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Case Report / Olgu Sunumu

Surgical repair of non-obstructive supracardiac total anomalous pulmonary venous connection in an adult patient

Erişkin bir hastada obstrüktif olmayan suprakardiyak total pulmoner venöz dönüş anomalisinin cerrahi onarımı

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ABSTRACT

Total anomalous pulmonary venous connection is an uncommon congenital heart malformation with abnormal drainage of all pulmonary veins into the systemic venous system. Despite its very low incidence, it is usually a pediatric cardiac emergency and rarely allows survival into adulthood without surgical correction in infancy. Herein, we report one of the oldest cases from Turkey who was successfully operated for non-obstructive, supracardiac total anomalous pulmonary venous connection.

Keywords: Adulthood, non-obstructive, supracardiac, total anomalous pulmonary venous connection.

ÖZ

Total pulmoner venöz dönüş anomalisi, pulmoner venlerin tamamının anormal bir şekilde sistemik venöz sisteme döküldüğü, nadir bir doğuştan kalp anomalisidir. İnsidansı çok düşük olmasına rağmen, çocuklarda genellikle kardiyak acilidir ve bebeklik döneminde cerrahi düzeltme yapılmadığı taktirde, nadiren erişkinlik yaşına ulaşılabilir. Bu yazıda, obstrüktif olmayan, suprakardiyak total pulmoner venöz dönüş anomalisi nedeniyle başarılı bir şekilde ameliyat edilen Türkiye'nin en ileri yaş olgularından biri sunuldu.

Anahtar sözcükler: Erişkinlik dönemi, obstrüktif olmayan, suprakardiyak, total pulmoner venöz dönüş anomalisi.

Total anomalous pulmonary venous connection (TAPVC) accounts for 2% of all congenital heart defects and often requires the differential diagnosis in cyanotic newborns, children, and adults with pulmonary overcirculation.[1] Among the four types of TAPVC, the most common (~50%) type is the supracardiac type in which all pulmonary veins drain into a common pulmonary venous chamber (cPVC) that finally terminates in the superior vena cava (SVC). Identifying all pulmonary veins and confirming their drainage is one of the essential components for a complete congenital cardiac examination required for a successful surgical treatment for TAPVC in all ages.[2]

Herein, we report one of the oldest cases from Turkey who was successfully operated for non-obstructive, supracardiac TAPVC.

CASE REPORT

A 23-year-old male presented with dyspnea on moderate exertion for two months before his admission. He had a history of isolated interatrial septal defect (ASD) diagnosis at two years of age; however, he had no further follow-up until presentation. On physical examination, mild central cyanosis and clubbed fingers were present. The jugular venous pulse was prominent in the sitting position. There was a hyperdynamic right ventricular impulse, fixed-splitting of second heart sound, and a 2°/6° systolic ejection murmur along the left sternal border. There was also mild hepatomegaly. Chest X-ray revealed cardiomegaly and a widening of the superior mediastinum bilaterally, producing a "snowman" appearance (Figure 1). An electrocardiogram revealed sinus rhythm with

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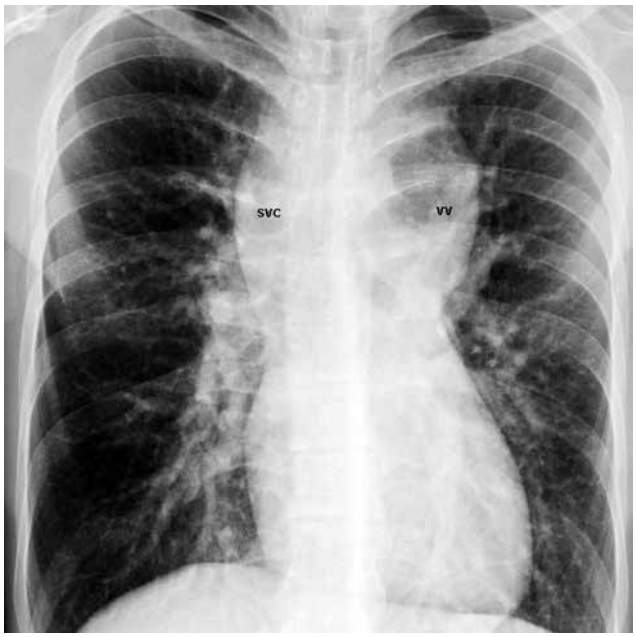

Figure 1. Snowman appearance on chest X-ray.
SVC: Superior vena cava; VV: Vertical vein.

