Left Pulmonary Artery Agenesis and Right Aortic Arch Presenting with Pulmonary Artery Hypertension: Original Image

Pulmoner Arteryel Hipertansiyon ile Başvuran Sol Pulmoner Arter Agenezisi ve Sağ Arkus Aorta Olgusu

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Unilateral pulmonary artery agenesis is a rare anomaly and has been reported at a rate of 1/200.000[1] and it is generally related with congenital heart defects such as tetralogy of Fallot, right aortic arch and septal defects. Absence of right pulmonary artery is seen more frequently than the left one.[2-6] Approximately 30% of patients do not have complaints.[1,7] When symptomatic, patients present with chest pain, pleural effusion, recurrent pulmonary infections, dyspnea or exercise limitation and hemoptysis.[6] Complications of the absence of pulmonary artery are massive hemoptysis, pulmonary hypertension, respiratory failure, right heart failure, bronchiectasis and include necrotizing bronchopneumonia.[7] A variety of methods such as computed tomography, magnetic resonance imaging, bronchogram, perfusion scintigraphy, echocardiography and cardiac catheterization can be used in diagnosis.[7] Here, we present a case of six-year old boy who was admitted to outpatient clinic with a complaint of fatigue, and then he was referred to our center due to pulmonary hypertension. His medical history included operation due to small perimembranous ventricular septal defect when he was 4-year-old and there was no catheter-angiography prior to the operation. Chest X-ray demonstrated a contracted left hemithorax with reduced lung volume, left mediastinal shift, hyperinflated right lung and marked dilation of the pulmonary conus (Figure 1). His echocardiography

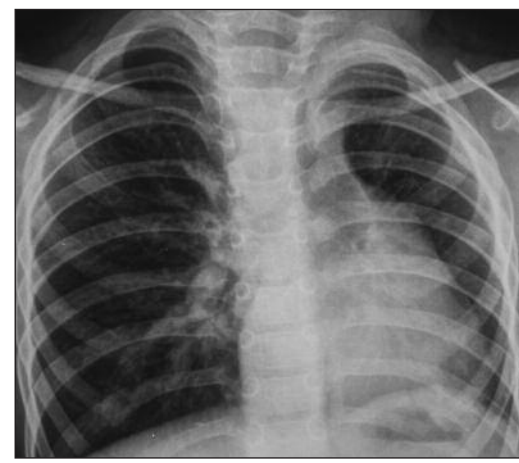

FIGURE 1: Chest X-ray demonstrated a contracted left hemitorax with reduced lung volume, left mediastinal shift, hyperinflated right lung and marked dilation of the pulmonary conus.

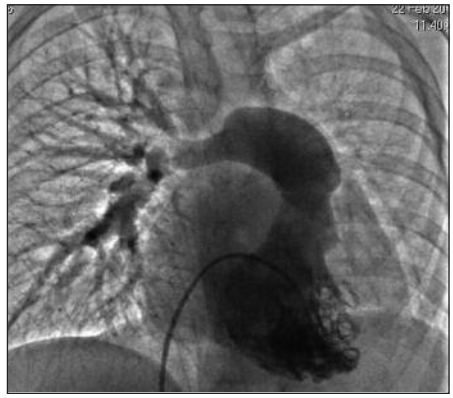

FIGURE 2: Right ventriculography demonstrated a large pulmonary artery which continues just with the right branch of pulmonary artery.

revealed right aortic arch, dilated right atrium and ventricle, enlarged main pulmonary artery with absence of left pulmonary artery. There was no residual shunt in the interventricular septum. Diagnostic catheter-angiography showed a large main pulmonary artery which continues just as the right branch of pulmonary artery (Video 1, Figure 2) and right aortic arch (Video 2, Figure 3). Mean pulmonary artery pressure was 38 mm Hg. Left lung parenchymal density was normal in high resolution computed tomography and there was subsegmental linear atelectasis at the posterior of lower lobe. Lung perfusion scintigraphy showed complete absence of left lung perfusion. In patients with isolated unilateral absence of pulmonary artery, the systemic collaterals to the affected lung come from the bronchial, intercostal, subclavian or subdiaphragmatic arteries.[8] Cardiac magnetic resonance angiography demonstrated absence of the left pulmonary artery and right aortic arch. There was no collateral artery branching from the aorta which supplied left lung (Figure 4). We supposed that left lung is supplied by the bronchial arteries. However, we didn't document such an association. Absence of pulmonary artery diagnosis is commonly not possible or has a delayed diagnosis in these patients due to asymptomatic course or lack of suspicion.[7] No treatment is mandatory for asymptomatic cases.[2,7] Surgical management can be performed in patients with accompanying cardiac anomalies. Selective embolization of systemic collaterals, or even pneumonectomy could be options for cases with massive hemoptysis. Revascularization is considered for selected cases.[2,7] Pulmonary artery hypertension either is still present immediately after surgery or has recurred several months or years after surgery in the absence of significant immediate postoperative residual lesions.[9] Long term vasodilator therapy may improve survival unless revascularization is performed. We did not perform surgery in our case, because there was no bronchiectasis on the hypoplasic lung and additional vascular anomalies. Treatment of pulmonary artery hypertension was initiated and patient was discharged for follow up.

