Patent foramen ovale presenting with platypnoea-orthodeoxia syndrome and stroke after multi-organ resection

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SUMMARY
Platypnoea-orthodeoxia syndrome (POS) is defined by oxygen desaturation and dyspnoea in upright position that improves by lying down. It results from a right to left shunt at the intracardiac or intrapulmonary level. A 53-year-old ovarian cancer patient presented with POS that was refractory to oxygen therapy. The symptoms began after an extensive abdominal and pelvic surgery as treatment of her cancer with a complex hospital course. A patent foramen ovale was found with the use of transoesophageal echocardiography. A percutaneous closure was done with positive outcome and dyspnoea disappearance. In this case with its challenging clinical setting, we present a unique clinical scenario of an immediate postoperative POS syndrome. We address the different therapeutic modalities and the need for a multidisciplinary medical approach.

BACKGROUND
Platypnoea refers to a worsening of dyspnoea that occurs in the upright position and improves with recumbency and orthodeoxia refers to arterial oxygen tension drop by more than 5% or 4 mm Hg in the upright position. This could be attributed to intracardiac shunts related to congenital anatomical defects such as patent foramen ovale (PFO), atrial septal defect (ASD) or atrial septal aneurysm with septal fenestrations, with or without elevated right–left gradient. These heart defects are usually accompanied by thoracic or abdominal structural abnormalities in patients presenting with platypnoea-orthodeoxia syndrome (POS). POS could also be caused by pulmonary pathologies where there is ventilation/perfusion mismatch or pulmonary arteriovenous shunting.

POS is considered a rare condition and its true prevalence in the population is not known. Clinically, silent PFO is a term described in the literature for uncomplicated PFO. In a minority of cases, the structural defect is discovered after cerebral ischaemic events.

We present an unusual mechanism of POS mediated by PFO and presented immediately after extensive surgical operation where many abdominal and pelvic organs were removed.

CASE PRESENTATION
A 53-year-old woman known to have operated ovarian cancer with right salpingo-ovariectomy and adjuvant chemotherapy 1 year ago, history of deep vein thrombosis (DVT) on long-term anticoagulation and iron deficiency anaemia. She was hospitalised for a scheduled laparotomy for the treatment of her cancer where she underwent extensive abdominal and pelvic organ resections that included peritoneectomy, splenectomy, hysterectomy, left annexectomy, appendixectomy, colectomy, infracolic omentectomy and diaphragm scraping. In the immediate postoperative phase, she presented with abrupt aphasia. On physical examination,
Case report

Blood pressure was 100/60 mm Hg, heart rate 108 bpm, cardio-pulmonary auscultation within normal limits, no heart failure signs, aphasic patient, oxygen saturation 98% on 12 L oxygen mask on lying position. ECG found normal sinus tachycardia with no conduction or ischaemic changes. Sodium 137 mmol/L, potassium 3.8 mmol/L, creatinine 46 µmol/L, glomerular filtration rate 109 mL/min and haemoglobin 107 g/L.

INVESTIGATIONS

Brain CT found left middle cerebral artery territory stroke (figure 1). The work up showed PFO on transoesophageal echocardiography (figure 2), while there were no relevant findings with transthoracic echocardiography (TTE).

DVT was found on Doppler ultrasound of lower limbs for which an inferior vena cava filter was implanted due to embolisation of DVT despite therapeutic anticoagulation (figure 3). The hospital stay was characterised by a refractory hypoxia in the setting or standing position which did not respond to optiflow or non-invasive mechanical ventilation, and it disappears on lying position. The patient was diagnosed with POS syndrome. She was then transferred to our cardiology department. Afterwards, heart catheterisation revealed normal right atrial pressure (mean: 6 mm Hg), normal right ventricular pressure (23/8 mm Hg), normal mean pulmonary capillary wedge pressure (10 mm Hg) and left atrial pressure (5 mm Hg). Cardiac output was 6.3 L/min and cardiac index was 3.4 L/min by estimated Fick measurement. Oximetry showed no significant increase in oxygen saturation in right-heart chambers.

DIFFERENTIAL DIAGNOSIS

High index of suspicion is required to bring it up to deferential diagnosis. The evaluation needs to correlate between dyspnoea and upright posture. Patients can present with POS without underlying cardiac causes. However, similar right-to-left communication can be found in the form of pulmonary arteriovenous malformation or severe ventilation perfusion mismatching.6 This association is also observed in cases of chronic liver disease in the presence of hepatopulmonary syndrome where worsening of oxygen desaturation is observed on standing position.7 In our case, the aetiology of POS was a right-to-left shunt at the cardiac level with no associated extra-cardiac causes.

TREATMENT

After multidisciplinary team discussion, percutaneous foramen ovale closure was decided to be the strategy of choice for her refractory hypoxia with no other underlying aetiology and to prevent stroke recurrence. The evidence on the effects of PFO closure for the treatment hypoxia is limited to case reports; however, it is promising. Hypoxia was found to improve immediately following PFO closure.8

Our patient was transferred to the catheterisation laboratory where foramen ovale closure was performed using Amplatzer septal occluder (size: 35 mm) (figure 4). The procedure was under general anaesthesia and was guided by transoesophageal echocardiography (TOE) (figure 5). In postprocedural period, TTE showed interatrial prosthesis located in place with no residual shunt along with small pericardial effusion (figure 6).

OUTCOME AND FOLLOW-UP

The hypoxia subsided postoperatively and subsequent 3 months follow-up revealed positive outcomes with no hypoxia episodes.
The first event in our case was a stroke that necessitated inferior vena cava filter placement due to anticoagulation contraindication and the high risk of venous thromboembolism in postoperative period.18 Assuming that thrombi crossed the PFO, this means there is a right-to-left shunt consolidating the diagnosis of POS. Many precipitating factors in patients with POS and PFO were described in the literature as mentioned above; however, post extensive abdominal organs resection have not been reported previously. Nevertheless, the most probable theory in our case is that resection of several abdominal pelvic organs has led to a change in volume distribution with less organs accommodating blood and more volume returning to right-heart chambers with an associated septal displacement favouring right-to-left shunt even in the absence of persistent high right-heart pressures. The use of relevant diagnostic tools confirms the presence of PFO. PFO percutaneous closure has proven to be a safe and a successful treatment for patients with POS, with rapid symptom relief and low complication rates.19 Our patient showed immediate positive outcome with dyspnoea disappearance in post-procedural period. Three-month follow-up showed neither recurrence of dyspnoea nor cerebrovascular events.

CONCLUSION
A previously silent PFO presented with stroke and POS after extensive abdominal organs resection in a 53-year-old cancer patient. This was managed with percutaneous device closure with positive outcome in 3-month follow-up.

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