Case Report / Olgu Sunumu

Repair of a Partial Anomalous Pulmonary Venous Connection After Mitral Valve Replacement: A Case Report

Mitral Kapak Replasmanı Sonrası Kısmi Pulmoner Venöz Dönüş Anomalisinin Tamiri: Olgu Sunumu Kısmi Pulmoner Venöz Dönüş Anomalisi

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Abstract

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital disorder with an incidence 0.5% of all congenital cardiac defects. Usually seen with congenital cardiac defects such as sinus venosus type interatrial septal defects or patent foramen ovale. Patients are asymptomatic until later in life (usually not hemodynamically significant and it is usually well tolerated by patients) and incidentally diagnosed with routine chest film, central catheter misplacement or post mortem examinations. In open hearts surgeries appropriate preoperative diagnosis is crucial for the accompanying congenital heart defects. We report a case of a surgical treatment of PAPVC in a 56 year old woman who had a history of mitral valve replacement surgery 1 year ago.

Öz

Parsiyel pulmoner venöz dönüş anomalisi (PAPVC) nadir görülen bir konjenital hastalıktır. Tüm konjenital kalp kusurları içinde insidans %0,5'dir. Genellikle bu tür doğumsal kalp kusurları sinüs venosus tipi atriyal septal defekt veya patent foramen ovale ile birlikte görülmektedir. Hastalar hayatlarının sonraki dönemlerine kadar asemptomatik olabilmekte (genellikle hemodinamik olarak anlamlı olmayan ve genellikle iyi tolere edilen) ve tesadüfen akciğer filmi, santral venöz kateterin yanlış yerleştirilmesi sonrasında veya otopsi sırasında tanı konulabilmektedir. Açık kalp ameliyatları öncesinde eşlik eden konjenital kalp defektlerinin tanısı çok önemlidir. Biz 1 yıl önce mitral kapak replasmanı ameliyatı öyküsü olan PAPVC tanısı alan 56 yaşındaki kadın hastanın cerrahi tedavisini sunduk.

Keywords
Partial anomalous pulmonary venous connection, surgical treatment, diagnosis

Anahtar Kelimeler
Parsiyel pulmoner venöz dönüş anomalisi, cerrahi tedavi, tanı

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Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital disorder with an incidence 0.5% of all congenital cardiac defects. One to three of the pulmonary veins (PV) drains oxygenated blood into the right atrium (RA) instead of the left atrium (LA) through the innominate vein (IV), superior vena cava (SVC), inferior vena cava (IVC), vena azygos or coronary sinus. Persistent left SVC is the most common cause with an incidence of 4.5% (1,2). Usually seen with congenital cardiac defects such as sinus venosus type atrial septal defect (ASD) or patent foramen ovale (sinus venosus type ASD are usually corrected by baffled patch closing). Isolated PAPVC without any other abnormality is very rare (3,4). Patients are asymptomatic until later in life (usually not hemodynamically significant and it is usually well tolerated by patients) and incidentally diagnosed with routine chest film, central catheter misplacement or post mortem examinations. Computed tomography (CT) scanning, magnetic resonance (MR), venography and catheterization of the pulmonary artery are used for diagnosing. We describe the surgical technique of reimplantation of the left SPV into the LA in a patients with PAPVC to the IV via a vertical vein without ASD with a history of (mitral valve repair) MVR surgery.

