

Anomalous Origin of the Right Pulmonary Artery from Ascending Aorta Associated with Aortopulmonary Window and Interrupted Aortic Arch: Successful Surgical Correction with a Single 3D Patch

Çıkan Aortadan Anormal Kaynaklanan Sağ Pulmoner Arter, Aortopulmoner Pencere ve Kesintili Arkus Aorta Birlikteliği: Tek 3D Yama ile Başarılı Cerrahi Düzeltme

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ABSTRACT

Anomalous origin of the right pulmonary artery from the ascending aorta (AORPA) is a rare congenital heart defect and generally associated with other cardiac anomalies. Patients with AORPA have high pulmonary flow results in congestive heart failure and unilateral pulmonary hypertension. Thus, it is frequently a fatal malformation in the neonatal or early infancy periods if early surgical repair is not performed. Herein, we report a 1.5 month-old girl who was diagnosed with AORPA associated with aortopulmonary window and interrupted aortic arch and successfully operated using a single pericardial xenograft 3D patch.

Key Words: Anomalous origin of the right pulmonary artery from ascending aorta, Aortopulmonary window, Congenital heart defect, Interrupted aortic arch

ÖZET

Çıkan aortadan anormal kaynaklanan sağ pulmoner arter (ÇAAKPA) nadir görülen bir konjenital kalp defekti olup genellikle başka kalp anomalilerine eşlik eder. ÇAAKPA'lı hastalarda artmış pulmoner kan akımı sonucunda konjestif kalp yetmezliği ve tek taraflı pulmoner hipertansiyon gelişir. Bu nedenle cerrahi düzeltme yapılmayan hastalar genellikle yenidoğan ya da erken süt çocukluğu döneminde hayatını kaybederler. Bu yazıda, ÇAAKPA, aortopulmoner pencere ve kesintili arkus aorta birlikteliği saptanan ve tek 3D ksenograft yama kullanılarak başarılı bir şekilde opere edilen 1.5 aylık kız hasta sunulmaktadır.

Anahtar Sözcükler: Çıkan aortadan anormal kaynaklanan sağ pulmoner arter, Aortopulmoner pencere, Konjenital kalp hastalığı, Kesintili arkus aorta

INTRODUCTION

Anomalous origin of a branch pulmonary artery from ascending aorta in the presence of a main pulmonary artery arising separately from the heart is a quite rare congenital cardiac anomaly. This condition should be distinguished from other heart defects associated with an anomalous arterial supply to the lungs such as patent ductus arteriosus, major collaterals between the systemic and pulmonary circulation and truncus arteriosus (1). Herein, we report an infant who had anomalous

origin of the right pulmonary artery from ascending aorta (AORPA) associated with other complex congenital cardiac defects.

CASE REPORT

A 1.5-mo-old girl was admitted to our clinic with complaints of dyspnea and growth retardation. Her physical examination revealed poor weight gain (weight <3rd percentile), tachypnea

(respiratory rate: 92/min) and tachycardia (heart rate: 160/min). We also noticed suprasternal, intercostal and subcostal retractions and a grade 3/6 systolic murmur which was audible all over the precordium and hepatomegaly. Echocardiography revealed that the right pulmonary artery was originating from the posterior aspect of the ascending aorta while the left pulmonary artery was normally originating from the pulmonary trunk. Multislice computed tomographic (CT) angiography confirmed AORPA, aortopulmonary window, patent ductus arteriosus and interrupted aortic arch (Figure 1A,B). The blood flow of lower part of the body was supplied by a large (5 mm) ductus arteriosus. Cardiac catheterization revealed a pressure of 80/45 mmHg in the ascending aorta, 80/40 mmHg in the right pulmonary artery and 75/45 mmHg in the left pulmonary artery. Due to congestive heart failure symptoms, the patient was given medical therapy including furosemide, enalapril, digoxin and fluid restriction. Then she underwent cardiac surgery. During surgery, the ascending aorta was opened and directed to descending aorta by using a single pericardial xenograft patch with three different axes. Also, the aortopulmonary window was closed with this patch and the main pulmonary artery and branch pulmonary arteries were left behind the patch. The continuity of systemic blood flow was thus achieved, the systemic and pulmonary blood flows were separated and the right pulmonary artery was connected to main pulmonary artery by this single 3D patch (Figures 1A,B; 2A,B). Then, aortotomy was closed with a pericardial graft. The patient's postoperative course was uneventful. CT angiography which was performed one month after the operation revealed a moderate stenosis between posterior aspect of the patch and posterior wall of RPA, at the level of pulmonary bifurcation (Figure 2A,B). It resulted in 25 mmHg peak gradient by Doppler echocardiography without any hemodynamic disturbance. The

clinical symptoms and findings related to congestive heart failure resolved and she began to gain weight after the operation.

