Tonsillar cyst: an unusual cause of ventilatory failure

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DECLARATIONS

Competing interests
None declared

Funding
None

Ethical approval
Written informed consent to publication has been obtained from the patient or next of kin

Guarantor
JL

Contributorship
JL wrote most of the manuscript; MR provided the diagrams and undertook lung function testing; RM treated the patient and supervised the writing of the article

Acknowledgements
None

Reviewer
Mat Daniel

This case demonstrates the importance of considering curable causes of ventilatory failure despite its common occurrence in patients with chronic obstructive pulmonary disease (COPD).

Introduction

COPD is a common disease that places a considerable burden on society; estimates based on disability life-years (DALY) suggests that COPD will become the fifth leading cause of disability worldwide by 2020.[1] The term 'exacerbation of COPD' describes an acute deterioration in the patient's baseline symptoms (notably dyspnoea, cough or sputum production) and a necessity to change their usual medications.[2] 'Exacerbations' with accompanying ventilatory failure in patients with severe COPD are a frequent cause of admission to hospital. We report an individual admitted in ventilatory failure who was initially diagnosed as having an exacerbation of COPD, but examination and investigations in clinic revealed that a tonsillar cyst was causing obstructive sleep apnoea, which upon removal led to substantial improvements.

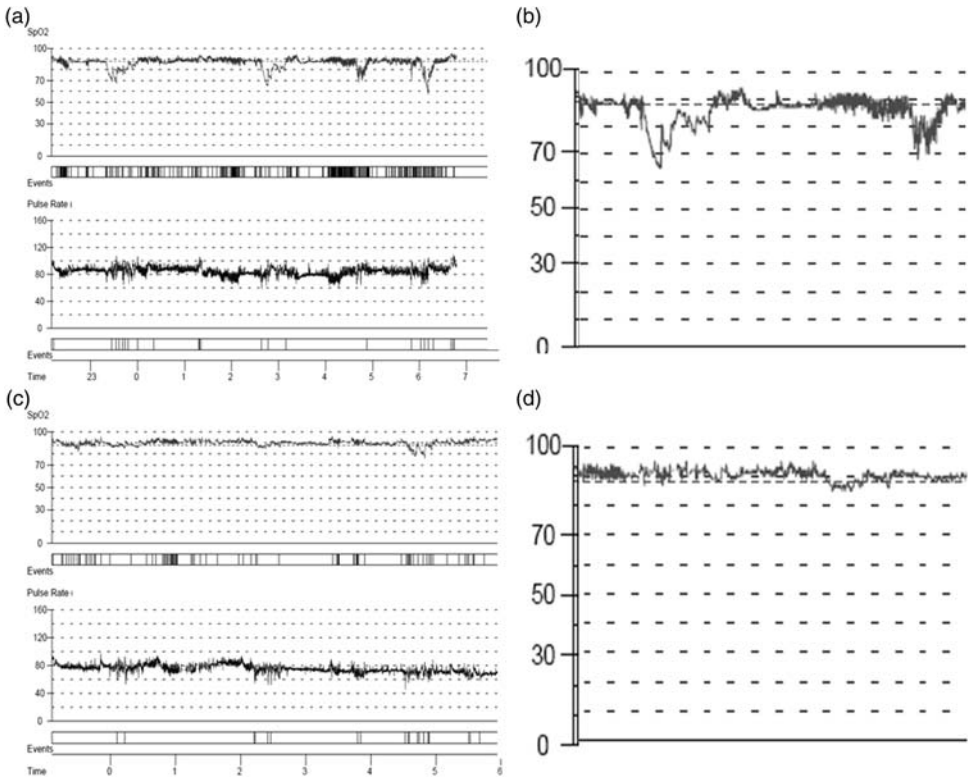
Case report

A 64-year-old woman was referred to the COPD clinic. She had a 22-pack–year smoking history and had recently presented as an emergency with drowsiness. She was found to have a significant ventilatory failure causing respiratory acidosis (FiO_2 0.4 pH 7.206 (normal range 7.35–7.45), $paCO_2$ 12.18kPa (4.67–6kPa), paO_2 16.69kPa (9.3–14kPa), HCO_3 35.4mmol/L (21–32mmol/L), base excess (BE) 3.8 mmol/L (+/– 2.5mmol/L). She required intensive care with bi-level positive pressure ventilatory support, nebulized bronchodilators, corticosteroids and antibiotics. Prior to discharge, her arterial blood gas showed pH 7.416, $paCO_2$ 6.68kPa, paO_2 7.89kPa, HCO_3 31.5mmol/L, BE 5.6 mmol/L breathing air. The working diagnosis from the admitting team was acute-on-chronic ventilatory failure due to pneumonia and COPD. On review in clinic it was noted that her CTPA during admission revealed only moderate upper-lobe centrilobular emphysema and no pulmonary emboli, with only minor consolidation in the lingula.