Case Report

Fifty-six year-old woman admitted our hospital with continuing complaints of dyspnea [New York Heart Association (NYHA) class 2], fatigue and palpitation after exercise for 10 months. She had a history of MVR surgery (2013), hypertension, diabetes and dyslipidemia. Physical examinations showed pretibial edema, venous distension and hepatojuguler reflux. The heart rate was 85 bpm and sinus rhythm in electrocardiogram. The blood pressure was 125/75 mmHg. First heart sound normal, second heart sound was harsh. Peripherical pulses were normal. Laboratory tests were within normal limits. The chest X-ray showed enlarged pulmonary arteries and increased pulmonary vascularity with a double density sign on the left paratracheal area. The transthoracic and transesophageal echocardiography [Transesophageal echocardiography (TEE)] were performed. Normal left ventricle, severely dilatation on the right heart chambers and severe pulmonary hypertension (PH) was found. There was no shunt on the atrial septum. A cardiac catheterization was performed, severe PH (mean pulmonary artery pressure 50 mmHg) and any significant findings in the coronary bed was detected. On computed tomography (CT) scan of the thorax showed a PAPVC with drainage of the left upper PV into the IV via a vertical vein (Figure 1). Based on these findings we decided to perform redo surgery. After cannulation of the right common femoral artery and vein patient was connected to the heart-lung machine then median sternotomy was performed, exploration was made with blunt and sharp dissection. After switching to bicaval venous cannulation, a clamp was placed on the ascending aorta. Subsequently a single dose of antegrade isotermic blood cardioplegia was administered to arrest the heart. Cardiopulmonary bypass (CPB) was performed with moderate hypothermia. The anomalous PV with vertical vein, SVC, left IV were dissected and encircled (Figure 2). The anomalous vein was ligated at the junction of the IV to facilitate the repositioning. Due to the left atrial appendix had been ligated for atrial fibrillation prior to the MVR surgery, partial clamp was placed in the left atrial wall. Longitudinal incision was made in the LA. Using a 10 mm Dacron greft anastomosis was performed between the left SPV and the LA (Figure 3). The patient was weaned off CPB with appropriate inotropic support.

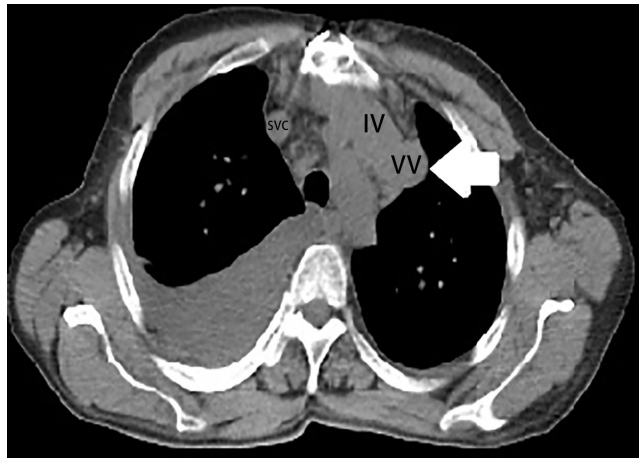

Figure 1. Partial anomalous pulmonary venous connection with drainage of the left superior pulmonary vein into the innominate vein via a vertical vein (arrow) and superior vena cava on computerized tomography scan

Discussion

PAPVC is a rare congenital anomaly and usually clinically silent that one or more PV drain into a systemic vein such as SVC, IVC, azygos vein, IV or directly to the RA rather than the LA since the early embryonic period. The incidence of PAPVC is 0.4-0.7% in total population. 10% are left sided and in 3% there is a connection between left lung and the left brachiocephalic vein. In 80-90% of patients are presented with an ASD (85% are sinus venosus type and 10-15% are secundum type). The severity of symptoms and complications depend on the shunt flow and extent of the left-right short circuit and the presence of other cardiac or pulmonary abnormalities (5). Diagnosis is made by echocardiography, computed tomography angiography, magnetic resonance imaging or cardiac catheterization and also pulmonary angiography provides a more detailed image. In this case, the anomalous venous return was not diagnosed by TEE on the prior MVR surgery. On account of continuing complaints of dyspnea (NYHA class 2), fatigue and palpitation after exercise for 10 months we decided to repeat both transthoracic and TEE. Unfortunately no anomalous was detected except severe PH, then chest CT angiography and cardiac catheterization was performed.

