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Unilateral pulmonary artery agenesis: clinical and laboratory findings of four cases and diagnostic clues for pediatricians

Tek taraflı pulmoner arter yokluğu: Çocuk uzmanlarına tanı koymada ipuçları, dört olgunun klinik ve laboratuvar bulguları

Introduction

Congenital unilateral absence of a pulmonary artery (UAPA) is a rare anomaly (1). The embryological explanation for the origin of UAPA is the involution of the proximal sixth aortic arch and persistence of the connection of the intrapulmonary-pulmonary artery to the distal sixth aortic arch. Some patients with isolated UAPA are completely asymptomatic; others may have severe pulmonary hypertension, pulmonary hemorrhage, congestive heart failure and cyanosis. Chest x-ray may show an absent hilar shadow, a shrunken affected lung, and a shift of the mediastinal structures to the affected side (2, 3). Subsequently, an echocardiography may confirm the diagnosis. Computerized tomography (CT) and magnetic resonance imaging (MRI) can show collateral arteries (1, 2, 4). Pulmonary artery pressure and collateral arteries can be revealed by cardiac catheterization (2, 3, 5, 6). In the present report, four cases with UAPA were discussed.

Case Reports

The demographic and clinical features of four cases are summarized in Table 1. Chest x-rays revealed absent hilar shadow and a shift of the mediastinal structures to the affected side (Fig. 1a). Echocardiography showed UAPA in all cases. Additional echocardiographic findings were coarctation of aorta (CoA) with patent ductus arteriosus (PDA) in case 1, abnormally partially venous return anomaly in case 2 and mitral valve prolapse (MVP) in case 4. Also, right aortic arch was showed in three of cases. Cardiac catheterization and imaging revealed UAPA (Fig. 1b) and many collateral arterial formations originating from descending aorta, common carotid artery and left subclavian artery (Fig. 2a, b, Fig. 3a). However, left lung of case 2 was not imaged, vertical vein was observed on MRI (Fig. 3b).

Discussion

The exact prevalence of UAPA is unknown, but the current literature estimates 1:200.000 individuals (1). In the literature, more than 300 cases have been reported since 2010 and absence of right pulmonary artery is more often than the left one (2-7). Recurrent pulmonary infections, decreased exercise tolerance and mild dyspnea during exertion are the most common symptoms (3, 7). Also, some patients with isolated UAPA can be presented with severe pulmonary hypertension, pulmonary hemorrhage, congestive heart failure and cyanosis. Eventually, in our cases, newborns with cyanosis had a severe form of the disease. So, we have suggested that the severity of disease may be associated with young age and the symptom of cyanosis.

Congenital heart defects that have been associated with UAPA are the followings; tetralogy of Fallot, ventricular septal defect (VSD), right aortic arch, truncus arteriosus, patent ductus arteriosus (PDA), CoA, subvalvular aortic stenosis, transposition of the great arteries and scimitar syndrome (2, 3, 5). Right pulmonary artery originating from ascending aorta is commonly associated with atrial septal defect (ASD) (8, 9). Also, left pulmonary artery originating from aorta associated with ASD was reported in the literature (8). Additionally, in a recent study, it was reported that the incidence of pulmonary hypertension was 86% among the patients with UAPA plus PDA and these patients died of

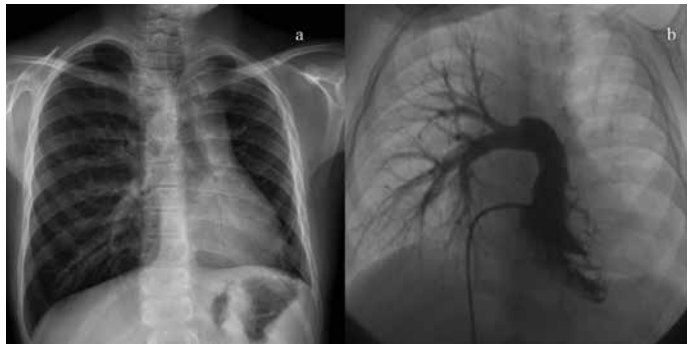

Figure 1. Decreased left pulmonary vascular frames and hemithorax volume, displacement of the left side of the thorax, hyperinflation of the right lung and deviation of trachea to the left side on chest X-ray (a) and absence of the left pulmonary artery on cardiac catheterization (b)