incomplete right bundle branch block. Arterial blood gas analysis revealed 80% oxygen saturation in room air. Laboratory tests were normal except for 17.8 mg/dL hemoglobin and transaminase levels (serum aspartate aminotransferases/alanine aminotransferases) 43/37 IU. Transthoracic echocardiogram revealed enlarged right atrium (RA) and right ventricle (RV) with a non-restrictive (48 mm) ASD. The pulmonary artery trunk was quite dilated. Pulmonary artery pressure was 25/15 mmHg. The pulmonary veins were not thoroughly visualized. Exact anatomy of the pulmonary veins was unable to be obtained by cardiac catheterization either. Three-dimensional images of computed tomographic angiography (CTA) of the chest (Figure 2) revealed that cPVC, collecting all pulmonary veins, was located behind the left atrium (LA) from which a vertical vein (VV) originated to reach the brachiocephalic vein (BCV) terminating into the SVC. Systemic and the pulmonary veins were quite dilated. The small LA did not receive any pulmonary vein.

A written informed consent was obtained from the patient. He underwent an elective surgery through a median sternotomy. During the operation, following aorto-bicaval cardiopulmonary bypass (CPB), the patient was cooled to 30°C. The aorta was cross-clamped and cardiac arrest was induced with antegrade cold cardioplegic solution. The heart was retracted to the cephalad and right, exposing the left posterior pericardial area. The VV and the cPVC were found behind the pericardium, adjacent to the back of the LA.

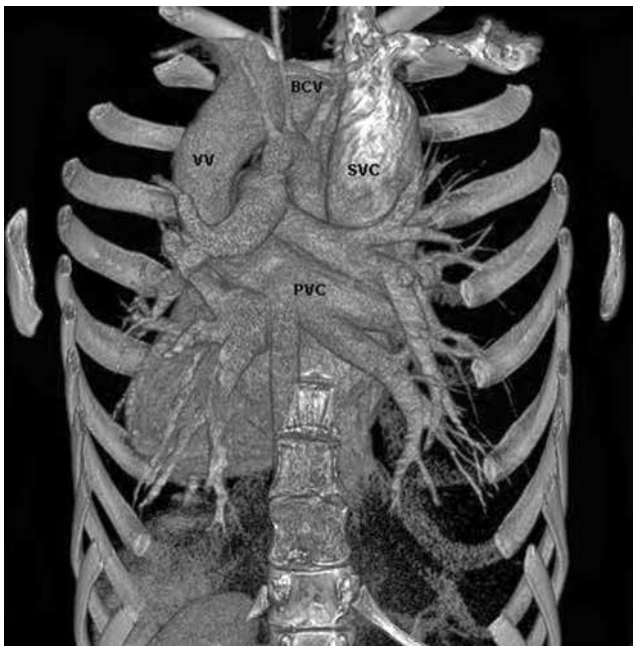

Figure 2. Anomalous connection of pulmonary veins with systemic vein.
SVC: Superior vena cava; VV: Vertical vein; BCV: Brachiocephalic vein; PVC: Pulmonary venous chamber.

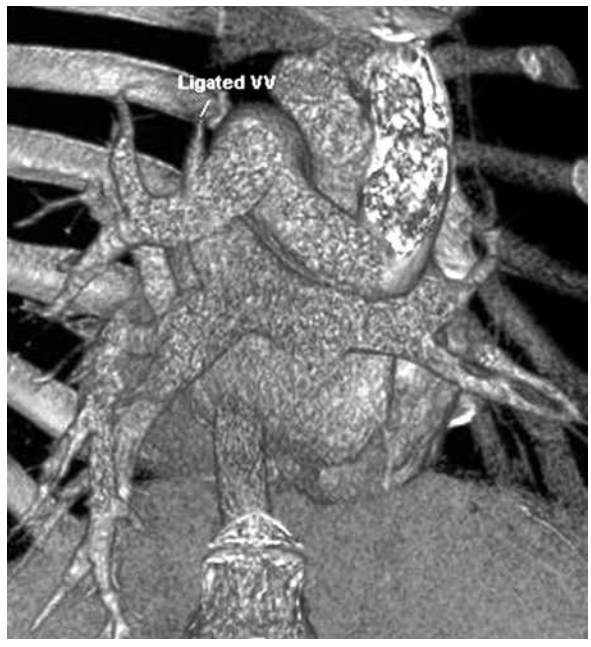

Figure 3. A postoperative three-dimensional, computed tomography angiography three-dimensional image. Ligated vertical vein.
VV: Vertical vein.

