Case report

Intermittent cardiac shunt mimicking pulmonary embolism and COPD

K. SCHWARZ1, I. DU RAND2, S. AUSTIN3 and H. ROUTLEDGE1

From the 1 Cardiology Department, Worcestershire Royal Hospital, Charles Hastings Way, WR5 1DD, 2 University Hospital Birmingham, Selly Oak, Birmingham, B29 6JD and 3 Respiratory Department, Worcestershire Royal Hospital, Charles Hastings Way, WR5 1DD, UK

Address correspondence to Dr K. Schwarz, Department of Cardiology, Worcestershire Royal Hospital, Charles Hastings Way, Worcester, Worcestershire WR5 1DD, UK. email: konstantin.schwarz@gmx.net

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A 79-year-old ex-smoker was admitted because of cyanosis, following a recent chest infection. He described longstanding mild exertional dyspnoea, attributed to chronic obstructive pulmonary disease (COPD), for which he was prescribed inhalers and long-term steroid therapy. His co-morbidities included hypertension, renal impairment, polymyalgia and three repairs of an infrarenal aortic aneurysm.

He appeared cushingoid and cyanosed, with a respiratory rate of 22/min, oxygen saturations 72%, blood pressure 95/50 mmHg and heart rate 95/min. His arterial blood gas analysis revealed pH 7.45, pO2 5.1 and pCO2 4.9 mmHg. Chest X-ray and CT pulmonary angiogram revealed mild emphysematous change, but no evidence of pulmonary embolism (PE). As smaller subsegmental emboli could not be excluded, anticoagulation was initiated. Transthoracic echocardiography (TTE) was normal other than a dilated aortic root. Notably, both right heart function and pulmonary artery pressure were normal.

Throughout his admission, frequent emergency calls were made because he was hypoxic and unresponsive to increased inspired oxygen. Saturations fluctuated between 72 and 99% on air, initially attributed to inaccurate pulse oximetry with postural hypotension and subsequently to his presumed COPD. Later intermittent central cyanosis with hypoxia in the upright position, which recovered when recumbent aroused suspicion of the platypnoea-orthodeoxia syndrome (POS) (Figure 1). Further investigation with tilt transoesophageal echocardiography (TOE) confirmed an intra-cardiac right-to-left shunt and a dilated thoracic aorta. The posturally dependent shunt was through a patent foramen ovale (PFO) within a mobile, aneurysmal interatrial septum (Figure 2).

Platypnoea and orthodeoxia describe breathlessness and hypoxia associated with upright posture. The syndrome is caused by intermittent intra-cardiac, or less frequently intrapulmonary, right-to-left shunting.

The extraordinary finding is of a large right-to-left shunt occurring in the absence of increased right heart pressures. The ‘water flows uphill’ if two conditions coexist: an ‘anatomical’ inter-atrial communication [PFO or atrial septal defect (ASD)] and a ‘functional’ component that causes deformity of the atrial septum with upright posture. The deformation, often a displacement of the septum horizontally, allows preferential flow of blood from the inferior vena cava to the left atrium. As in our patient this may be caused simply by dilatation of the ascending aorta. Other functional components allowing posturally dependent shunting in the
absence of an inter-atrial pressure gradient are a persistent Eustachian valve, tilting of the heart after pneumonectomy, kyphoscoliosis, a loculated pericardial effusion, constrictive pericarditis or diaphragmatic paralysis.\textsuperscript{4} Platypnoea-orthodeoxia caused by intrapulmonary right-to-left shunting is described with arteriovenous malformations, the hepatopulmonary syndrome and with an increased postural ventilation–perfusion mismatch seen in COPD and autonomic neuropathy (with postural hypotension). More than one factor may be responsible in a single patient. POS is rarely diagnosed, but considering that PFO is found in 27\% at autopsy\textsuperscript{5} and that orientation of the septum may be modified by age and hypertension-related aortic root dilatation, its prevalence may be underestimated.\textsuperscript{2}

As a young man, when our patient ran as National Service 100-yard-champion, his PFO was an asymptomatic normal variant. With increasing age and enlargement of his aorta, the PFO became a rogue accomplice in the aetiology of his profound hypoxia. Detailed study of

**Figure 1.** Cushingoid features and ecchymoses due to long-term steroids and anticoagulation; central cyanosis in upright position.

**Figure 2.** TOE showing mobile aneurismal inter-atrial septum and dilated aortic root (A, B). Recumbent posture with absence of inter-atrial shunt (C) and upright posture (tilt table) inducing right-to-left shunt through PFO in the mobile atrial septum (D).
patient’s previous notes showed three interesting observations. Two years earlier, while recovering from an aortic aneurysm repair, the patient developed sudden respiratory failure and a CTPA performed for presumed PE was negative. The patient did not admit to any symptoms consistent with a clinical diagnosis of COPD and a 6 min walk test suggested possible orthodeoxia behaviour already, few months prior to his acute admission.

It is likely that occasional patients are wrongly treated for PE or COPD. Goddart described 11 patients undergoing PFO/ASD closure for symptomatic hypoxia with normal pulmonary artery pressures. In nine cases, the initial investigations were for pulmonary embolus, all of which were negative. Absence of pulmonary hypertension should precipitate the search for a shunt as a potential cause.

Diagnosis of POS caused by intra-cardiac shunting requires postural pulse oximetry and bubble contrast TTE or TOE (preferably on a tilt-table). Treatment can be offered in form of percutaneous or surgical PFO/ASD closure.

References