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Case Report / Olgu Sunumu

A case of mixed drainage total anomalous pulmonary venous return in an adult patient

Erişkin bir hastada miks drenaj total anormal pulmoner venöz dönüş olgusu

Murat Meriç, Serkan Yüksel

Department of Cardiology, Medicine Faculty of Ondokuz Mayıs University, Samsun, Turkey

ABSTRACT

Total anomalous pulmonary venous return is observed in 1.5 to 3% of congenital heart diseases. Urgent surgical intervention is required following the diagnosis, since severe heart failure is accompanied by cyanosis from the first days of life in most of the patients. This rare congenital heart disease is often managed in adulthood with surgery. Patients who survive until adulthood without a surgical intervention have a large atrial septal defect. A 36-year-old female patient was admitted with complaints of fatigability, shortness of breath, and cyanosis of the lips. Total anomalous pulmonary venous return, secundum-type atrial septal defect, ventricular septal defect, and pulmonary hypertension were diagnosed by transthoracic echocardiography and catheter angiography. After medical treatment, surgery was planned for the patient who responded to the pulmonary vasoreactivity test with iloprost.

Keywords: Adult congenital heart disease; atrial septal defect; pulmonary hypertension; total anomalous pulmonary venous return; ventricular septal defect

ÖZ

Total anormal pulmoner venöz dönüş, doğuştan kalp hastalıklarının %1.5-3'ünde görülür. Hastaların çoğunda hayatın ilk günlerinden itibaren siyanoza ciddi kalp yetmezliği eşlik ettiği için, tanıyı takiben acil cerrahi girişim gerekmektedir. Bu nadir doğuştan kalp hastalığı, genellikle erişkin dönemde cerrahi ile tedavi edilir. Cerrahi girişim olmaksızın erişkin yaşa kadar ulaşan hastalarda büyük bir atriyal septal defekt mevcuttur. Otuz altı yaşındaki kadın hasta çabuk yorulma, nefes darlığı ve dudaklarda morarma yakınmaları ile başvurdu. Transtorasik ekokardiyografi ve kateter anjiyografide total anormal pulmoner venöz dönüş, sekundum tip atriyal septal defekt, ventriküler septal defekt ve pulmoner hipertansiyon saptandı. İloprost ile yapılan pulmoner vazoreaktivite testine yanıt veren hastaya tıbbi tedavi sonrasında cerrahi planlandı.

Anahtar sözcükler: Erişkin doğuştan kalp hastalığı; atriyal septal defekt; pulmoner hipertansiyon; total anormal pulmoner venöz dönüş; ventriküler septal defekt.

Total anomalous pulmonary venous return (TAPVR) is one of the rare congenital heart anomalies characterized by the opening of all four pulmonary veins into the right atrium, or into one or more venous formations associated with the right atrium. It is seen in 1.5 to 3% of congenital heart diseases.[1] Total anomalous pulmonary venous return occurs in four different forms according to the drainage site of the pulmonary veins: supracardiac, cardiac, infracardiac, and mixed type. The most frequent type is supracardiac (45 to 50%), followed by cardiac (25 to 30%), infracardiac (15 to 20%), and mixed type (5%). The mixed type appears to be the combination of the three other types.[1] Cyanosis and severe heart failure are observed in most of the patients from the first days of life. Obstruction of pulmonary venous return is the most important factor that identifies the severity of the symptoms. There is a limited number of reports in the literature for this congenital heart disease, which is quite rare in adulthood.[2-5] In this article, we present a mixed drainage TAPVR case, who reached the adulthood.

CASE REPORT

A 36-year-old female patient was referred to cardiology clinic with complaints of early fatigability, shortness

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Correspondence: Murat Meriç, MD. Ondokuz Mayıs Universitesi Tıp Fakültesi, Kardiyoloji Anabilim Dalı, 55280 Kurupelit, Samsun, Turkey.
Tel: +90 362 - 312 19 19 e-mail: drmeric@hotmail.com

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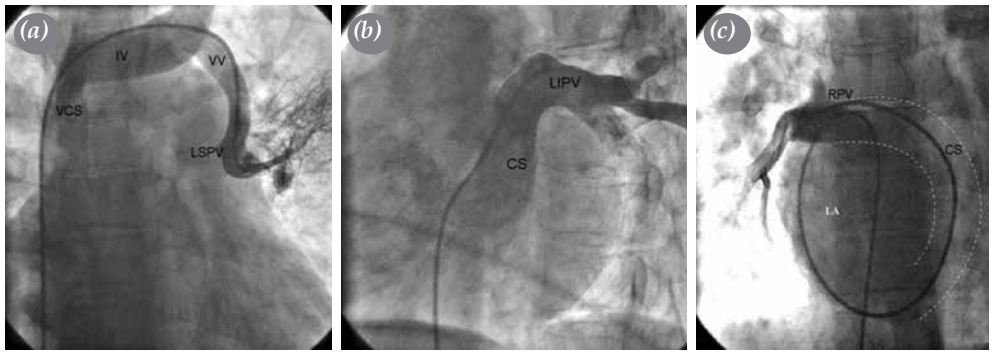

Figure 1. (a) An anteroposterior view showing flow of left superior pulmonary vein into innominate vein via vertical vein after selective injection into left superior pulmonary vein. **(b)** A left anterior oblique view showing flow of left inferior pulmonary vein into coronary sinus after selective injection into the inferior pulmonary vein. **(c)** Flow of right pulmonary veins into coronary sinus after selective injection into right pulmonary veins.

