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Case Report

Introducing a Rare Case: Unilateral Absence of the Pulmonary Artery Branch Associated With Large Patent Ductus Arteriosus in an Adult Patient

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ABSTRACT

Unilateral absence of the pulmonary artery (UAPA) is a rare congenital cardiovascular anomaly with a wide array of symptoms. An 18-year-old man was referred to our hospital with dyspnea on exertion and central cyanosis. Transthoracic and transesophageal echocardiographic examinations revealed severe right ventricular enlargement, large main and left pulmonary arteries, severe pulmonary hypertension, and a large patent ductus arteriosus (PDA). The right pulmonary artery could not be seen; consequently, UAPA was considered. Computed tomography angiography confirmed the diagnosis. Our case is a rare condition with UAPA associated with patent ductus arteriosus, diagnosed in adulthood. It underscores the need for awareness of this anomaly for early diagnosis and treatment. ***(Iranian Heart Journal 2022; 23(4): 125-130)***

KEYWORDS: Unilateral absence of pulmonary artery, Patent ductus arteriosus, Pulmonary hypertension, Congenital heart disease

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Unilateral absence of the pulmonary artery (UAPA) is a rare congenital cardiovascular anomaly with diverse presentations. It was first described by Frentzel in 1868.[1-3] Patients with isolated UAPA in the absence of other cardiovascular malformations usually survive into adulthood and experience only minor, if any, symptoms.[4,5] A late presentation in life, as with the patient described herein, is uncommon, and delayed diagnosis may be the result of atypical symptoms earlier in life that remain unrecognized. Such an anomaly can present a wide array of symptoms.[6] Routine investigations such as chest X-rays (CXR) and echocardiography may suggest the diagnosis of UAPA, but confirmation needs computed tomography (CT) scans and magnetic resonance angiography. An early diagnosis of UAPA and appropriate and timely intervention may improve the outcome.[1,2]

Case Report

An 18-year-old man was referred to our hospital (Rajaie Cardiovascular, Medical, and Research Center) for outpatient

evaluation and advanced echocardiography by a fellow of echocardiography due to severe enlargement and severe dysfunction of the right ventricle (RV).

The patient had a history of dyspnea on exertion class II and central cyanosis in strenuous activities. Recently, because of hard work during military service, his symptoms manifested themselves markedly. On physical examination, he had clubbing in his hands and feet, and the O_2 saturation level of his lower extremities was lower than that of his upper extremities. In laboratory tests, he had erythrocytosis, iron deficiency, and thrombocytopenia. Transthoracic echocardiography (TTE) revealed normal left ventricular (LV) size with moderate dysfunction (LV ejection fraction =40%), severe RV enlargement with severe dysfunction, a D-shaped LV in both systole and diastole, a large main pulmonary artery (PA), a large left PA, severe pulmonary hypertension (PH), and a systolic pulmonary pressure (sPAP) of about 90 mm Hg. The right PA was not seen in TTE. Nonetheless, due to the faint signal Doppler of tricuspid regurgitation, we assumed that the sPAP might have been underestimated. The suprasternal view in the Doppler study depicted a patent ductus arteriosus (PDA) with turbulent continuous flow and systolic dominance.

The patient's CXR had some findings in favor of UAPA, too, consisting of a smaller right hemithorax, ipsilateral hemidiaphragm elevation, highly diminished pulmonary vascular markings, contralateral lung hyperinflation, and herniation beyond the midline (Fig. 1).

The patient underwent transesophageal echocardiography, which revealed a small patent foramen ovale and a large tortuous PDA (Fig. 2). The point was that the transesophageal echocardiographic examination, too, did not show the right PA. Even contrast injection from the peripheral vein filled the right heart, the main PA, and the left PA; still, there was no evidence of the right PA. We, therefore, suggested that the absence of the right PA be considered. Other modalities were then employed to confirm our diagnosis. The patient underwent CT angiography, which confirmed the absence of the right PA (Fig. 3) and visualized the tortuous PDA (Fig. 4).

The patient underwent right and left heart catheterization, which revealed severe PH (sPAP=140 mm Hg), many collaterals, a large PDA, and the absence of the right PA (Fig. 5). Accordingly, he was scheduled for medical treatment.

