Undiagnosed adult congenital heart disease presenting with postpartum dyspnoea: a case report

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Abstract

Patients with adult congenital heart disease are born with structural heart defects who survived into adulthood. Occasionally, complex lesions remain undiagnosed, potentially causing substantial cardiovascular health problems at young age. Here, the case is presented of a patient with subacute heart failure 1 week postpartum, revealing the diagnosis of aortic coarctation (CoA) with patent ductus arteriosus (PDA). A 34-year-old woman presented to the emergency department with severe hypertension and exercise-related dyspnoea 1 week postpartum. An initial diagnosis of pulmonary embolism was made after detection of a solitary opacity in the pulmonary artery (PA) on CT pulmonary angiography. Symptoms persisted despite anticoagulant treatment. Thorough clinical and echocardiographic reassessment unmasked the diagnosis of severe CoA with PDA, which was treated with percutaneous dilatation and stenting. Follow-up consultation 4 weeks later showed an asymptomatic patient with normalized blood pressure. The puerperium is a high-risk period to develop hypertensive heart failure for mothers with pre-existing heart disease, due to mobilization of extracellular fluid to the intravascular compartment. Undiagnosed CoA should always be ruled out in case of unexplained postpartum hypertension. When detecting a solitary opacity in the PA, a PDA with associated heart defects should be excluded by further investigations. This opacity is located at the orifice of the PDA in the PA and is probably a flow effect, which results from the mix of contrast-free with contrast-rich blood.

Keywords  Adult congenital heart disease; Aortic coarctation; Patent ductus arteriosus; Acute heart failure; Postpartum hypertension; Postpartum dyspnoea; Case report

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Introduction

Patients with adult congenital heart disease (ACHD) are born with structural defects of the heart and/or great vessels who survived into adulthood. It is estimated that approximately 3.5 million ACHD patients live in Europe and the US alone.¹ At least 10% of ACHD patients remain undiagnosed until adulthood.² Most of these have simple defects, e.g. small atrial or ventricular septal defects, Ebstein’s anomaly, etc.¹,³ Occasionally, more complex lesions remain undiagnosed due to rather vague or mild symptoms. In this case report a 34-year-old woman is presented with occult CoA and PDA, unmasked in the postpartum period. Several studies have shown that the diagnosis of non-critical CoA is often delayed, despite the presence of specific symptoms such as therapy-resistant arterial hypertension at young age, cardiac murmurs, and differential blood pressure (BP) between upper and lower extremities.⁴-⁶ When left untreated, apparently mild cases of CoA may in time lead to hypertensive complications such as compensatory left ventricular hypertrophy (LVH), left heart failure, aortic dissection and premature coronary and cerebral atherosclerosis. CoA can occur as isolated heart defect, but is often associated with other conditions of which a bicuspid aortic valve is the most frequent.⁷,⁸ Less frequent associations are aortic arch hypoplasia, subaortic stenosis, mitral valve abnormalities, ventricular and atrial septal defects and PDA.⁹ CoA is detected in 5–8% of all congenital heart defects. Up to 18% of patients treated for CoA have a concomitant PDA.¹⁰

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Figure 1 Transverse (left panel) and coronal (right panel) images of the CT pulmonary angiography through the solitary opacity (red arrow). The opacity is located in the pulmonary trunk next to the orifice of the left pulmonary artery.

Figure 2 Transthoracic echocardiographic images. Colour Doppler ultrasound shows flow turbulence at the distal pulmonary artery (upper left panel, high parasternal short axis view, red arrow) suggesting the presence of a small PDA. A visible stenosis is detected in the proximal descending aorta with notable flow turbulence on colour Doppler (upper right panel, suprasternal view, red arrow), suggestive for a CoA. Continuous wave Doppler shows a high maximum pressure gradient of 79 mmHg over the stenosis with a prominent diastolic tail, illustrating severe CoA (lower middle panel, suprasternal view, red arrow).
Case report

A 34-year-old woman was admitted to the emergency department (ED) with general fatigue and progressive exercise-related dyspnoea since 3 days, 1 week postpartum (A0P2G2). In the last trimester of her pregnancy, she was diagnosed with gestational hypertension for which she was treated with methyldopa (250 mg, three times daily). She had no other relevant medical history.