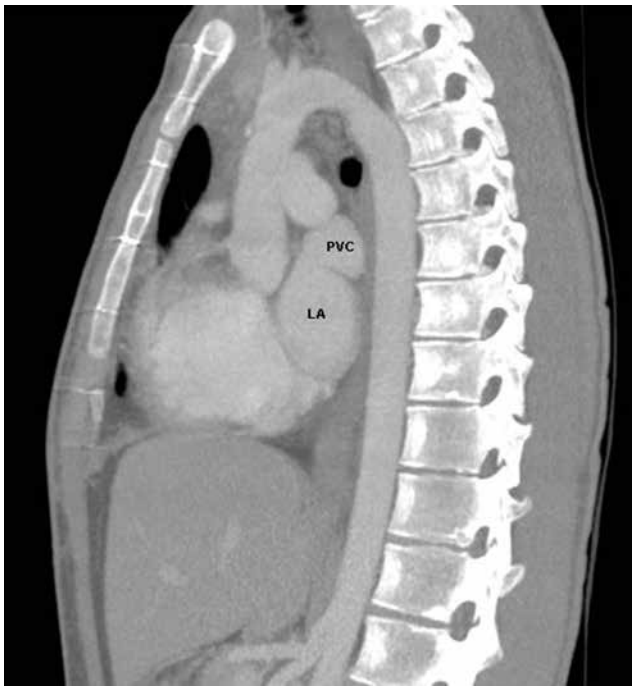

Figure 4. A postoperative three-dimensional, computed tomography angiography image. LA anastomosis with pulmonary venous chamber.

PVC: Pulmonary venous chamber; LA: Left atrium.

A longitudinal incision was made into both anterior wall of the cPVC and the posterior wall of the LA. Extended side-to-side anastomosis was constructed with continuous suture technique. The interatrial defect was closed with a Dacron patch (50×20 mm). After weaning from CPB, hemodynamic stability allowed us to ligate the ascending VV. Postoperative course was uneventful. He was discharged seven days after the operation. Control thoracic CTA one year following the operation revealed ligated VV (Figure 3) with a large anastomosis leading the cPVC to the LA (Figure 4).

DISCUSSION

Most patients with TAPVC have symptoms during the first year of life, and 80% die before one year of age, if not treated. One of the two main factors responsible for survival beyond infancy and childhood of non-operated patients is a large ASD which prevents pulmonary overcirculation and regulates the cardiac output and systemic oxygenation, and the other one is a non-stenotic pulmonary venous drainage which prevents pulmonary hypertension.

In patients with non-obstructive TAPVC, pulmonary arterial pressure may be normal. The shear effect of the high blood flow on the pulmonary endothelium is similar to that observed in patients with large ASDs. Although muscular arteries and arterioles often have prominent medial hypertrophy, many patients do not develop significant pulmonary vascular obstructive disease even in adulthood. The oldest reported patient in the literature with a TAPVC repair was 57 years old.[3] This patient is one of the oldest patients operated for TAPVC in Turkey.[4]

Echocardiography is the first-line choice for the diagnosis of isolated TAPVC. The pulmonary venous confluence can be sometimes difficult to visualize and patients can be easily misdiagnosed with isolated ASD, unless TAPVC is suspected. The reconstructed three-dimensional CTA images revealed important information and was a guide for planning of the surgical treatment in our patient showing the precise size, site, and distance between the cPVC and LA.

Surgical repair is the only treatment for TAPVC, although it may be hazardous in adults due to the large and friable pulmonary and systemic veins. The main goal of surgery is to provide a non-obstructive route for pulmonary venous drainage into the LA, closure of ASD, and removal of the alternative routes of pulmonary venous drainage. There are two anatomic exposure techniques for restoring the normal relationship in supracardiac TAPVC: the posterior and the biatrial (lateral) approach. In our case, we preferred the posterior one, as it is safe, effective, and provides excellent exposure without associated late atrial arrhythmias.[5]

On the other hand, whether the VV is to be ligated is still controversial. Some authors prefer to leave the ascending VV patent to decompress the left side of the heart or to take oxygenated blood to the liver, respectively.[6] In addition, following rerouting the pulmonary veins to the LA, most VVs may close spontaneously due to high resistance, and interventional therapy can be used on those which stay open in the late postoperative phase. In our case, we ligated the VV.

In conclusion, adult patients with an atrial septal defect and cyanosis require a detailed examination to exclude total anomalous pulmonary venous connection in whom the closure of the defect would end with mortality. The survival of our patient to this age can be attributed to non-obstructive, supracardiac pathology with a large atrial septal defect; however, the mechanism which protects the overflowed and congested lung from pulmonary hypertension is still unclear. Increasing age an unfavorable factor for successful surgical outcomes. However, careful perioperative assessment of this small group of patients with multiple imaging modalities would exclude anatomical and physiological factors which adversely affect the outcome.

Declaration of conflicting interests

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