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Case report

Platypnoea–orthodeoxia after left total knee replacement

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SUMMARY

Platypnoea–orthodeoxia syndrome (POS) is a rare disorder characterised by both dyspnoea (platypnoea) and arterial desaturation (orthodeoxia) in the upright position, with improvement in the supine position. We report an unusual case in which an 82-year-old woman developed severe hypoxaemia with POS after left total knee replacement. A significant difference in alveolar–arterial blood gas oxygen tension was demonstrated, and hypoxaemia failed to respond to 100% oxygen supply. A patent foramen ovale with a right-to-left shunt was evident on transoesophageal echocardiogram employing colour Doppler and agitated normal saline studies. Interestingly, the patient's symptoms resolved within 6 months with ongoing chest physiotherapy, without surgical or medical intervention.

BACKGROUND

Platypnoea–orthodeoxia syndrome (POS) is a rare disorder that is characterised by both dyspnoea (platypnoea) and arterial desaturation (orthodeoxia) in the upright position, with improvement in the supine position.[1] This condition has been reported in association with pulmonary, hepatic and cardiac diseases. We report an unusual case of POS associated with patent foramen ovale (PFO) after left total knee replacement (TKR). Without a high degree of suspicion, diagnosis may be delayed, and POS should be initially suspected in patients with positional hypoxaemia and dyspnoea. The disorder should be biochemically confirmed by arterial blood gas (ABG) analysis, particularly, by calculation of the alveolar-to-arterial (A–a) oxygen gradient, which is a standard measure of oxygenation; this measure corresponds to the difference between the amount of oxygen in the alveoli (ie, the alveolar oxygen tension (PAO_2)) and the amount of oxygen dissolved in the plasma (PaO_2). Calculating the A–a difference helps in narrowing the differential diagnosis, as hypoxaemia with a wide A–a gradient that is difficult to correct with 100% oxygen supplementation can be explained only by the presence of a right-to-left shunt.

CASE PRESENTATION

An 82-year-old woman was electively admitted for left TKR. Her medical history included acute myocardial infarction 10 years prior, as well as hypertension, left total hip replacement and a stable, 4.5 cm ascending aorta aneurysm.

She was taking the following medications: telmisartan 40 mg once a day, aspirin 100 mg once a day, metoprolol 50 mg two times per day, rosuvastatin 10 mg once a day, rabeprazole 20 mg and lercanidipine 10 mg. Her preoperative anaesthesia assessment included an ECG which showed a right bundle branch block, as well as a chest X-ray and transthoracic echocardiogram, which showed borderline left atrial diameters, normal left ventricular size and systolic function, and an estimated ejection fraction >50%. There were no overt regional wall motion abnormalities. Mild degenerative changes of the aortic and mitral valves were observed, along with mild aortic valve insufficiency. The ascending aorta was dilated (approximately 5 cm). There were no respiratory symptoms, and spirometry did not show significant abnormality.

The patient underwent TKR. On the first day after the operation, she developed dense right arm weakness and left homonymous hemianopia. There were no other obvious focal neurological deficits. CT and brain MRI showed innumerable T2 FLAIR hyperintensities, demonstrating high diffusion-weighted imaging signals that were consistent with multifocal acute infarcts. These were predominantly cortically/subcortically distributed in both the right and left cerebral hemispheres within the frontal, parietal and occipital regions; additionally, there was a small focus within the left cerebellar hemisphere, favouring a central embolic cause. The patient was managed according to an acute stroke unit pathway, moved to a bed with telemetry monitoring and clopidogrel was added to her medications.

Following surgery, the patient complained of increasing shortness of breath and was noted to be hypoxic on room air, up to 86%. A physical examination did not reveal any other significant findings. Her blood tests remained within normal ranges. CT pulmonary angiogram did not show pulmonary embolism; however, it showed left lower lobe atelectasis and evidence of right ventricular strain. The contrast was seen refluxing into the hepatic veins, which was consistent with right atrial dysfunction. During admission, progressive dyspnoea at rest prompted several emergency calls and subsequent admission to a high-dependency bed. Extensive investigations were undertaken to determine possible causes of her symptoms, including sepsis, pulmonary embolism, pulmonary hypertension and heart failure. Blood tests remained unchanged, apart from a transient rise in serum troponin, which was treated as NON-ST elevation myocardial infarction

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Table 1 Oxygen saturation patterns with varying postures

Posture	Saturation with 2 L O_2 via nasal prong (%)	Saturation with 12 L O_2 via non-rebreather mask (%)
Lying in bed	89–93	93–95
Standing	77–80	85

for medical management only. CT of the pulmonary artery and transthoracic echocardiography were repeated, which showed normal right ventricular size with moderate systolic dysfunction, moderate right ventricular hypertrophy and significant pulmonary hypertension. No intracardiac thrombus was visualised. Two weeks postsurgery the patient achieved marginal improvement, requiring 2 L of oxygen via nasal prong; therefore, she was transferred to the rehabilitation ward.