IV: Innominate vein; VCS: Vena cava superior; VV: Vertical vein; LSPV: Left superior pulmonary vein; LIPV: Left inferior pulmonary vein; CS: Coronary sinus; RPV: Right pulmonary veins; LA: Left atrium.

of breath, and cyanosis of the lips. On physical examination, central cyanosis and tachypnea were present. The oxygen saturation was 89%, the arterial blood pressure was 100/65 mmHg, and the right ventricular heave was prominent on palpation. On auscultation, S2 was accentuated and there was a 2/6 systolic murmur best heard in left third intercostal space. Examination of other systems revealed normal findings. Cardiomegaly and an increase in pulmonary vascularity were detected on telecardiography. Electrocardiography revealed a right axis deviation and right ventricular hypertrophy. On echocardiographic examination, the right atrium, right ventricle, and main pulmonary artery were large with right ventricular hypertrophy. The pulmonary artery systolic pressure, which was calculated from tricuspid insufficiency using the Bernoulli equation, was 80 mmHg. The coronary sinus was enlarged and the blood flow was increased in the apical four-chamber and the parasternal long axis views. A large secundum type atrial septal defect

Table 1. Cardiac catheter findings

Measured from	O_2 saturation (%)		Pressure	
	Before ilomedin	After ilomedin (10^{th} min)	Before ilomedin	After ilomedin (10^{th} min.)
Superior vena cava	80	82		
Right atrium	82	89	11	11
Inferior vena cava	72	75		
Right ventricle	83	84	133/0-3	
Pulmonary artery	88	91	126/57-85	101/40-60
Aorta	92	94	136/73-92	114/72-88
Pulmonary wedge			16	13

Table 2. Flow/resistance ratios during catheterization

Flow/resistance	Qp	Qs	Qp/Qs	Pulmonary resistance (Rp)	Systemic resistance (Rs)	Rp/Rs
Before iloprost	14.5	7.9	1.8	7.24	14.0	0.51
After iloprost	20.0	10.0	2.0	4.10	13.4	0.30

Qp: Pulmonary flow; Qs: Systemic flow; Rp: Pulmonary resistance; Rs: Systemic resistance.

(ASD) and a small muscular ventricular septal defect (VSD) were shown. The pulmonary vein related to the left atrium was not observed in the subcostal and apical four-chamber positions. On cardiac catheterization and angiography, it was observed that a vertical vein was filled after the injection to the innominate vein in anteroposterior projection, and the left superior pulmonary vein drained into the innominate vein via the vertical vein. By accessing through the coronary sinus, three pulmonary veins (two right pulmonary veins and one left inferior vein) were combined and opened into the coronary sinus (Figure 1). Pulmonary artery pressure was at a systemic level, and catheter findings and the flow/resistance ratio before and after the vasoreactivity test with iloprost are summarized in Table 1 and 2. Following medical treatment, the patient was scheduled for surgery. A written informed consent was obtained from the patient.

DISCUSSION

Total anomalous pulmonary venous return is a congenital anomaly which leads to cyanosis due to the complete mixture of systemic and pulmonary venous blood.[1] In all types of TAPVR, all of the systemic and pulmonary venous returns finally come to the right atrium. A part of the systemic and pulmonary venous return passes into the pulmonary circulation, and the other part passes into the systemic circulation via an ASD or patent foramen ovale.[6] If the ASD is large, the distribution of pulmonary venous blood into the pulmonary and systemic circulations depends on the compliances of the right and left atria and ventricles and systemic and pulmonary vascular resistances.[2-5] If the ASD is restrictive, the right atrium is loaded and systemic and pulmonary venous congestion occurs.[1-6] In the obstructive type of TAPVR, pulmonary venous hypertension, pulmonary edema, pulmonary hypertension, and right heart failure occur.[1-6] Obstruction is due to the restrictive type of ASD or compression of the channels of the anomalous pulmonary venous return. The incidence of venous obstruction is the highest in the infracardiac type and the lowest in the cardiac type.[6] The hemodynamic condition depends on the amount of pulmonary blood flow and resistance, the size of the ASD, and the degree of pulmonary hypertension. The majority of the patients, who maintain their lives without surgical treatment, have large ASDs, as in our case. The physical examination findings are very similar with large ASDs. Mild cyanosis and exercise intolerance are observed. As in our case, mild alterations in pulmonary vascular resistance, blood flow, and arterial oxygen can be also observed in patients who survive until the age of 20 years.[7] Feng et al.[8] and Ogawa et al.[9] reported similar cases with TAPVR and large ASDs. The authors speculated that survival benefit of their patients was due to large ASD or broad pulmonary venous confluence. Similarly, in our case, a large ASD combined with a small muscular VSD might lead to survival benefit and less prominent symptomatology in her young periods of life. Over time, as compliance of the right ventricle decreases, right-to-left shunting across the ASD and VSD gradually increases, leading to symptoms (i.e., cyanosis or dyspnea).

The exact diagnosis of TAPVR is made by selective pulmonary angiography along with oxygen saturation measurements. In addition, it has been shown that magnetic resonance imaging angiography can be also a reliable method for the diagnosis of TAPVR. In the present case, we performed catheter angiography to determine the indications for surgery due to the presence of a large ASD and VSD in addition to pulmonary hypertension symptoms. After oral bosentan therapy, surgical repair was planned for our case, in whom pulmonary hypertension was detected during catheterization and who responded to the vasoreactivity test using iloprost.

In conclusion, the onset of symptoms, diagnosis, and treatment may be delayed in patients with total anomalous pulmonary venous return due to the anatomical characteristics of this anomaly. In selected patients, if surgical repair is performed in proper settings, it yields successful results and good prognosis.

Declaration of conflicting interests

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