

Partial Anomalous Pulmonary Venous Return Presenting with Adult Onset Thalamic Infarction

Talamik İnfarktüs ile Başvuran Erişkin Hastada Parsiyel Pulmoner Venöz Dönüş Anomalisi

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ABSTRACT Partial pulmonary venous return anomaly (PPVRA) is defined as one or more of the pulmonary veins drain into the right atrium or a systemic vein such as superior vena cava (SVC), inferior vena cava (IVC), azygos, hepatic or portal vein. Diagnosing of PPVRA is often clinical challenging. Onset clinical manifestation and progression of the disease vary significantly depending on the amount of the intra-cardiac shunt. Signs or symptoms may include dyspnea, atrial arrhythmia, right heart failure and pulmonary hypertension, transient ischemic attack (TIA), or stroke. In this article, we aimed to present a 39-year-old female patient who underwent Warden Procedure with the diagnosis of PPVDA.

Keywords: Thalamic infarction; sinus venosus atrial septal defect; partial anomalous pulmonary venous return; intra-cardiac shunt

ÖZET Parsiyel pulmoner venöz dönüş anomalisi (PPVDA), bir ya da daha fazla pulmoner venin sağ atriyuma ya da superior vena cava (SVC), inferior vena cava (IVC), azygos, hepatic ya da portal ven gibi sistemik bir damar içine drene olması ile tanımlanır. PPVDA'nın teşhisi genellikle klinik açıdan zordur. Başlangıçtaki klinik görünüm ve hastalığın ilerlemesi, kardiyak şant miktarına bağlı olarak önemli ölçüde değişmektedir. İşaretler veya semptomlar dispne, atriyal aritmi, sağ kalp yetmezliği ve pulmoner hipertansiyon, geçici iskemik atak (TIA) veya inmeyi içerebilir. Bu yazıda PPVDA tanısıyla Warden Prosedürü uyguladığımız 39 yaşında kadın hastayı sunmayı amaçladık.

Anahtar Kelimeler: Talamik infarktüs; sinüs venozus atriyal septal defekt; parsiyel anormal pulmoner venöz dönüş; intrakardiyak şant

Partial pulmonary venous return anomaly (PPVRA) is described as drainage of one or more pulmonary veins into the right atrium or to a systemic vein. This abnormal venous return may be towards the superior and inferior vena cava, the azygos vein, the portal or the hepatic veins.¹ This cardiac anomaly is a very rare. The prevalence of this, 0.1-0.2% in adult population reported.² Mostly, abnormal right pulmonary veins (especially right upper pulmonary veins) are opened to the right atrium (RA) or superior vena cava (SVC). The left-sided PPVRA may be drained into the innominate vein, hemiazygos vein and coronary sinus.¹ PPVRA is commonly associated with atrial septal defect (ASD) and other congenital heart anomalies.¹ PPVRA is seen in approximately 10-15% of ostium secundum ASD cases and 85% of patients with sinus venosus ASD.³

In this article, we aimed to present a 39-year-old female patient operated for PPVRA.

CASE REPORT

A 39-year-old female patient was admitted to a neurologist with a complaint of diplopia six months ago. Brain magnetic resonance (MR) imaging showed infarct area consistent with late acute-subacute phase and diffuse contrast enhancement in the anterior half of the left thalamus (Figure 1A, 1B). Then acetylsalicylic acid (ASA) 100 mg and Rivaroxaban 20 mg once a day were started. The patient's diplopia complaint has disappeared. Additionally, subclinical deep vein thrombosis (DVT) was detected in the lower extremity doppler. At the same time, the etiology that caused this condition was also investigated. Transthoracic echocardiography (TTE) showed ASD and mild pulmonary hypertension sign was seen [mean pulmonary arterial pressure (PAP) 35 mmHg]. Qp: Qs ratio of 1:5, mild tricuspid regurgitation. In Transesophageal echocardiography (TEE), sinus venosus type ASD was observed in the interatrial septum adjacent to the SVC. Thorax MR imaging, approximately 7 mm

in diameter defective appearance (ASD) in interatrial septum. Right pulmonary veins were found to connect to the ASD region. A total of five pulmonary veins were seen in the right upper lobe and middle lobe of the lung. Four of the pulmonary veins of the right upper and middle lung lobes were seen opening into the lateral SVC just above the right atrium and SVC junction (Figure 2A, 2B, 2C, 2D). In addition, there is a pulmonary vein trunk from the right lung to the left atrium. