

A case of late postoperative pulmonary arterial hypertension: a rare entity

Geç ameliyat sonrası pulmoner arteriyel hipertansiyon olgusu:
Nadir bir antite

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Late postoperative pulmonary arterial hypertension is a rare and controversial entity and the mechanism of its development is not currently known. In this article, we report a 27-year-old female patient with late postoperative pulmonary hypertension diagnosed 16 years after the correction surgery of patent ductus arteriosus. The fact that the pulmonary artery pressure values have been within normal limits in the previous medical records suggests that she is a late pulmonary artery hypertension case.

Key words: Congenital heart disease; pulmonary hypertension/ diagnosis.

Pulmonary arterial hypertension associated with congenital heart disease (CHD-PAH) is different from pulmonary arterial hypertension (PAH) and poses a significant threat to patients in terms of both morbidity and mortality.^[1] The development of CHD-PAH is more frequent in surgically unrepaired patients,^[1] but, unfortunately, even for those patients who have undergone repair, postoperative reactive pulmonary hypertension can develop.^[2] Moreover, late postoperative PAH, albeit rare, is another possibility for clinical presentation.^[3] Late postoperative PAH is distinguished from postoperative reactive PAH in that it refers to the development or persistence of pulmonary hypertension after the immediate postoperative period despite what seems to be adequate surgical repair. Typically this condition is attributed to the late timing of the anatomic shunt correction, miscalculation of the likelihood of surgical correction, or longstanding effects of stable but elevated right ventricular afterload that lead to recalcitrant remodeling.^[2] Herein, we present a case of late postoperative PAH diagnosed 16 years after the correction of patent ductus arteriosus.

Geç ameliyat sonrası pulmoner arteriyel hipertansiyon nadir ve tartışmalı bir antitedir ve gelişim mekanizması halen bilinmemektedir. Bu yazıda, patent duktus arteriyozus düzeltme ameliyatından 16 yıl sonra geç ameliyat sonrası pulmoner arteriyel hipertansiyon tanısı konulan 27 yaşındaki bir kadın olgu sunuldu. Hastanın geçmiş tıbbi kayıtlarında pulmoner arter basıncının normal sınırlarda olması bu olgunun bir geç ameliyat sonrası pulmoner arteriyel hipertansiyon olgusu olduğunu düşündürmektedir.

Anahtar sözcükler: Doğuştan kalp hastalığı; pulmoner hipertansiyon/tanı.

CASE REPORT

A 27-year-old female patient was admitted to our clinic complaining of easy fatigability and dyspnea that had started six months ago. She also had been experiencing multiple syncopal attacks, the last one happened two days before her admission. Sixteen years before, she had undergone patent ductus arteriosus closure. According to the previous patient discharge form, her pulmonary pressure had been within normal limits at the time of preoperative diagnostic cardiac catheterization (systolic, mean, and diastolic pressures were 30, 18 and 12 mmHg, respectively), and her postoperative period had been uneventful. On her physical examination, a 2-3/6 grade systolic murmur was heard along the left lower parasternal border. An electrocardiogram (ECG) showed a right ventricular hypertrophy and right axis deviation. Transthoracic echocardiography (TTE) revealed mild to moderate tricuspid regurgitation, severe pulmonary hypertension (systolic pulmonary pressure: 98 mmHg calculated from peak tricuspid regurgitant velocity), a D-shaped left ventricle, and enlarged right

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Figure 1. High resolution computed tomographic view of the patient.

heart chambers. We did not find any residual ductal flow. Other diagnostic evaluations examining rheumatologic, hematologic, and pulmonary functions, including the diffusion capacity of carbon monoxide, yielded no evidence for other diseases that might cause PAH, such as scleroderma or parenchymal lung diseases. Her high-resolution chest computed tomography (CT) was normal, except for mild enlargement of the pulmonary truncus and the left and right main pulmonary arteries (Figure 1). However, ventilation-perfusion scintigraphy revealed hypoperfusion on her left lung, suggesting chronic thromboemboli (Figure 2). Her six-minute walk test was 300 m. We performed cardiac catheterization and detected no residual shunt. Pulmonary angiography revealed enlarged left and right main pulmonary arteries



Figure 2. Ventilation-perfusion scintigraphy of the patient.