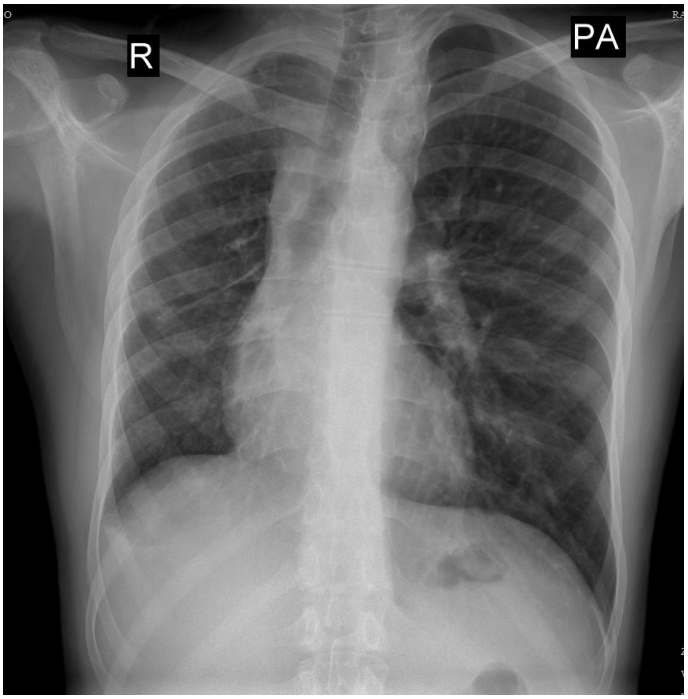

Figure 1: The chest X-ray of the patient shows an ipsilateral hemidiaphragm elevation (the yellow arrow), a smaller right hemithorax (the blue arrow), a contralateral lung hyperinflation, and herniation beyond the midline (the orange arrow).

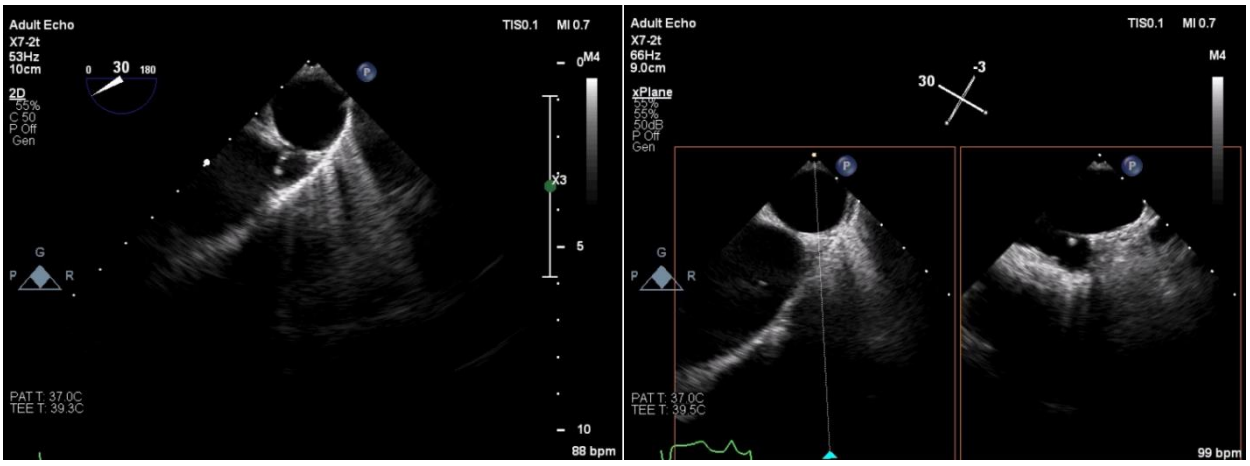

Figure 2: The transesophageal echocardiography images (the upper esophageal view) show a patent ductus arteriosus (the orange arrow).

