


Reversible platypnoea–orthodeoxia syndrome in post-tuberculosis bronchial stenosis

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Keywords

Atelectasis, bronchial stenosis, orthodeoxia, platypnoea, ventilation–perfusion mismatch.

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Abstract

Bronchial stenosis is known to complicate endobronchial tuberculosis despite medical therapy. It is often associated with dyspnoea. In severe cases, bronchial stenosis results in airflow obstruction, impaired secretion clearance, and can lead to respiratory failure. We present an unusual observation of platypnoea–orthodeoxia syndrome in a young woman with acute atelectasis due to post-tuberculosis bronchial stricture. Imaging revealed complete middle and right lower lobe atelectasis with a partially aerated right upper lobe. In the sitting posture, there was positional worsening of dyspnoea associated with an increase in the alveolar–arterial oxygen gradient and shunt fraction. The likely mechanism was due to gravitational difference in ventilation–perfusion matching. The platypnoea–orthodeoxia syndrome was reversible following balloon dilatation of the bronchial stenosis and expansion of the collapsed lung.

Introduction

Bronchial stenosis is known to complicate endobronchial tuberculosis (TB) despite medical therapy [1]. It is often associated with dyspnoea and can result in respiratory failure if left untreated. We present an unusual observation of platypnoea–orthodeoxia syndrome (POS) in a case of acute lung collapse secondary to post-TB bronchial stricture, which was reversible following successful bronchoscopic intervention.

Case Report

Our patient was a young and healthy woman. She had a history of pulmonary TB treated medically (2HREZ/4R3H3). The TB infection was complicated by right main bronchus (RMB) stricture requiring endoscopic balloon dilatation 6 months prior to the current presentation.

The patient presented to the Emergency Department with acute dyspnoea associated with right-sided back pain. The symptoms were preceded by a 3-day history of non-productive cough without constitutional symptoms. Clinical examination revealed right-sided reduced chest expansion, dullness to percussion, reduced air entry, and vocal resonance. She was in mild respiratory distress and

was hypoxaemic despite 35% supplemental oxygen (see Table 1). Chest X-ray showed complete “white out” of the right lung field with an ipsilateral deviated trachea (see Fig. 1A). Computed tomography (CT) scan of the thorax (pulmonary angiogram protocol for chest pain and raised serum d-dimer) showed almost complete atelectasis of the right lung due to a RMB stricture with no evidence of a pulmonary embolism. The right upper lobe remained partially aerated. Contrast material was present within the pulmonary vessels and in the atelectatic lobes (see Fig. 1C and D).

We noticed that the patient preferred to lie completely flat, and there was associated improvement in oxygen saturation (SpO_2) in the supine posture (see Table 1), allowing the patient to be weaned off supplemental oxygen, albeit mildly tachypnoeic (SpO_2 was 93% on room air; respiratory rate 24/min). However, when rising to a sitting position, she immediately became more tachypnoeic, and SpO_2 rapidly declined to 79%. SpO_2 quickly returned to baseline when she resumed the supine posture.

We proceeded to perform rigid bronchoscopy under general anaesthesia, which confirmed pin-point stenosis at the proximal RMB. Serial balloon dilatation improved RMB lumen size and allowed for expansion of the

Table 1. Arterial blood gas results pre- and post-balloon dilatation of right main bronchus stenosis.

	Pre-procedure	Pre-procedure	Post-procedure
Posture	Sitting	Supine	Semi-recumbent
Supplemental oxygen	VM 35%	NP 4 L/min	NP 2 L/min
pH	7.38	7.40	7.37
PaCO ₂ , mmHg	28	27	34
PaO ₂ , mmHg	57	88	125
Bicarbonate, mmol/L	18	17	21
Base excess	-8	-6	-5
SaO ₂ , %	89	97	99
Haemoglobin, g/dL	14.3	14.3	14.3
A-a O ₂ gradient*, mmHg	158	114	25
Shunt fraction [†] , %	33.2	12.6	1.5

*Calculated alveolar-arterial oxygen gradient, A-a O₂ gradient = $FiO_2 (Patm - PH_2O) - PaCO_2/0.8 = FiO_2(713) - PaCO_2/0.8$.

[†]Calculated shunt fraction = $(CcO_2 - CaO_2)/(CcO_2 - CvO_2)$ [2] where pulmonary capillary and mixed venous oxygen content were estimated.

VM, venturi mask; NP, nasal prong; PaCO₂, partial pressure of carbon dioxide in arterial blood; PaO₂, partial pressure of oxygen in arterial blood; SaO₂, oxygen saturation of haemoglobin in arterial blood; FiO₂, fraction of inspired oxygen (estimated additional 3% above atmospheric FiO₂ for every 1L/min increment in supplemental oxygen flow via nasal prong); CcO₂, pulmonary capillary oxygen content; CaO₂, arterial oxygen content; CvO₂, mixed venous oxygen content.