(Figure 3). Systolic, mean, and diastolic pressures for the pulmonary artery, right ventricle, and right atrium were 99-61-42 mmHg, 90-30-0 mmHg, and 7-5-4 mmHg, respectively, with normal left ventricular end-diastolic pressure. A vasoreactivity test with iloprost was negative. Based on this data, we prescribed bosentan 62.5 mg bid; however, her transaminases increased at the two-week control, and we shifted her medication to an iloprost inhaler (nine times a day) and warfarin for a target an international normalized ration (INR) level between 1.5-2.5. Her follow-up visit at week 12 revealed a drop in pulmonary artery systolic pressure, which measured 73 mmHg in TTE. Furthermore, she improved from functional class III to II.

DISCUSSION

An important problem we were confronted with in this case was determining the exact cause of pulmonary arterial hypertension. It might have been a manifestation of “missed” pulmonary arterial hypertension already present at the time of operation. Indeed, the relatively late correction of the PDA in an 11-year-old patient could be supportive of preoperative PAH, but the discharge notes related to preoperative cardiac catheterization did not confirm this possibility. Reactive PAH may be another possible clinical scenario. We were not able to exclude the presence of reactive PAH right after corrective surgery but speculated that the absence of symptoms, such as dyspnea, for at least 15 years pointed to a more recent development of the disease. Although ventilation-perfusion lung scintigraphy

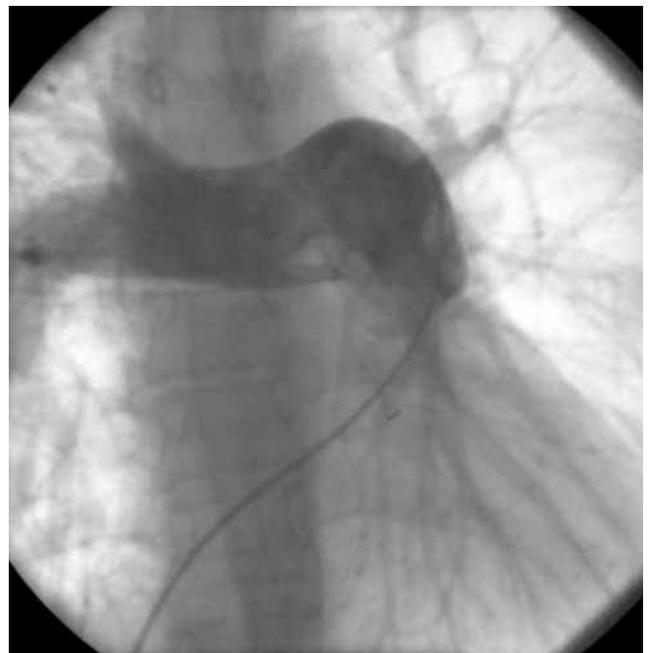


Figure 3. Pulmonary angiography of the patient.

revealing diffuse hypoperfusion of the left lung denotes a chronic thromboembolic event or left pulmonary artery hypoplasia, the angiographic appearance of the left pulmonary artery in this patient was normal, and no parenchymal pathology was seen on high-resolution chest CT. We decided hypoperfusion of left lung was not the result of chronic thromboemboli but the consequence of microvascular thrombus formation in the natural course of the existing pulmonary hypertension.^[4]

Differentiation between primary and late postoperative PAH was not possible in our case because the patient also had characteristics suggestive of idiopathic origin, such as female gender and young age. Nevertheless, we thought that her previous history of congenital heart disease correction supported the diagnosis of late postoperative PAH. Information about this disease is very scarce. Delayed surgical correction, as in our case, is one of the factors posited for its development. The presence of a high-pressure pulmonary vascular bed over a long period of time may have been an underlying trigger for the late emergence of PAH. More in-depth knowledge about this disease is needed for its prevention. Unfortunately, the rarity of

late postoperative PAH seems to be a major obstacle for collecting enough clinical data to achieve this goal.

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