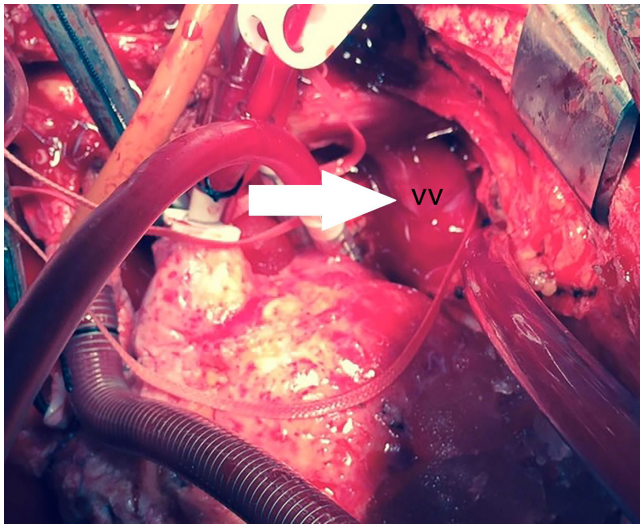

Figure 2. Externel view of the vertical vein (arrow)

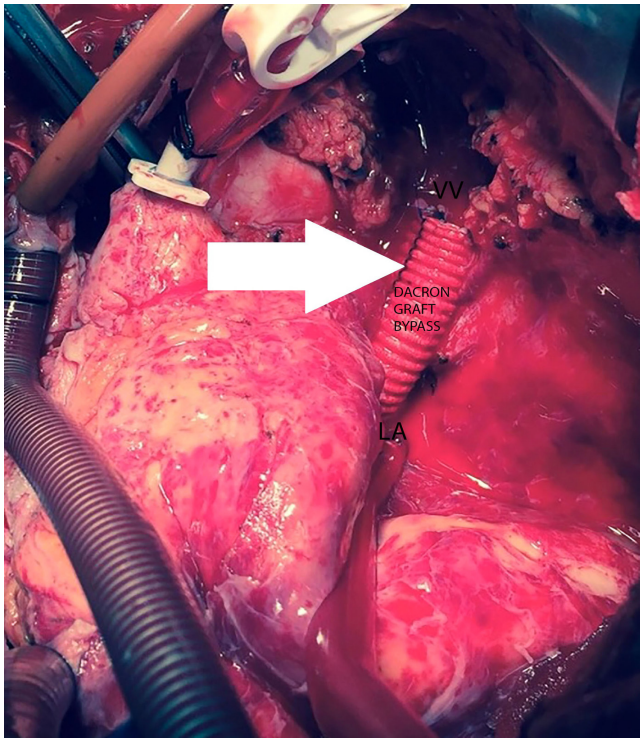

Figure 3. Intraoperative image. Interposition of dacron graft between innominat vein and the left atrium (arrow)

In 42.8% of patients are presented with PH. Besides, some authors determined that using echocardiography may fail in diagnosing due to outflow of all PV to the LA can not be differentiated (6). PAPVC can cause hemodynamically significant right to left shunt (Qp: Qs N 1.5:1), right ventricular failure or pulmonary vascular obstructive disease therefore, this identity must be treated surgically. Surgical treatment results of PAPVC surgery are generally good (7). Timing of the surgery is essential.

In conclusion, in open hearts surgeries appropriate preoperative diagnosis is crucial for the accompanying congenital heart defects. During open heart surgery PV's are not investigated routinely by surgeons whether there is a congenital defect or not. On account of this cardiologists should be more skeptical for overlooked congenital heart defects. On the other hand timing of cardiac surgery is essential to prevent right heart failure and improve patient's quality of life.

Ethics

Informed Consent: We received indormed consent from the patient.

Peer-review: Internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: S.B.D., M.K.D., Concept: S.B.D., Design: S.B.D., Data Collection or Processing: S.B.D., Analysis or Interpretation: S.B.D., Literature Search: S.B.D., Writing: S.B.D., M.K.D.

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