DISCUSSION

AORPA is more common than the left variant (AOLPA) and makes up 70-80% of these cases (2,3). The anomalous right PA usually arises from the posterior aspect of the ascending aorta close to the aortic valve. Less commonly, it originates from just proximal to the innominate artery or a patent ductus arteriosus (2,4). It is proposed that failure in leftward migration of the embryonic branch pulmonary artery to reach the main pulmonary artery portion after its joining the trunco-aortic sac occurs in patients with AORPA (2). Also, an association has been shown with DiGeorge syndrome in these patients. Our patient did not have other clinical and laboratory manifestations of this syndrome. However, we could not perform a genetic testing for 22q11.2 deletion syndrome in our patient in the perioperative period and she missed the follow-up later on.

AORPA is generally associated with other cardiac anomalies such as patent ductus arteriosus and aortopulmonary septal defect. While tetralogy of Fallot and aortic arch anomalies are more common in AOLPA (2), AORPA can be accompanied with interrupted aortic arch, as in our patient. Achieving an accurate diagnosis cannot be possible with echocardiography in some cases. CT angiography with 3D reconstruction is a preferable method for defining cardiovascular anatomy and the relationship between vascular structures. It is safer than conventional angiography, and also reduces radiation exposure and procedure time (5).

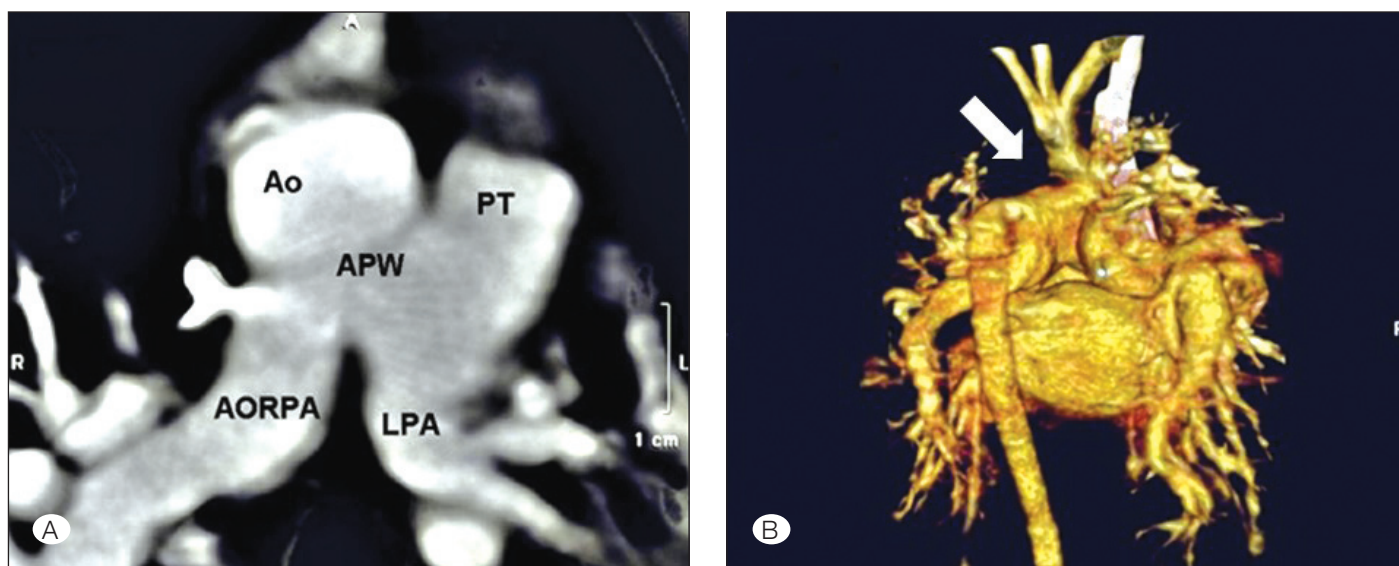


Figure 1: Pre-operative multislice CT images of the patient. **A)** showing AORPA and large aortopulmonary window. **B)** Interruption of the aortic arch (arrow). (**Ao:** ascending aorta, **AORPA:** Anomalous origin of the pulmonary artery from the ascending aorta, **APW:** aortopulmonary window, **LPA:** left pulmonary artery, **PT:** pulmonary truncus).

