Surgical Correction in A Late Presentation of Mixed-Type of Total Anomalous Pulmonary Venous Drainage

Taufik Nur Yahya1*, Arief Rakhman Hakim2, Heroe Soebroto2, Erdyanto Akbar2

1Resident of Thoracic, Cardiac, and Vascular Surgery, Universitas Airlangga / Soetomo Hospital, Surabaya, Indonesia
2Attending of Department of Thoracic, Cardiac, and Vascular Surgery, Universitas Airlangga / Soetomo Hospital, Surabaya, Indonesia

Corresponding author. Email: taufiknuryahya.md@gmail.com

ABSTRACT

Total anomalous pulmonary venous drainage (TAPVD) is a rare congenital heart disease, characterized by the failure of all four pulmonary veins to form a direct connection to the left atrium; instead, they drain into the right heart through different routes of systemic venous return. The incidence of TAPVD ranges from 0.6 to 1.2 per 10,000 live births. A 14-year-old male presented with dyspnea and leg edema on moderate exertion. Physical examination showed cyanosis, clubbing finger, and systolic murmur. Echocardiography showed situs solitus, presence of innominate vein, presence of persistent left superior vena cava, pulmonary veins drained to coronary sinus and superior vena cava, secundum atrial septal defect, and moderate tricuspid regurgitation. Cardiac catheterization and cardiac computed tomography confirmed the diagnosis of mixed-type TAPVD by demonstrating the left superior pulmonary vein drained to superior vena cava through vertical vein while three other pulmonary veins to coronary sinus. Surgery was performed under general anaesthesia, median sternotomy approach, and bicaval to ascending aorta cardiopulmonary bypass. Vertical vein was ligated, transected, and anastomosed to left atrial appendage. Coronary sinus was unroofed and rerouted to left atrium. Atrial septal defect was closed with pericardial patch. The procedure took aortic cross-clamp time of 31 minutes and cardiopulmonary bypass time of 110 minutes. The patient was then transferred to the intensive care unit with stable hemodynamic. The patient was discharged 3 days after surgery. TAPVD requires urgent surgical correction within the first few months of life due to a high mortality rate if left untreated. This case is unique because the patient had been asymptomatic until he entered his teenage years. TAPVD can present late, as in our case, in the setting of a widely patent atrial septum without obstruction to pulmonary venous return. The objective of this study is to demonstrate a good result in surgical correction of mixed-typed TAPVD despite late repair.

Keywords— total anomalous pulmonary venous drainage, mixed type, congenital heart disease

INTRODUCTION

Total anomalous pulmonary venous drainage (TAPVD) is a rare congenital heart disease, which occurred in 0.6 to 1.2 per 10,000 live births [1]. TAPVD is characterized by the failure of all four pulmonary veins (PVs) to form a direct connection to the left atrium (LA); instead, they drain into the right heart through different routes of systemic venous return. An atrial septal defect (ASD) is usual-

ly present to send blood from RA to LA and LV. Darling et al, proposed a classification of TAPVD into four types [2]:

Type I supracardiac (45-55% of total cases): four pulmonary veins drain to the superior vena cava or its tributaries

Type II intracardiac (20-30% of total cases): pulmonary veins empty directly into right atrium, typically via coronary sinus

Type III infracardiac (13-25% of total cases): pulmonary veins connect to a vertical vein that runs through the diaphragm and eventually drain to the systemic venous system, such as the portal or inferior vena cava.

Type IV mixed (5% of total cases): characterized by having at least two of the above types

Physiologically, TAPVD is divided into two types: obstructive and nonobstructive. The division depends on the presence or absence of an obstruction to the pulmonary venous return. The infracardiac type is usually obstructive while the majority of the cardiac and supracardiac types are non-obstructive [3].

Case Report

A 14-year-old male presented with dyspnea and leg edema on moderate exertion. The symptoms exacerbated with physical activity, especially when he played football, and improved with resting. The symptoms were felt about one year prior to hospital admission. The patient had a past medical history of recurrent respiratory tract infection and delayed walking. At admission, the physical examination showed cyanosis, clubbing finger, and systolic murmur.

