recognised early and managed appropriately.

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Competing interests None.

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