A significantly elevated BP of 186/80 mmHg was measured on admission, with oxygen saturation of 91%, heart rate of 92 bpm and normal temperature (36.5°C). Clinical examination revealed a systolic heart murmur over the left infraclavicular region, bibasilar crackles and mild bilateral malleolar pitting oedema. The electrocardiogram showed normal sinus rhythm with criteria for LVH. Plain film radiography demonstrated a dilated hilar vasculature and mild bilateral pleural effusion. The blood results showed elevated D-dimers (1333 ng/mL, norm.: <500 ng/mL) and NT-pro-BNP (426 pg/mL, norm.: <125 pg/mL). Echocardiography showed a non-dilated left ventricle with normal function and moderate concentric hypertrophy, without signs of pulmonary hypertension or right ventricular failure.

A CT pulmonary angiography (CTPA) was performed because of high suspicion of pulmonary embolism (PE). It showed a solitary opacity of 7 mm in the PA next to the origin of the left pulmonary artery (LPA) (Figure 1). A venous duplex ultrasound showed no signs of deep venous thrombosis of the lower extremities. A diagnosis of postpartum PE was made and the patient was admitted. A therapeutic dose of low-molecular-weight heparin was started. Because of signs of hypertensive heart failure (NYHA Class II), furosemide

Figure 3 3D reconstruction of the CT angiography of the aorta. Remark the well-developed collateral circulation (red arrows) with a large calibre of the left subclavian (LSA), left common carotid (LCA), internal mammary (IMA), and intercostal arteries. Ao, aorta; PA, pulmonary artery.
Follow-up consultation took place 6 weeks later, revealing that symptoms persisted. Focused clinical re-evaluation showed a large differential systolic BP between the right arm (178 mmHg) and the right leg (100 mmHg). Echocardiographic reassessment with continuous wave Doppler (suprasternal view) showed a maximum pressure gradient of 79 mmHg in the upper descending aorta with prominent diastolic tail, suggestive for CoA (Figure 2). Furthermore, retrograde flow turbulence was observed in the distal PA in the high parasternal short axis view, suggestive for a small PDA. A CT aortic angiography was performed to confirm the diagnosis (Figure 3), showing a high-grade stenosis in the proximal descending aorta, compatible with severe CoA. Furthermore, a PDA was visible just proximally to the coarctation. The ductus was connected to the PA near the origin of the LPA, where the solitary opacity was detected. With these findings, the initial diagnosis of lung embolism was abandoned, and anticoagulation was stopped.

The patient was referred to a specialized centre for catheterization, where the coarctation was dilated and stented with a covered stent of 45 mm length (CCP 8Z45) on a 20 mm balloon (Figure 4). It was anticipated that the stent would obstruct flow down the PDA, resulting in its closure. Dual antiplatelet therapy was started after procedure, i.e. aspirin for 6 months and clopidogrel for 3 months. Follow-up consultation took place 1 month later. Her symptoms had greatly improved, and her BP was normalized. She even managed to stop antihypertensive treatment a few weeks before consultation. Echocardiographic reassessment showed a pressure gradient of 14 mmHg over the stent without any evidence of a PDA. Diuretics were stopped, as the central venous pressure was estimated to be normal, and clinically, there were no signs of heart failure anymore.

Discussion

This patient had undiagnosed ACHD consisting of CoA with PDA. She presented with hypertensive heart failure 1 week postpartum. Postpartum hypertension complicates approximately 2% of pregnancies. It typically occurs in the first 2 weeks postpartum due to mobilization of extravascular fluid to the intravascular compartment. This fluid shift and increase of BP must have triggered subacute heart failure in a patient with LVH and associated diastolic dysfunction due to untreated CoA.

CTPA showed a solitary opacity in the PA, first diagnosed as PE. However, the presence of a PDA at the same location suggests it is no embolism, but probably a flow effect, composed of contrast-free blood in the PDA surrounded by contrast-rich blood in the PA. An alternative hypothesis is local thrombus formation rather than embolization. Several case reports of patients with PDA have reported the presence of local thrombi in the PA, almost exclusively located near the opening of the ductus. At this point, flow turbulence is maximum, exerting shear stress on the vessel wall and facilitating endothelial injury. Traumatized endothelium triggers local inflammation and is prone to thrombus formation by platelet adhesion and activation of the coagulation cascade.

This case report implies several interesting conclusions. First, it is important to recognize the puerperium as high-risk period for mothers with pre-existing heart disease to develop heart failure, due to shift of relatively large
amounts of extracellular fluid to the intravascular compartment. Second, undiagnosed ACHD always needs to be included in the differential diagnosis of a patient with postpartum dyspnoea. When she presents with unexplained postpartum hypertension, CoA should be ruled out. Third, a solitary opacity in the PA visualized on CTPA should raise suspicion of the presence of a PDA with associated heart defects and should be investigated further.

Conflict of interest
None declared.

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