Laboratory findings were within normal limits while posteroanterior chest x-ray suggested excessive pulmonary blood flow and heart enlargement. Echocardiographic examination showed situs solitus, AV-VA concordance, presence of innominate vein, presence of left superior vena cava (PLSVC), PVs drained to coronary sinus (CS) and superior vena cava (SVC), absence of patent ductus arteriosus (PDA), dilatation of right atrium (RA) and right ventricle (RV), secundum atrial septal defect (ASD) with diameter of 1,62 cm, mild mitral regurgitation, and moderate tricuspid regurgitation with pressure gradient of 37,43 mmHg. Cardiac catheterization and cardiac computed tomography (Fig. 1) confirmed the diagnosis of mixed type TAPVD by demonstrating the left superior PV drained to SVC through vertical vein while three other PVs to CS, in addition to presence of innominate vein and PLSVC. Gradients between RA/LA and RV/PA were 2 mmHg and 7 mmHg, respectively. Pulmonary vascular resistance index was calculated at 2,99 WU.m2. The pulmonary-to-systemic flow ratio is 1.17.

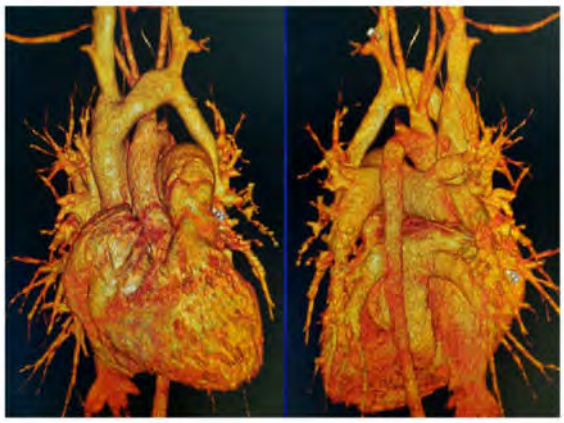

Figure 1. Cardiac computed tomography demonstrates the left superior PV drained to SVC through vertical vein while three other PVs to CS; also shown the presence of innominate vein and PLSVC

The aim of the surgery is to redirect all pulmonary veins to the LA through wide and nonrestrictive connection. Surgery was performed under general anaesthesia, median sternotomy approach, bicaval to ascending aorta cardiopulmonary bypass (CPB), antegrade cardioplegia and superficial cooling.

Vertical vein was ligated, transected, and anastomosed to left atrial appendage (LAA). CS was unroofed and rerouted to LA. ASD was closed with a pericardial patch (Fig. 2). The procedure took aortic cross-clamp time of 31 minutes and CPB time of 110 minutes. After surgery, the patient was transferred to the intensive care unit (ICU) with stable hemodynamic. The postoperative care was uneventful. The patient was discharged on post operative day 3.

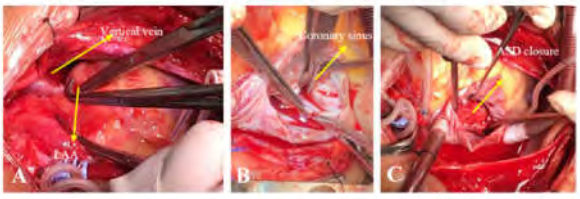

Figure 2. (A) Vertical vein anastomosed to LAA; (B) Unroofing CS; (C); ASD closure with pericardial patch

Discussion

Isolated TAPVD is incompatible with life. It requires ASD coexistence to allow oxygenated blood supply to the left chambers. The smaller the ASD, the higher the RA pressure and the lower the blood supply to the LA and the cardiac output [4].

Surgical repair should begin as soon as the diagnosis is made. Pulmonary venous drainage is usually obstructed in type I and III TAPVD. Because there is no conservative palliative treatment for pulmonary edema caused by high capillary pressure, severe hypoxia and metabolic acidosis occur, this condition needs a surgical emergency in the neonate [5].