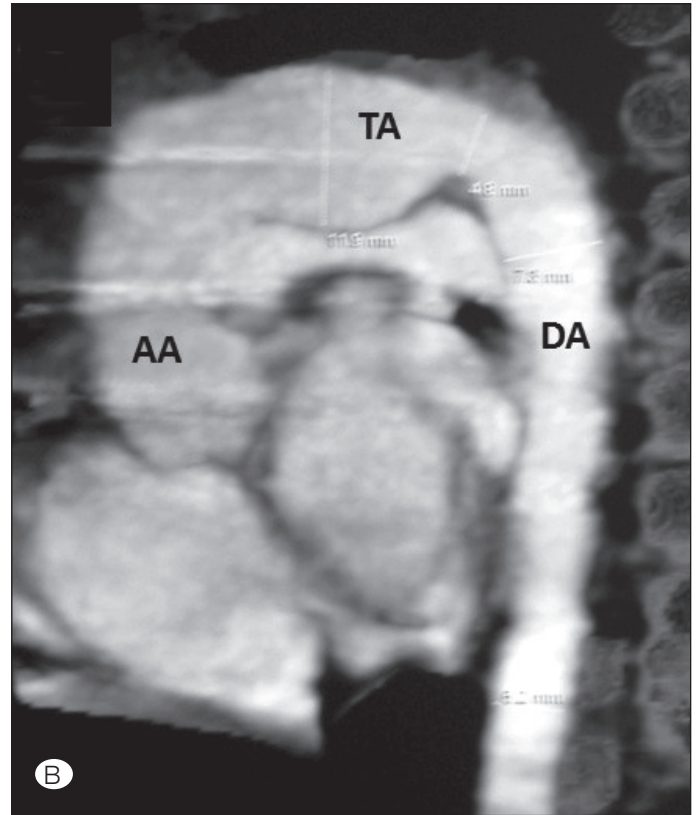
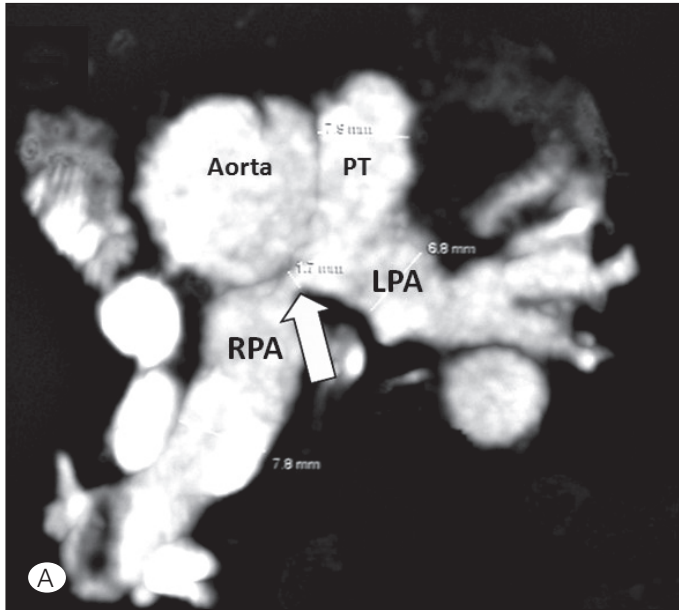


Figure 2: Postoperative CT findings of the patient. **A)** showing separation of systemic and pulmonary flows and RPA-main pulmonary artery connection (arrow indicates moderate RPA stenosis). **B)** showing ascending and descending aorta continuity via ductus arteriosus. (**AA:** Ascending aorta, **DA:** descending aorta, **LPA:** left pulmonary artery, **PT:** pulmonary truncus, **RPA:** right pulmonary artery, **TA:** transverse aorta).

While the lung associated with the normally originating PA receives the entire cardiac output from the right ventricle, the other lung is exposed to both pressure and volume overload due to shunting from the aorta in affected patients. Thus, congestive heart failure and pulmonary hypertension develops rapidly and it is frequently a fatal malformation in the neonatal or early infancy periods if early surgical repair is not performed. Similarly, a severe congestive heart failure occurred in our patient in the first month of her life and we observed a dramatic clinical response after surgery. Although direct implantation is the surgical procedure of choice whenever feasible, various surgical techniques have been described (implantation using a synthetic graft, autologous pericardial patch, and homograft interposition) (6). In our case, we preferred to use the patent ductus arteriosus because of its hard-textured wall. Thus, single 3D patch provided a pulmonary circulatory correction, continuity of ascending and descending aorta via patent ductus arteriosus and separation of systemic and pulmonary circulation as well.

In conclusion, AORPA is a rare but usually fatal congenital cardiac anomaly in the first year of the life. Although echocardiography can be sufficient for diagnosis, CT angiography provides a detailed demonstration of the defect and concomitant anomalies.

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