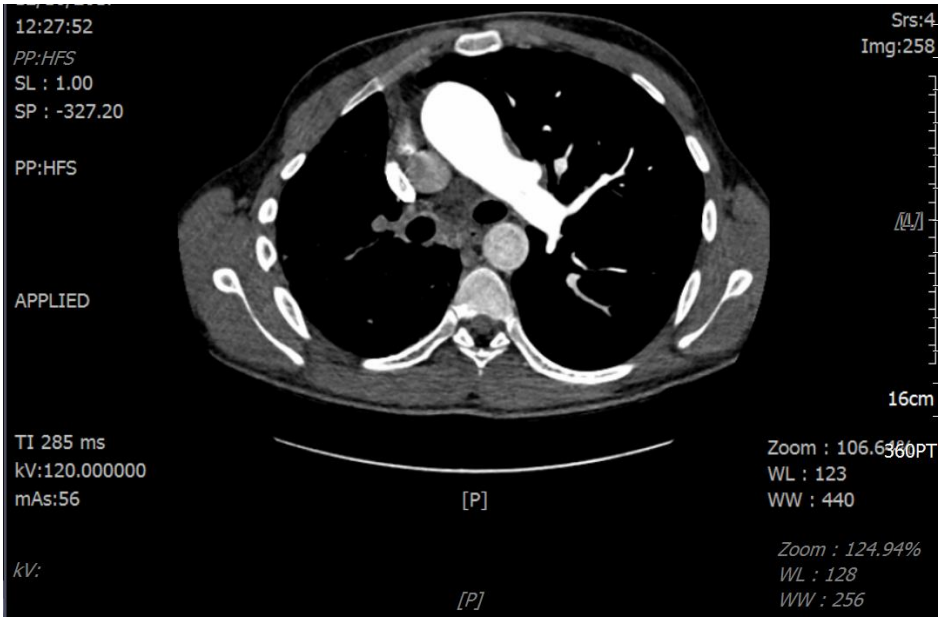

Figure 3: The chest computed tomography angiogram demonstrates the absence of the right pulmonary artery (the orange arrow).

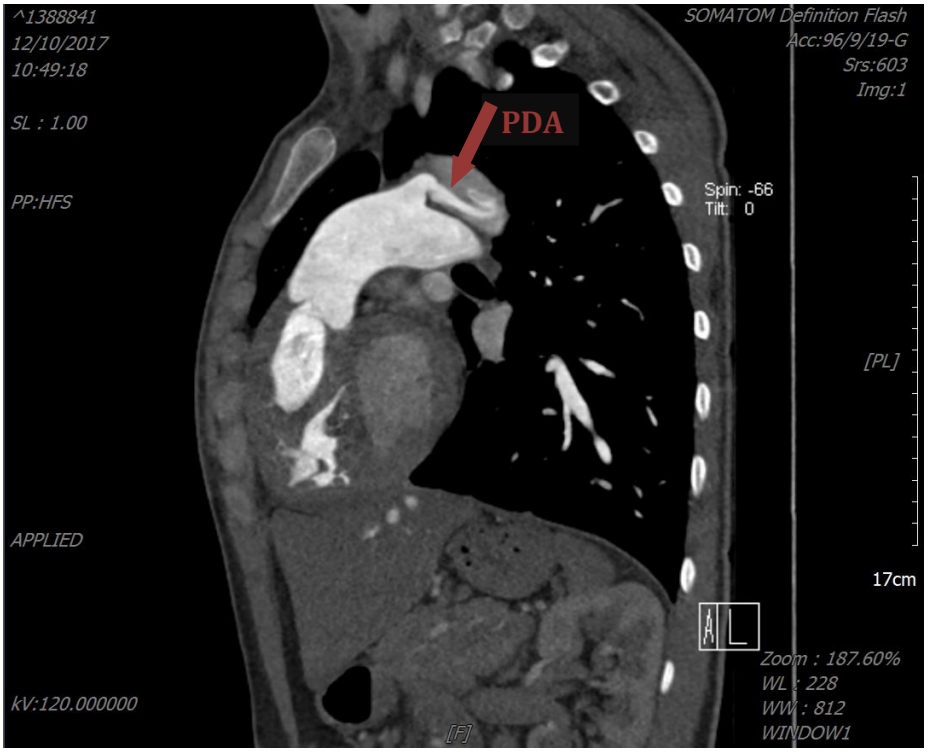

Figure 4: The chest computed tomography angiogram demonstrates the patent ductus arteriosus (PDA) (the orange arrow).