Interestingly, in the rehabilitation ward, emergency calls were activated repeatedly, daily in the morning for persistent hypoxia; her symptoms and oxygen requirement worsened when she sat in a chair, compared with lying flat in bed; moreover, standing caused a significant drop in oxygen saturation, which prevented her participation in the physiotherapy programme (table 1).

INVESTIGATIONS

ABG analysis confirmed hypoxaemia, and the A–a oxygen gradient increased to 38.6 (while the normal value for her age is 19.3). Despite 100% supply of oxygen via a non-rebreathing mask, a minimal improvement was demonstrated with repeated ABG analysis (table 2). A ventilation–perfusion (V/Q) scan was normal and secondary causes of pulmonary hypertension were negative. The telemetry did not detect atrial fibrillation; repeated CT aortography ruled out aortic dissection and no significant pathology was observed in the arterial phase of the abdomen aside from a trace of free fluid located dependently/posteriorly in the presacral space. Venous Doppler ultrasound of the lower limbs assessed the deep and superficial veins from the calf to the groin. No ultrasonic signs of an acute deep veins thrombosis (DVT) were noted in the calf, popliteal fossa or thigh veins. The presence of a wide A–a gradient with a minimal response to 100% supply of oxygen prompted us to suspect a right-to-left intracardiac shunt. A transoesophageal echocardiogram revealed a PFO, with a right-to-left shunt confirmed by a bubble study (figure 1).

TREATMENT

The patient continued inpatient physiotherapy and occupational therapy rehabilitation programme for 2 months, with significant improvement, and was discharged to a healthy ageing unit on warfarin and 1.5 L of oxygen via nasal prong, awaiting cardiology follow-up with possible intervention.

OUTCOME AND FOLLOW-UP

Our patient's symptoms exhibited remarkable improvement with a continuous rehabilitation programme and remained

Table 2 Arterial blood gas analyses before and after oxygen supply

	On room air	On 100% oxygen supply
Temperature	36.5°C	36.5°C
PCO_2	25 mm Hg	22 mm Hg
PO_2	65 mm Hg	75 mm Hg
Oxygen saturation	85%	95%
A–a gradient	38.6 mm Hg	539 mm Hg

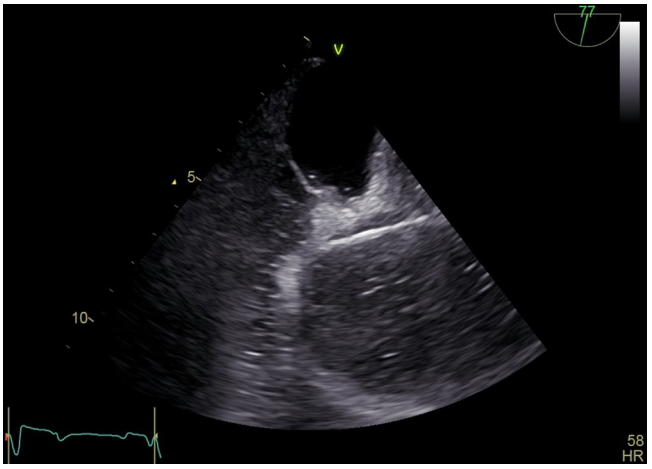

Figure 1 A transoesophageal echocardiogram demonstrating strongly positive microbubble study.

satisfactory at the regular 3-month outpatient visit. She achieved good recovery from stroke without significant residual neurological deficit and was weaned from oxygen and discharged from the healthy ageing unit to her home, where she lived alone and was independent. To our surprise, at 6 months after surgery, her oxygen saturation was 91%–93% on room air without a significant postural drop. The patient remained asymptomatic at her annual cardiology follow-up.

DISCUSSION

POS has been reported in association with PFO and has been described with intrapulmonary shunting, as well as with disorders such as pericardial effusion, constrictive pericarditis, emphysema, amiodarone pulmonary toxicity, pneumonectomy, abdominal disorders (cirrhosis or ileus) and vascular disorders (abdominal aortic aneurysm).[1] Two components are required.[2]

The first component is an interatrial shunt (such as a PFO, an atrial septal defect or a fenestrated atrial septal aneurysm) or intrapulmonary shunting, as in hepatopulmonary syndrome and pulmonary arteriovenous malformations, which may occur in patients with cirrhosis.