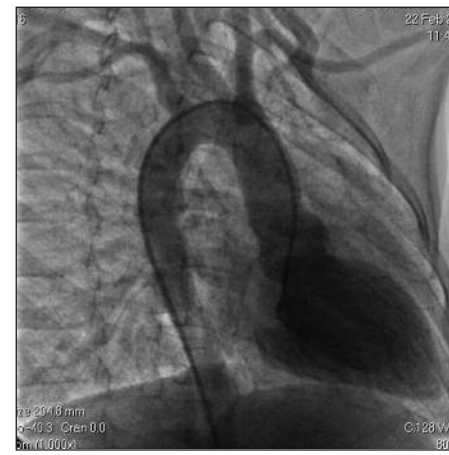

FIGURE 3: Left ventriculography showed right sided aortic arch and no collateral arteries originated from the aorta.

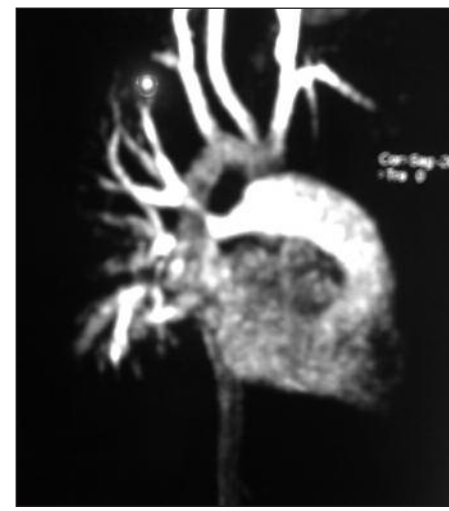

FIGURE 4: MR-angiography demonstrated absence of the left pulmonary artery and right aortic arch.

REFERENCES

1. Ten Harkel AD, Blom NA, Ottenkamp J. Isolated unilateral absence of a pulmonary artery: a case report and review of the literature. Chest 2002;122(4):1471-7.
2. Ergün P, Erdoğan Y, Türkkanı M, Gökçek A. [Isolated unilateral absence of a pulmonary artery: an unusual cause of recurrent pulmonary infection]. Turkiye Klinikleri J Arch Lung 2003;4(4):221-4.
3. Paç A, Ozme S, Celiker A, Ozkutlu S. Absent pulmonary valve syndrome with agenesis of the left pulmonary artery. Turk J Pediatr 1994;36(3):249-53.
4. Pool PE, Vogel JH, Blount SG. Congenital unilateral absence of a pulmonary artery. The importance of flow in pulmonary hypertension. Am J Cardiol 1962;10:706-32.
5. Karataş Z, Alp H, Altın H, Baysal T. Unilateral pulmonary artery agenesis: clinical and laboratory findings of four cases and diagnostic clues for pediatricians. Anadolu Kardiyol Derg 2012;12(7):603-5.
6. Bouros D, Pare P, Panagou P, Tsintiris K, Siafakas N. The varied manifestation of pulmonary artery agenesis in adulthood. Chest 1995;108(3):670-6.
7. Okutan O, Ugan H, Kaya H, Taş D, Demirer E, Apaydın M, et al. [Isolated unilateral absence of pulmonary artery: a case report]. Turkish Journal of Thoracic and Cardiovascular Surgery 2010;18(1):67-70.
8. Oakley C, Glick G, McCredie RM. Congenital absence of a pulmonary artery. Report of a case, with special reference to the bronchial circulation and review of the literature. Am J Med 1963;34:264-71.
9. Galiè N, Hoeper MM, Humbert M, Torbicki A, Vachiery JL, Barbera JA, et al. ESC Committee for Practice Guidelines (CPG). Guidelines for the diagnosis and treatment of pulmonary hypertension: the Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS), endorsed by the International Society of Heart and Lung Transplantation (ISHLT). Eur Heart J 2009;30(20):2493-537.