Abstract

Pulmonary hypertension due to congenital heart disease continues to be a diagnostic challenge despite modern diagnostic modalities. Herein, we report a 26-year-old woman with an incidentally documented patent ductus arteriosus and Eisenmenger syndrome. She presented with progressive dyspnea and exercise intolerance which was initially attributed to pulmonary embolus. She was started on macitentan and tadalafil therapy aiming to reduce the pulmonary vascular resistance with consideration for heart–lung transplantation should any further deterioration occur.

Keywords: Eisenmenger syndrome, Patent ductus arteriosus, pulmonary hypertension

INTRODUCTION

The ductus arteriosus is an essential connection between the pulmonary artery and aorta; it is necessary for proper fetal circulation. Functional closure of the ductus arteriosus occurs soon after birth. After 3 months of age, the persistence of duct between the aorta and pulmonary artery is named patent ductus arteriosus (PDA) which accounts for 6% to 11% of all congenital heart anomalies. PDA can lead to irreversible pulmonary hypertension (PH) with development of Eisenmenger syndrome which is a rare clinical finding in adult patients. We report a case of PDA with PH in a woman which was incidentally discovered on cardiac catheterization.

CASE REPORT

A 26-year-old woman presented to our emergency department with progressive dyspnea lasting several months. She had a history of three times spontaneous abortion. She initially was consulted with the chest disease department. After a thorax computerized tomography (CT) scan was performed, she was hospitalized with the diagnosis of subsegmentary pulmonary embolus [Figure 1]. On physical examination, blood pressure was 110/60 mm Hg and heart rate was 82 beats/min. Her respiratory rate was 24 breaths/min, with an oxygen saturation of 79% by pulse oximetry while breathing room air. A transthoracic echocardiogram (TTE) revealed an estimated systolic pulmonary artery pressure of 115 mm/hg with the right heart chamber dilatation. On auscultation,

Figure 1: Transvers computerized tomography image

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How to cite this article: Koza Y, Birdal O, Pirimoğlu B, Taş H. A 26-year-old woman with worsening dyspnea: Look closer, think critically. J Cardiovasc Echography 2020;30:116-8.
there was a loud P2 with a soft holosystolic murmur in the tricuspid area and a long diastolic murmur in the pulmonary area. The electrocardiogram revealed right ventricle (RV) hypertrophy, and chest X-ray showed a prominent central pulmonary trunk [Figure 2]. Serologic markers for connective tissue disease and thrombophilia panel were unremarkable. A right heart catheterization revealed severe pulmonary arterial hypertension with a mean pulmonary artery pressure of 76 mm/hg and a pulmonary capillary wedge pressure of 10 mm/Hg [Video 1].

**DISCUSSION**

Despite modern clinical and imaging innovations, PH due to congenital heart defects continues to be occult clinically and, in many cases, is still detected incidentally. PDA is a congenital heart defect in which the ductus arteriosus that typically connects the aorta to the left pulmonary artery fails to spontaneously close after birth.[2-4]

In the absence of a high velocity shunt at PDA level in Eisenmenger patients, it may be difficult to diagnose a PDA on echocardiography. In PDA patients with Eisenmenger syndrome, the shunt is bidirectional, mainly from right-to-left, low-velocity, and with no associated continuous murmur.[3,4]

Clinical manifestations of PDA are dependent on size of the ductus, the age of the patient, the pressure differential across the ductus, and the presence or absence of PH. The vast majority of patients may remain asymptomatic throughout life or it may be diagnosed accidentally or when it becomes symptomatic. The incidence of silent PDA is as high as 1 in 500 patients.[4,5]

In PDA with left-to-right shunting, TTE with color flow imaging provides a sensitive diagnostic tool with the use of the suprasternal notch or parasternal short-axis approach to image a jet in the main pulmonary artery. However, in cases of PDA associated with PH and a right-to-left shunt, the sensitivity of TTE has been estimated as low as 12%.[2-4]

The development of Eisenmenger syndrome from PDA is uncommon. As in the present case, development of Eisenmenger syndrome abolishes the classic “machinery” murmur of PDA, resulting in difficulty in arriving at a diagnosis. Deoxygenated blood from the RV is directed from the PDA to the aorta distal to the left subclavian artery, thus causing clubbing and cyanosis in the lower, but sparing the upper extremities. However, cyanosis may occur intermittently and is only present when pulmonary pressure is severe enough to reverse the shunt.[5-7] On closer examination, our patient was noted to have digital clubbing limited to her lower limbs but she had no cyanosis.

In the present case, PDA was not visualized on echocardiography and was overlooked on CT angiogram. The presence of PDA was incidentally detected on catheterization. On the right heart catheterization, attempts to engage the pulmonary artery directed the Multipurpose Amplatz catheter to the descending aorta. Her pulmonary artery and aortic pressures were nearly equal. CT images were reevaluated and PDA was visualized [Figure 3a-c]. Three months after the macitentan and tadalafil treatment, her exercise capacity improved and an increase of 70 m in 6-min walk distance was observed. The patient is currently tolerating double PH-specific therapy with a good functional status.

**Conclusion**

The most important learning point of this case is the necessity of a comprehensive medical history and physical examination. A detailed patient information is mandatory before the radiological examination. A congenital heart defect should always be sought and excluded in all patients presenting with PH. Due to the absence of a high velocity shunt at PDA level in Eisenmenger patients, large PDAs can be missed on routine echocardiography. More importantly, the presence of PDA should be investigated before patients become symptomatic.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.
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Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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