CASE REPORT

Platypnoea orthodeoxia after hepatic surgery

Muhammad Asrar ul Haq,^{1,2} Vivek Mutha,^{1,2} Ahmed Al-Kaisey,¹
William J van Gaal^{1,2}

¹Department of Cardiology,
The Northern Hospital,
Melbourne, Victoria, Australia
²Department of Medicine,
University of Melbourne,
Melbourne, Victoria, Australia

Correspondence to
Dr Muhammad Asrar ul Haq,
muhammad.asrar@
unimelb.edu.au

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SUMMARY

Platypnoea orthodeoxia is a rare clinical syndrome characterised by dyspnoea and deoxygenation exhibited in upright position with improvement in supine posture. Previously described aetiologies include cardiac (pericardial effusion or constrictive pericarditis), pulmonary (pneumonectomy, emphysema or amiodarone toxicity), abdominal (cirrhosis or ileus) or vascular (abdominal aortic aneurysm) causes. We report an unusual case of platypnoea orthodeoxia associated with patent foramen ovale and partial hepatic resection.

BACKGROUND

Platypnoea orthodeoxia is a rare clinical syndrome characterised by dyspnoea and deoxygenation exhibited in upright position, for example, sitting or standing, and improves in supine posture. We report an unusual case of platypnoea orthodeoxia associated with patent foramen ovale (PFO) and partial hepatic resection.

CASE PRESENTATION

A 53-year-old woman was admitted electively for left hemihepatectomy due to recurrent ascending cholangitis and intrahepatic cholelithiasis. Her medical history included insulin-dependent type 2 diabetes mellitus, diverticular disease and dyslipidaemia. There were no previous cardiac or respiratory symptoms of note. The postoperative course was complicated by acute renal impairment, sepsis and bile leak. On week 5 postoperatively, the patient complained of increasing shortness of breath and was noted to be hypoxic, desaturating up to 78% on room air. Her physical examination did not reveal any other significant finding. Her blood test results remained unchanged, with a moderate degree of renal impairment (creatinine of 250 µmol/L). CT pulmonary angiogram did not suggest any pulmonary pathology.

Extensive investigations were undertaken looking for various potential causes of her symptoms including sepsis, chest infection, pulmonary embolism and hepatic congestion without much success. Interestingly, her symptoms were worse on standing up which stopped her from participating in the physiotherapy programme. She denied dyspnoea when in bed. Her oxygen saturation patterns (table 1) demonstrated minimal improvement with supplemental oxygen. A significant change with posture was however noticed.

INVESTIGATIONS

A transthoracic echocardiogram was performed which showed normal left ventricular systolic and

Table 1 Oxygen saturation patterns with varying postures

Posture	Saturation on room air (%)	Saturation with supplemental oxygen (%)
Lying in bed	93–95	NA
Sitting	80–83	85
Standing	78–80	85

diastolic function, normal valves, no pericardial effusion, normal pulmonary artery pressure and no evidence of constrictive or restrictive pathology. It however was strongly positive for microbubble contrast across the intra-atrial septum, in semiupright position. A subsequent transoesophageal echocardiogram revealed a small tunnel PFO measuring 0.24 cm (figure 1; video 1) and confirmed a change in haemodynamics with posture with left-to-right shunt on supine changing to right-to-left shunting on erect position.

OUTCOME AND FOLLOW-UP

The PFO was successfully closed using an Amplatzer device 25 mm. Her symptoms improved remarkably and the renal function returned to baseline. The patient participated in the rehabilitation programme with good results, and remains well on 1-year follow-up with an oxygen saturation of above 95% on room air and no significant change with posture.

DISCUSSION

Platypnoea orthodeoxia is a rare condition which may be associated with cardiac, pulmonary or hepatic diseases.¹ Clinical features may include disabling postural hypoxia in upright posture with dramatic improvement in supine position and minimal effect of supplemental oxygen. To the best of our knowledge, this is the first case of this syndrome associated with subdiaphragmatic surgery reported in literature. We postulate that changes in hepatic blood flow dynamics secondary to surgery in association with congenital PFO may have caused this syndrome.

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.^{2 3} Normally, the left atrial pressure is 5–8 mm Hg higher than the right atrial pressure; this pressure difference is thought to prevent right-to-left shunting through an anatomical defect. Transient shunting of blood through this anatomical defect can occur in special situations, such as

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Figure 1 (A) Transthoracic echocardiogram (4 chamber view) demonstrating strongly positive microbubble study with contrast completely opacifying left ventricle. (B) Transoesophageal echocardiogram demonstrating small PFO. (C) Intraoperative colour Doppler of atrial septum. (D) Final result post-Amplatzer device implantation (arrow). LA, left atrium; LV, left ventricle; PFO, patent foramen ovale; RV, right ventricle.

Valsalva manoeuvre. Therefore, it is important to understand that PFO alone should not cause symptoms.^{4 5}

Platypnoea orthodeoxia was first described by Burchell *et al*² in 1949. Both the anatomical defect (eg, PFO or atrial septal defect) as well as the functional component producing a right-to-left shunt on upright posture are required to cause this syndrome. Potential causes described previously are cardiac (pericardial effusion or constrictive pericarditis), pulmonary (pneumonectomy, emphysema or amiodarone toxicity), abdominal (cirrhosis or ileus) or vascular (abdominal aortic aneurysm).³ The diagnosis can sometimes be difficult but should be suspected in patients with hypoxia that is more pronounced on sitting or standing especially when hypoxia is not corrected with supplemental oxygen. The treatment is aimed at treating the cause. If symptoms are still present, then surgical closure of the anatomical defect should be considered.

Video 1 Transthoracic echocardiogram (4 chamber view) demonstrating strongly positive microbubble study with contrast completely opacifying left ventricle in upright posture.

Learning points

- ▶ Platypnoea orthodeoxia is a rare condition which may cause disabling postural hypoxia in upright posture with improvement in supine position and minimal effect of supplemental oxygen.
- ▶ An anatomical defect (eg, patent foramen ovale or atrial septal defect) and a functional component producing a right-to-left shunt on upright posture are required to cause this syndrome.
- ▶ Potential aetiologies include cardiac (pericardial effusion or constrictive pericarditis), pulmonary (pneumonectomy, emphysema or amiodarone toxicity), abdominal (cirrhosis or ileus) or vascular (abdominal aortic aneurysm).
- ▶ Closure of the anatomical defect may be required if treatment of causative pathology does not succeed or is not possible.

Competing interests None.

Patient consent Obtained.

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