For type II TAPVD, which are generally non-obstructive forms of TAPVD, surgery can be elective in the neonatal phase or during the first six months of life before irreversible pathological changes occur in the pulmonary vascular bed. In these patients, a hemodynamic study was indicated mainly to evaluate the physiology of the vascular pulmonary bed and to monitor the echocardiographically inconclusive course of one of the pulmonary veins [5].

Mixed-type TAPVD is the least common subtype, accounting for approximately 5% of all patients with TAPVD. Furthermore, due to the unpredictability of the pulmonary venous connection, mixed TAPVD is the most difficult form and requires a combination of techniques including individualized anastomosis of the pulmonary veins. To successfully manage highly variable anatomical pulmonary vein connections, an individualized operation must be designed. Therefore, the first questions to be addressed are a correct preoperative understanding of the anatomy and an accurate description of the abnormally draining pulmonary veins [6].

In our case, the patient was asymptomatic until adolescence. Lack of abnormal drainage blockage, including significant atrial septal defect and maintenance of vascular resistance, is an important factor contributing to the long-term life of our patients. Similar cases where TAPVD patients were asymptomatic due to the absent of obstruction had been reported before. In those cases, the patients survived the surgery [5][7].

Surgical correction of mixed type TAPVD was performed as a combination of techniques, including the individualized anastomosis of pulmonary veins. As in our case, the surgery consisted of ligation and transection of vertical vein, which was then anastomosed to LAA and unroofing and rerouting CS to LA. ASD then was closed with pericardial patch. The procedure took aortic cross-clamp time of 31 minutes and CPB time of 110 minutes. For surgeons familiar with the correction of TAPVD in neonates, surgical treatment in older patients shows that it is not more technically difficult and can pro-

vide excellent long-term survival [8].

Conclusion

Typically, TAPVD requires urgent surgical correction within the first few months of life due to a high mortality rate of 80% by 1 year, if left untreated. This case is unique because the patient had been asymptomatic until he entered his teenage years. TAPVD can present late, as in our case, in the setting of a widely patent atrial septum without obstruction to pulmonary venous return. In this case, the prognosis is better despite late repair.

References

1. M. D. Reller, M. J. Strickland, and T. Riehle-Colarusso, "Prevalence of congenital heart defects in metropolitan Atlanta, 1998-2005". J Pediatr, no. 6, pp. 807-813, December 2008.
2. R. C. Darling, W. B. Rothney, J. M. Craig, "Total pulmonary venous drainage into the right side of the heart: report of 17 autopsied cases not associated with other major cardiovascular anomalies." Lab Invest, no. 6, pp. 44-64, January 1957.
3. M. K. Park and M. Salamat, Park's Pediatric cardiology for practitioners, Elsevier Health Sciences, 2020
4. F. A. Atik, P. E. Irun, M. Barbero-Marcial, E. Atik, "Total anomalous drainage of the pulmonary veins - Surgical therapy for the infradiaphragmatic and mixed anatomical types". Arq Bras Cardiol, no 82, pp. 259-263, 2004
5. W. V. D. A. Vicente, P. S. Dias-da-Silva, L. D. M. Vicente, S. Bassetto, M. M. D. Romano, C. A. Ferreira, and A. J. Rodrigues, "Surgical correction of total anomalous pulmonary venous drainage in an adult". Arquivos brasileiros de cardiologia, no. 87(5) pp. 172-175, November 2006
6. M. Xiang, C. Wu, Z. Pan, Q. Wang, and L. Xi.. Mixed type of total anomalous pulmonary venous connection: diagnosis, surgical approach and outcomes. Journal of Cardiothoracic Surgery, No. 15, pp 1-7, December 2020.
7. M. Thawabi, S. Studyvin, A. Hawatmeh, R. Verma and M. Cohen, "Late presentation of total anomalous pulmonary venous connection". Journal of the American College of Cardiology, No. 69, pp 2235-2235, March 2017.
8. A. Serraf, E. Belli, D. Roux, M. Sousa-Uva, F. Lacour-Gayet, C. Planché, "Modified superior approach for repair of supracardiac and mixed total anomalous pulmonary venous drainage". Ann Thorac Surg. No. 65, pp. 1391-1393, May 1998.