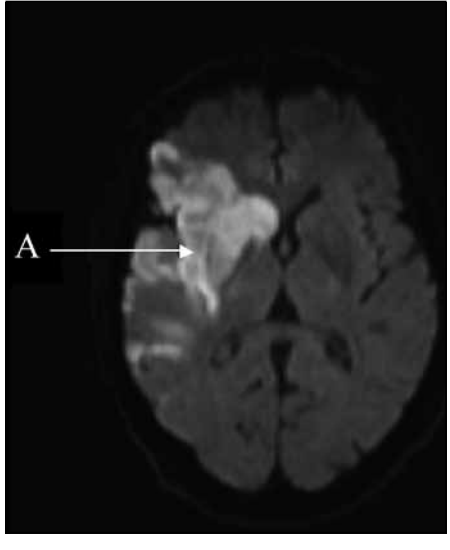
Pulmonary and paradoxical embolism with platypnoea-orthodeoxia

Introduction

This article outlines the case of a 62-year-old man who presented with a deep vein thrombosis, pulmonary embolus and right middle cerebral artery territory ischaemic stroke in association with a patent foramen ovale with right-to-left shunting. Despite anticoagulation, he was noted to have desaturation and tachypnoea when upright, without other symptoms or cardiovascular compromise, that resolved on lying down. The article describes the syndrome of platypnoea-orthodeoxia, characterized by breathlessness, hypoxia and/or cyanosis while in the upright position that improves on lying down, its relationship to this case, and its resolution by closure of the patent foramen ovale.

Figure 1. Diffusion-weighted brain magnetic resonance imaging showing acute right middle cerebral artery territory infarction (A).



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Discussion

Platypnoea-orthodeoxia, first described by Burchell et al (1949), is characterized by breathlessness, hypoxia and/or cyanosis while in the upright position that improves on lying down. It manifests as a result of shunting of deoxygenated blood from the right to the left side of the heart through intracardiac or intrapulmonary shunts in the absence of elevated right heart pres-

Figure 2. Computed tomography pulmonary angiography showing a pulmonary embolus involving the right main pulmonary artery (A).

Case Report

A 62-year-old man presented to the emergency department having woken up with left-sided weakness. He denied any previous medical history, family history, recent travel or use of cigarettes, alcohol, prescribed or recreational medications. Physical examination revealed left-sided hemianopia, hemineglect, hemiplegia and hemianaesthesia, and a swollen left calf. Cardiorespiratory and abdominal examinations were unremarkable. Laboratory investigations demonstrated elevated D-dimers 9.9 mg/litre (normal range <0.55 mg/litre), the presence of lupus anticoagulant and respiratory alkalosis with hypoxia on arterial blood gas analysis (pH 7.51, partial pressure of carbon dioxide 3.9 kPa, partial pressure of oxygen 5.9 kPa, bicarbonate 24 mmol/litre). Electrocardiography and chest X-ray were normal. Brain magnetic resonance imaging (*Figure 1*) demonstrated acute right middle cerebral artery territory infarction and computed tomography pulmonary angiography (*Figure 2*) showed a pulmonary embolus involving the pulmonary trunk extending into the right and left main pulmonary arteries. Leg venous Duplex ultrasound revealed a left popliteal vein thrombosis. Carotid Duplex ultrasound was normal. Transoesophageal echocardiography demonstrated a patent foramen ovale with right-to-left interatrial shunting.

He was treated with intravenous heparin and oxygen. Despite this, episodes of desaturation (96% to 85%) and tachypnoea (18–34 breaths/min) were noted on sitting up from the supine position, without other symptoms or cardiovascular compromise. This resolved on lying down. A diagnosis of platypnoea-orthodeoxia was made. The patent foramen ovale was closed percutaneously leading to resolution of these symptoms. Oral anticoagulation was subsequently provided to prevent further thromboembolic events.

sures. A number of different mechanisms have been proposed depending on the anatomical abnormality. In the setting of an interatrial shunt, as in this case, it has been suggested that the orthodeoxia results from stretching of the interatrial communication while upright, and this may be enhanced by persistence of a vestigial Eustachian valve at the junction of the inferior vena cava and right atrium that directs blood flow from the right to left atrium (Cheng, 1992).