The second component is a functional component that promotes abnormal shunting when the patient rises from a recumbent to an upright position. This could be mediated by a deformity in the atrial septum (that promotes shunt flow) and/or the right atrium, which increases streaming of blood from the inferior vena cava through the defect. An elevation in right atrial pressure causing right-to-left shunting is usually required, although blood may flow from right to left even when the right atrial pressure is normal, which typically occurs with persistent eustachian valves.[1]

PFO is thought to be present in 25% of the population; however, clinically significant right-to-left shunting is rare and requires raised right atrial pressure.[3] Riddles *et al*[4] reported a case of POS following laparoscopic cholecystectomy. Other case studies also indicate that surgery is a common precipitant for the development of POS.[5–8] While various mechanisms have been proposed for a transient elevation in right heart pressure, the underlying mechanics that maintain the shunt are not well understood.[7] Possible explanations for mechanisms that maintain postoperative right-to-left shunting remain speculative. Riddles *et al*[4] suggested that 'a common postoperative complication of atelectasis may increase pulmonary vascular resistance and cause a V/Q mismatch'. While the patient in the present

case study had left lower lobe atelectasis, it is unlikely to be the only explanation. Literature describing POS is scarce outside the realm of intra-abdominal or intrathoracic surgery where surgery has been deemed the precipitant.[4 8] The proposed mechanism in abdominal laparoscopic surgeries involved stretching of the interatrial septum, resulting from the dilated aortic root; upright positioning accentuated this stretching, with possible compression on the right atrium.[7] Our patient had a stable 4.5 cm ascending aorta aneurysm and an aortic root of 35 mm, with the presence of PFO, which represented the anatomical component. Advanced age represents an added risk factor[7] which may be due to a decline in connective tissue compliance, leading to postoperative anatomical distortion, as well as a change in the cardiac axis leading to a change in shunting.[4]

Another relevant issue in our patient is the presence of multifocal acute infarcts, which are predominantly cortically/subcortically based, favouring a central embolic cause. The distribution of these infarcts is not consistent with small vessel disease. The lack of intracardiac thrombus or atrial fibrillation directed us towards the presence of a PFO and a right-to-left shunt. It is difficult to attribute the POS to one pathology in this patient. However, the patient exhibited rapid postoperative improvement; this indicates a temporary and reversible insult that promoted the shunt dynamics. Two transient factors were involved in this case: (1) haemodynamic changes during surgery, which can increase myocardial oxygen demand resulting in ischaemia to the myocardium[9] and affecting right ventricular compliance; and (2) left lower lobe atelectasis, which resolved within 2 weeks postsurgery and coincided with a marked reduction in oxygen requirement. Therefore, we postulate that these two factors, in addition to the presence of an ascending aorta aneurysm and the patient's advanced age, caused changes in intracardiac blood flow dynamics. These changes, in association with congenital PFO, may have caused the syndrome in this patient.

Closure of the anatomical defect may not be necessary in all patients, as our patient participated in a rehabilitation programme with excellent results and remains in a satisfactory state at the 6-month follow-up, with an oxygen saturation >93% on room air; she also showed no significant change with posture. Hence, treatment of the causative pathology, such as atelectasis in this case, which resolved with chest physiotherapy and a period of recovery from surgery, rendered POS a temporary, reversible complication after surgery.

Learning points

- Platypnoea–orthodeoxia syndrome (POS) is a rare condition that may cause disabling postural hypoxia in the upright posture; it shows improvement in the supine position and minimal change with supplemental oxygen.
- A common postoperative complication of atelectasis may increase pulmonary vascular resistance and cause a ventilation–perfusion mismatch, and this complication in association with congenital patent foramen ovale may cause POS.
- In hypoxaemia, arterial blood gas and A–a gradient calculation can narrow the differential diagnosis; when a wide A–a gradient and hypoxaemia that does not respond to 100% oxygen are detected, a right-to-left shunt should be suspected. While CT of the pulmonary artery can rule out pulmonary embolism and other causes of extracardiac shunting, a bubble study echocardiogram must be the next step in the diagnostic investigation.
- Closure of the anatomical defect may be required if treatment of the causative pathology fails or is not possible.

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Contributors TA suggested the diagnosis and encouraged the investigations. He did the majority of the writing of the article. BJ contributed in writing the case and was involved directly in the care and management of the patient and followed up with the patient. FE gave general guidance and supported in writing the case discussion. GAA provided oversight. All authors gave their final approval before submission.

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Reminder of important clinical lesson