Table 1. Demographic and clinical features of cases

	Case 1	Case 2	Case 3	Case 4
Age	18-day-old	6-day-old	18-mount-old	11-years-old
Sex	Female	Female	Male	Male
Body weight	2.8 kg	2.7 kg	6.5 kg (< 3P)	38 kg
Complaints	Cyanosis	Cyanosis	Recurrent pulmonary infections	Chest pain and palpitation
Relativity of parents	None	Second degree	First degree	None
Oxygen saturation, %	65	76	92	93
Electrocardiography	Right axis	Right axis	Right axis	Normal axis
Absence of pulmonary artery	Right	Right	Left	Left
Chest X-ray	Decrease of the bronchovascular frames and deviation of trachea on the same hemithorax and increase of the aeration on the contra lateral			
Collateral arteries	-	DA	Common carotid artery and DA	LCA and DA
Right aortic arch	+	-	+	+
Additional echocardiographic findings	CoA, PDA, VSD, ASD	PAPVR	ASD	MVP, MR
Contralateral PAP, mmHg	60/10: mean 28	80/25: mean 60	50/20: mean 30	55/25: mean 35
Prognosis	Exitus	Being followed up in our clinic		

ASD - atrial septal defect, CoA - coarctation of the aorta, DA - descending aorta, LCA - left subclavian artery, MR - mitral regurgitation, MVP - mitral valve prolapse, PAP - pulmonary artery pressure, PAPVR - partial anomalous pulmonary venous return, PDA - patent ductus arteriosus, VSD - ventricular septal defect

pulmonary hypertension at early ages (7). Associated cardiac anomalies in our two cases (case 1 and 3) with ASD were CoA, PDA, VSD and right aortic arch respectively. Also, abnormal partially venous return anomaly was presented in case 2. In the literature the first reported patient with dextrocardia, absent of right pulmonary artery and MVP was an adult (10). Similarly in our case 4, absence of left pulmonary artery with MVP and moderate mitral regurgitation was detected as a first association in childhood.

On chest X-ray, ipsilateral grossly diminished pulmonary vascular markings, hemidiaphragmatic elevation, cardiac and mediastinal displacement, a small hemithorax, and contralateral lung hyperinflation can be detected. Absence of pulmonary artery can be showed by echocardiography. Anatomic details and distal pulmonary arteries can be visualized by MRI and high resolution CT. The presence of hilar arteries can be demonstrated by cardiac catheterization and pulmonary venous wedge angiography.

The incidence of pulmonary hypertension varies between 18-44% in isolated UAPA and it is much higher among patients who had UAPA with PDA (1-3, 5, 7, 8). This is because of that, the contralateral pulmonary vascular bed has insufficient elasticity to cope with the increased blood flow (4).

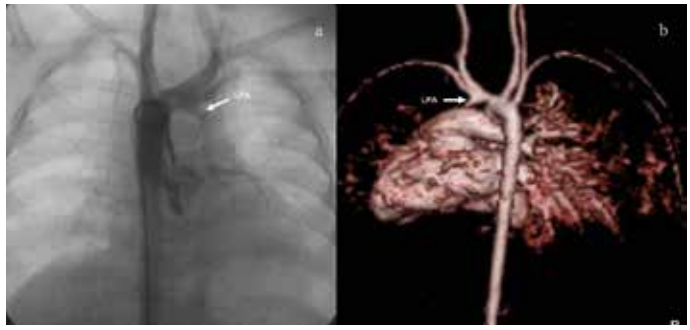

Figure 2. Left subclavian artery, common carotid artery and left pulmonary artery both originating from arcus aorta and right aortic arch views on cardiac catheterization (a) and cardiac magnetic resonance imaging (b)

LPA - left pulmonary artery

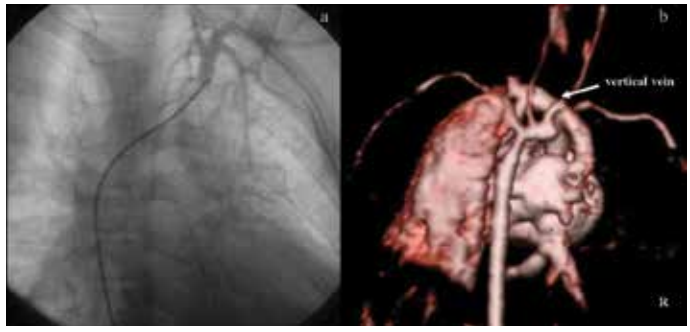

Figure 3. Multiple collateral arteries originating from left subclavian artery on cardiac catheterization (a) and abnormal partially venous return anomaly on the upper-right side of the lung with absence of left lung on cardiac magnetic resonance imaging (b)

Conclusion

Unilateral absence of a pulmonary artery should be considered in the patients manifested with cyanosis, recurrent respiratory infections or pulmonary hypertension. Early diagnosis can prevent further deterioration and associated comorbidities. Primarily, chest radiography and echocardiography are useful for the diagnosis of UAPA.

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Traumatic pulmonary valve hematoma; an unusual complication of pulmonary balloon valvuloplasty

Pulmoner balon valvuloplastinin nadir komplikasyonu; travmatik pulmoner kapak hematomu

Introduction

Pulmonary balloon valvuloplasty (PBV), initiated in 1982 by Kan et al. (1) was one of the first therapeutic procedures used catheters for the treatment of congenital heart disease. PBV is now recognized as the standard therapy for pulmonary valve stenosis (PS). Complications of PBV such as pulmonary regurgitation, annular laceration, pulmonary artery dissection, cardiac perforation have been reported. This is the presentation of the case with pulmonary valve hematoma that resulted as a complication following the treatment of PS after the application of PBV.