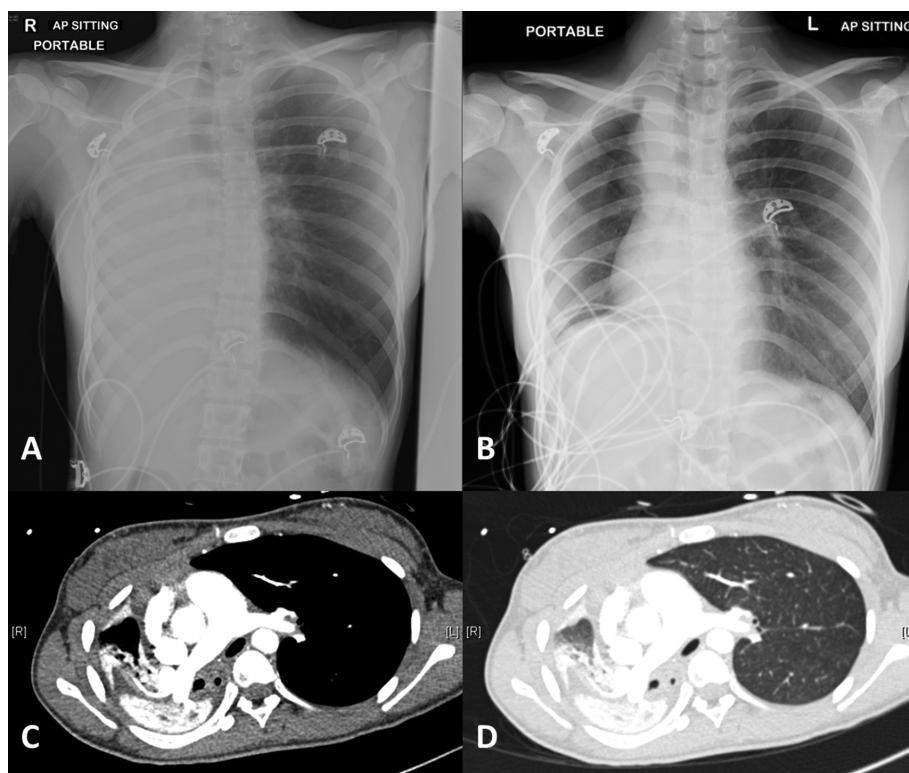


Figure 1. Chest X-ray and contrasted computed tomography (CT) thorax (pulmonary angiogram protocol) images. (A) and (B) Chest X-ray showing “white out” of the right lung field with an ipsilateral deviated trachea and expansion of the right lung following balloon dilatation of the right main bronchus. (C) and (D) Representative cuts of CT showing contrast enhanced pulmonary vessels in collapsed lobes, partially aerated right upper lobe, and otherwise normal left lung (pre-procedure).

atelectatic lung. Her symptoms and POS resolved (SpO₂ 99–100% in all postures) within an hour post-procedure. Calculated shunt fraction normalized (see Table 1). The patient was discharged from hospital 2 days post-procedure and planned for bronchial stenting on a later admission.

Discussion

POS is a rare clinical condition characterized by positional dyspnoea, oxygen desaturation >5%, or a decrease of blood partial pressure of oxygen >4 mmHg from supine to an upright posture [3]. Less than 250 cases have been reported in the literature. The majority of POS is due to

intra-cardiac shunt; patent foramen ovale and atrial septal defect are the two most commonly reported intra-cardiac shunts associated with POS. Pulmonary causes can be because of pulmonary anatomical shunts (e.g. pulmonary arteriovenous malformation or hepatopulmonary syndrome) or even rarer pulmonary parenchymal disease; these collectively contribute to approximately 13% of cases [3]. The underlying pathophysiological mechanism of POS is a right-to-left shunt. The reason for augmentation of the shunt is different depending on the underlying cause; it may be due to increased directional flow across the inter-atrial shunt or increased regional ventilation-perfusion (V/Q) mismatch [3,4]. In pulmonary disease, POS has been postulated to be due to increased dead space and physiological shunt in the erect posture [3].

To our knowledge, the observation of POS in acute atelectasis secondary to post-TB bronchial stricture has never been reported. The availability of arterial blood gases measured in various postures along with contrasted CT thorax allowed for deduction of the possible pathophysiology of POS in this case. CT pulmonary angiogram findings of contrast media within the collapsed lobes suggest continuing perfusion of the area without ventilation, resulting in right-to-left shunting. Although we know that hypoxic vasoconstriction occurs to divert blood from an under-ventilated lung, the mechanism is likely imperfect with persistent shunting of blood through the collapsed lung, similar to the finding of hypoxaemia in an anaesthetized individual undergoing one-lung ventilation [5]. In addition, there was no obvious increase lucency of lung parenchyma on the right when compared to the normal left lung (to suggest hypoxic vasoconstriction) on CT imaging.

We hypothesize that the orthodeoxia in this case was due to the gravitational increase in lower zone perfusion, resulting in increased shunting of blood through under-ventilated areas with concomitant increased V/Q mismatch in the partially aerated upper lobe (dead space or Zone 1 phenomenon) [3,5]. However, it is difficult to delineate the exact contribution of each mechanism and whether there was any component of anatomical shunt

in the absence of 100% oxygen breathing [2]. Partial improvement in oxygenation following oxygen supplementation suggests a definite component of V/Q mismatch. Bubble-contrast echocardiography and nuclear perfusion study in different postures would be ideal to confirm the above hypothesis, but the clinical urgency of the interventional procedure superseded further investigations. We think that the presence of an anatomical shunt is unlikely given normal shunt fraction following expansion of the collapsed lung.

In conclusion, we reported an interesting and unusual observation of POS in a case of acute atelectasis due to post-TB bronchial stricture. POS probably occurs in other causes of lung atelectasis, but it may be masked by the use of supplemental oxygen. Nonetheless, the case illustrates that post-TB bronchial stricture can result in significant physiological impairment in an otherwise healthy individual. Timely bronchoscopic intervention allows for normalization of physiology and, in this case, POS.

Disclosure Statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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