Patent foramen ovale affects approximately 25% of the population but very few patients suffer with platypnoea-orthodeoxia (Hagen et al, 1984). In addition to an 'anatomical' component, for platypnoea-orthodeoxia to occur, a 'functional' component must exist incorporating an additional cardiorespiratory disease process that potentiates the right-to-left shunting (Cheng, 2002). In this case, the authors propose that the pulmonary embolus involving the pulmonary trunk resulted in obstruction to the right ventricular tract outflow, potentiating the passage of deoxygenated blood from the right to left atrium across the patent foramen ovale. Although platypnoea-orthodeoxia is being increasingly recognized, pulmonary embolus as an associated 'functional' process has only been described once previously (Syed and Sabeen, 2004). It may be that unless a pulmonary embolus presents a sufficient obstruction to right ventricular tract outflow, it may not be a sufficient stimulus to right-to-left shunting. As in this case, closure of intracardiac or intrapulmonary shunts and treating underlying pulmonary conditions may cause resolution of the symptoms. BJHM

Burchell HB HHJ, Wood EH (1949) Reflex orthostatic dyspnea associated with pulmonary hypertension. *Am J Physiol* **159:** 563–4

Cheng TO (1992) Reversible orthodeoxia. *Ann Intern Med* **116:** 875

Cheng TO (2002) Mechanism of platypnea-orthodeoxia: what causes water to flow uphill? *Circulation* **105:** e47

Hagen PT, Scholz DG, Edwards WD (1984) Incidence and size of patent foramen ovale during the first 10 decades of life: an autopsy study of 965 normal hearts. *Mayo Clin Proc* **59:** 17–20

Syed FH, Sabeen FM (2004) Platypnea-orthodeoxia: Report of two cases and review of the literature. *South Med J* **97:** 657–62

LEARNING POINTS

- Platypnoea-orthodeoxia may occur in patients with underlying intracardiac or intrapulmonary communications who develop common cardiorespiratory disorders like pulmonary embolus which affect intrathoracic blood flow, so the general physician needs to be aware of it and its management.
- While this condition was initially thought to be rare it is being increasingly recognized, but pulmonary embolism as an associated 'functional' condition has only been described once previously.
- It is most commonly associated with intracardiac and intrapulmonary shunts. Orthostatic refractory hypoxaemia should alert the possibility of platypnoea-orthodeoxia.
- This condition is treated by managing the underlying cause, which substantially improves symptoms.
- Diagnosis may require contrast-enhanced echocardiography and postural manoeuvres or peripheral contrast tilt-table transoesophageal echocardiography, technetium-labelled macroaggregated albumin scanning, and pulmonary arteriography.

Ileo-ileal intussusception from Crohn's ileitis

Crohn's disease can cause abdominal pain and diarrhoea. A 33-year-old man presented with a 3-month history of recurrent abdominal pain and occasional diarrhoea. A computed tomogram of the abdomen and pelvis demonstrated ileal intussuception (*Figure 1*) and thickened terminal ileum (*Figure 2*). The patient was treated by laparoscopic reduction of the intussusception and a right hemicolectomy and was started on steroids. Histology of the resected specimen confirmed the presence of Crohn's disease.

Small bowel intussusception is caused by several conditions such as polyps, lymphoma, prominent Peyer's patches and Meckel's diverticulum. Only very few cases of intussusception associated with Crohn's disease have been reported in the literature. In this case, the computed tomography scan helped in the diagnosis that was confirmed histologically. BJHM

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Figure 1. Transverse section of computed tomography scan demonstrating ileo-ileal intussuception.

Figure 2. Transverse section of computed tomogram of the abdomen and pelvis demonstrating thickened terminal ileum.