Pulmonary function tests demonstrated an obstructive pattern: FEV_1 0.84l (33.8% of predicted), FVC 2.2l (75.2%), VC 2.34l (76.3%), FEV_1/VC 35.77% (46.5%), KCO 1.19mmol/min/kPa/L (80.4%), TLCO 4.82mmol/min/kPa (61.0%) compatible with COPD. On detailed questioning in clinic, she had never had any significant problems with breathlessness or exacerbations but felt she lacked energy. It was felt that her chronic ventilatory failure may not be solely due to her COPD and on further examination of her oropharynx there was an enlarged left tonsil. Her body mass index was 30.4 kg/m^2. She underwent nocturnal oximetry which showed evidence of significant nocturnal hypoventilation and a desaturation index of 41.7 per hour (Figures 1a and 1b). She had a hypoxic challenge test as she wished to undertake an aircraft flight but this was terminated at 9 minutes due to oxygen desaturation to 82% while breathing FiO_2 0.15 and her arterial blood gas post-test showed a pH 7.365, $paCO_2$ 6.18kPa and paO_2 5.68kPa. It was felt that she had obstructive sleep apnoea and ventilatory failure due to the enlarged tonsil with a lesser contribution from her COPD. She underwent urgent tonsillectomy which revealed benign lymphoid hyperplasia on histology. Postoperatively, her symptoms improved dramatically; her Epworth score fell from 22 preoperatively to 0. She and her husband reported a significant improvement in

Figure 1

(a) Overnight pulse oximetry measured prior to tonsillectomy. Saturations are the top trace and pulse rate is the bottom trace. A desaturation event was characterized as a drop in saturations by at least 4% for a minimum duration of 10 s. A pulse event was a change in rate by at least 6 bpm for a minimum of 8 s. Adjusted index SpO2 41.7, minimum SpO2 57, average SpO2 83.7, average pulse rate 84.7, low pulse rate 58, Epworth Score 22; (b) Measured oxygen desaturation prior to tonsillectomy between 02:00 and 05:00; (c) Overnight pulse oximetry measured 6 months post-tonsillectomy. Saturations are the top trace and pulse rate is the bottom trace. A desaturation event was characterized as a drop in saturations by at least 4% for a minimum duration of 10 s. A pulse event was a change in rate by at least 6 bpm for a minimum of 8 s. Adjusted index SpO2 11.9, minimum SpO2 76, average SpO2 87.8, average pulse rate 74.9, low pulse rate 45, Epworth Score 0; (d) Measured oxygen desaturation 6 months post-tonsillectomy between 01:00 and 03:00



her mood and energy, reflected in a marked improvement in her desaturation index to 11.9 per hour (Figures 1c and 1d), and arterial blood gases improved to paO_2 10.28, $paCO_2$ 4.85kPa and pH 7.412. Furthermore, she was able to complete her hypoxic challenge test with improved post-test arterial blood gases of pH 7.464, paO_2 8.3kPa, $paCO_2$ 4kPa, HCO_3 20.9mmol/L, BE −1.7 mmol/L. Pulmonary function tests showed little change six months postoperatively; FEV_1 0.88l (35.9% of predicted), FVC 1.14l (73.6%), indicating that her clinical improvement was not due to recovery in her lung function.

Discussion

There are many contributing factors to chronic ventilatory failure in COPD such as airflow obstruction, reduced gas exchange, respiratory muscle weakness and obesity hypoventilation.[3] Our patient with a history of COPD, presented with

acute-on-chronic ventilatory failure with decompensation due to a mild pneumonia. On review in a specialist COPD clinic, it was thought she had chronic ventilatory failure but her COPD did not sufficiently account for this. Detailed examination suggested that a tonsillar cyst was causing obstructive sleep apnoea and hypoventilation, which on removal led to resolution of symptoms and abnormalities in overnight oximetry, arterial blood gas and response to hypoxic challenge.

As a patient with COPD, being admitted to hospital led to an initial treatment of having an 'exacerbation' of her disease without further investigation of possible precipitants. Viral and bacterial pathogens, such as rhinoviruses and haemophilus influenzae, respectively, are common, and pollutants such as ozone, sulphur and nitrous oxide may also trigger exacerbations.[4]

Basic clinical history, examination and investigations will help with separating infectious exacerbations with differential diagnoses which are varied, and commonly include pneumonia, pneumothorax, pulmonary embolus and cardiac failure.[5] The arterial blood gas can be helpful in identifying patients with chronic ventilatory failure by the presence of a disproportionately raised bicarbonate level and raised arterial carbon dioxide level compared to a relatively normal pH. Common causes of chronic ventilatory failure are listed in Table 1.

Obstructive sleep apnoea is a clinical syndrome that describes the presence of episodes of apnoea and hypopnea with symptoms of daytime somnolence. The prevalence of this condition is estimated to be at about 4% of men and 2% of women in a cohort study of 402 middle-aged adults.[6] Co-existing COPD and obstructive sleep apnoea is known as overlap syndrome and is present in 1% of patients over the age of 40 years.[7] The aetiology of this is poorly understood. In adults, a range of surgical approaches, including tonsillectomy, uvulopalatal pharyngoplasty and tracheostomy in very severe cases have been advocated, but the role of surgery in the management of obstructive sleep apnoea remains uncertain.[8] Long-term treatment with domiciliary continuous positive airway pressure remains the mainstay of management because of its beneficial effects on daytime symptoms.[9]

Table 1
Common causes of chronic ventilatory failure

Category	*Examples*
Pulmonary	COPD
	Bronchiectasis
	Cystic fibrosis
Chest wall restriction	Obesity hypoventilation
	Scoliosis
	Thoracoplasty
Neuromuscular disease	Motor neurone disease
	Myotonic dystrophy
	Muscular dystrophy
Neurological disease	Spinal cord trauma
	Central hypoventilation

Conclusion

This case reminds us of the importance of assessing for causes of exacerbations and reversible causes of chronic ventilatory failure even in apparently common clinical scenarios, as identification of them can lead to a cure.

- Exacerbations of COPD are defined as an acute and sustained deterioration in a patient's symptoms that are beyond the normal day-to-day variability and have many possible causes.
- Chronic ventilatory failure is associated with a raised arterial carbon dioxide and bicarbonate with a relatively mild acidaemia.
- Causes of chronic ventilatory failure include COPD, bronchiectasis and extra-pulmonary conditions such as neuromuscular disorders.
- Long-term treatment with domiciliary continuous positive pressure airway support is the mainstay of management of patients with obstructive sleep apnoea.

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