Case Report

A 16-year-old female patient was hospitalized with the diagnosis of isolated PS. She defined dyspnea, palpitations and fatigue on exertion that started one year before her admission. The auscultation of the patient unveiled a 4/6 grade pansystolic murmur which was best heard at the right and left second intercostal spaces. The chest roentgenogram showed mild cardiomegaly. Electrocardiogram demonstrated right ventricular hypertrophy. The remainders of the laboratory data were normal. Transthoracic echocardiography revealed PS with an 80 mm Hg gradient at the valvular level.

The pressure gradient between the right ventricle and the pulmonary artery was measured to be 70 mm Hg and pulmonary valve annulus was delineated as 18 mm by catheterization. PBV had been performed using 16 mm balloon valvuloplasty catheters with a length of 3 and 4 cm (PDC520-TH-70282010/04) by the pediatric cardiology. However, pulmonary valve gradient persisted after PBV in the echocardiography but any suspicious mass over the pulmonary valve or subpulmonary muscle hypertrophy was reported.

The patient underwent an operation one month after PBV. The heart was exposed through a midline sternotomy and cardiopulmonary bypass instituted. Pulmonary valve was inspected through the supravalvar vertical pulmonary arteriotomy incision. Unexpected, a red colored, fluctuating mass of 1.5 cm×2 cm in diameter was seen over the anterior semi lunar cusp (Fig. 1a). The thin external capsule of the mass was ruptured with the manipulation of the forceps and dark red colored liquid was discharged. A hard pearl like mass with the dimensions of 0.5×0.5×0.5 cm was occurred (Fig. 1b) with in the pouch. A valvotomy was made by the scalpel just next to the annulus but suitable size Hegar dilatator could not pass through the infundibular area, so the incision over the pulmonary artery was then extended towards the infundibular area of the right ventricular outflow tract (RVOT) until an adequate enlargement was obtained. Resection of the fibromuscular ridge and myectomy was performed from the infundibulum. The RVOT was closed with a diamond shaped (3×2 cm) Dacron patch. The rest of the operation was completed in uneventfully. No complication was encountered during the postoperative follow-up period. A pulmonary infundibular gradient of 10 mm Hg was measured with transthoracic echocardiography at the postoperative 5th day. Postoperative course was uneventful and the patient was discharged after 1 week.

Microscopic examination of the partially excised pulmonary valve showed fresh bleeding and myxoid degeneration areas (Fig. 2a). Histology of the pearl like mass demonstrated dense fibrosis (Fig. 2b).

Discussion

Balloon dilation of the pulmonary valve is currently considered the therapeutic modality of choice for the treatment of PS in any group and any valvular morphology (2). According to the previous studies, the independent predictors of long term result after BPV in pediatric patients are: 1) valve morphology; 2) ratio of balloon to annulus diameter; and 3) immediate post dilation pressure gradient through pulmonary

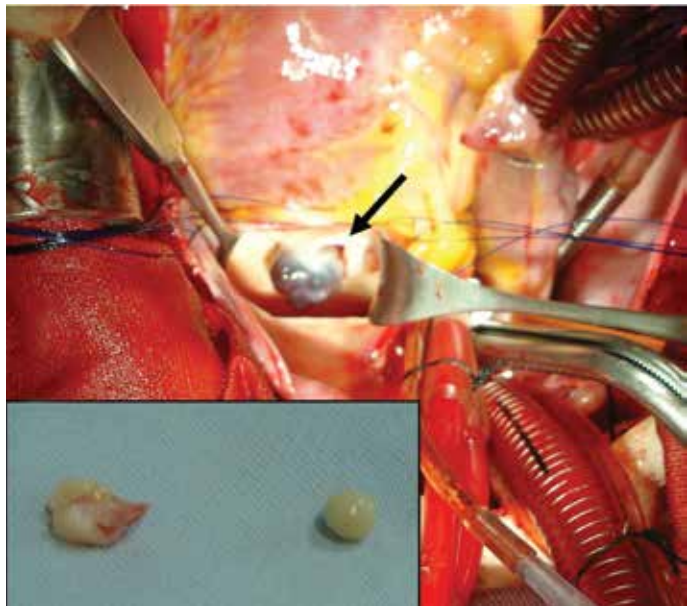

Figure 1. Intraoperative views: a) Arrow indicates red colored, 1.5 cmx2 cm in diameter, fluctuating mass over the anterior semilunar cusp through the supravalvar pulmonary vertical incision; b) Excised anterior semilunar pulmonary valve and hard fibrotic pearl like mass with the diameters of 0.5x0.5x0.5 cm