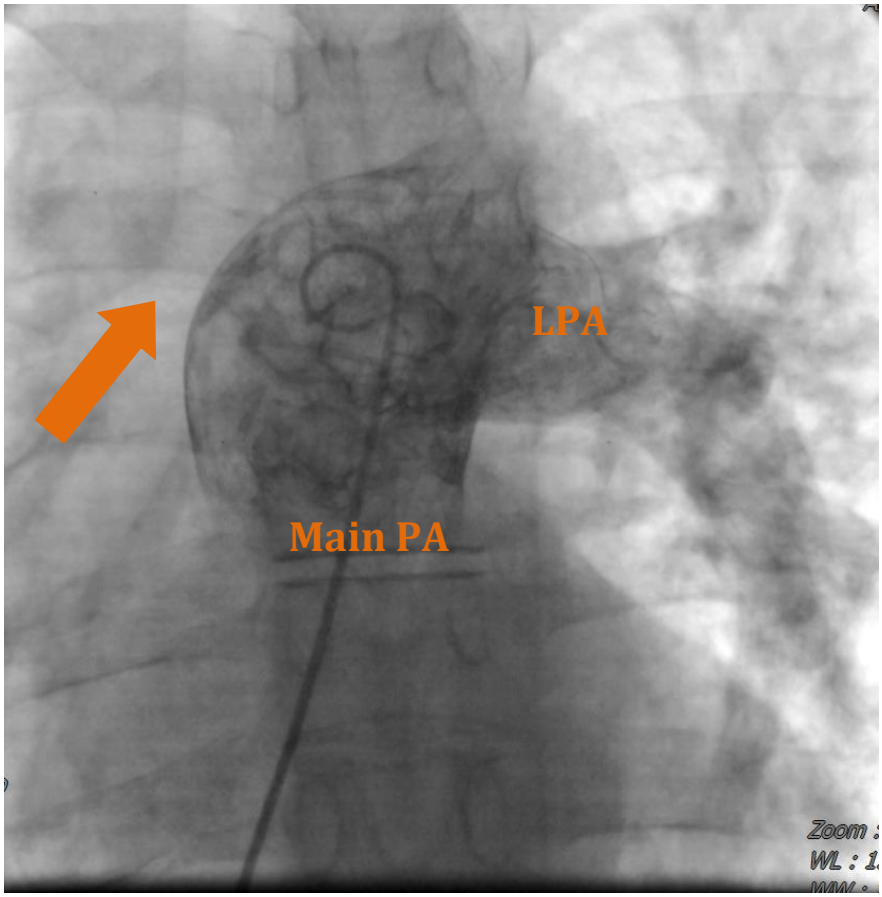

Figure 5: Right heart catheterization and contrast injection in the main PA shows the absence of the right pulmonary artery (the orange arrow).

PA, Pulmonary artery; LPA, Left pulmonary artery

DISCUSSION

UAPA is a very rare congenital cardiovascular malformation with a prevalence rate of about 1 in 200 000 young adults.[1,7] UAPA is twice as common on the right side. This condition is associated with cardiac malformations, such as tetralogy of Fallot, atrial septal defect, ventricular septal defect, transposition of the great vessels, coarctation of the aorta, right aortic arch, truncus arteriosus, PDA, and pulmonary atresia; additionally, it is sometimes an isolated malformation.[1,5]

Patients with UAPA have 2 types of presentations. The first type is seen in infants, usually presenting with congestive heart failure and PH. The other presentation type occurs in older patients, who are often asymptomatic. They tend to present with chest pain, pleural effusion, recurrent infections (37%), dyspnea or exercise intolerance (≈40%), PH (≈44%), hemoptysis (≈20%), and high-altitude pulmonary edema (≈10%). Moreover, some cases are detected during CXR incidentally. The prognosis depends on other associated cardiovascular anomalies and the severity of PH. Overall mortality in these cases is 7%. [1,5,7]

The diagnosis of UAPA is challenging and based on comprehensive history taking, physical examinations, and imaging tests. High suspicion is needed to make the diagnosis. A variety of imaging techniques are available to diagnose this malformation.

There is no consensus concerning the treatment of patients with UAPA. The

treatment is directed toward the clinical presentations. Some authors have recommended using serial echocardiography to follow up asymptomatic adults for the early detection of PH. Patients with PH can be treated medically with vasodilator agents. Alternatively, revascularization of the peripheral branches of the affected PA to the pulmonary hilum can be tried, and there are reports of successful revascularization procedures, mostly in childhood. Hemoptysis can be treated with embolization, lobectomy, or pneumonectomy. Severe infections may necessitate lobectomy or pneumonectomy. Any pulmonary surgery in a patient with UAPA may be complicated by the presence of systemic collaterals. It is necessary for these patients to be followed up closely, particularly for the observation of their pulmonary hemodynamics.[2,7-9]

Our case was a rare condition with UAPA and severe PH associated with PDA and was diagnosed in adulthood. The patient became a candidate for medical treatment due to systemic PH, and no other interventions were deemed beneficial for him.

CONCLUSIONS

As UAPA is a rare anomaly, the diagnosis needs high suspicion. Awareness of this anomaly will help us in early diagnosis and treatment. PH, a severe complication of this anomaly, increases the mortality rate significantly. Hence, early diagnosis may prevent this fatal complication and reduce morbidity and mortality through timely treatments.

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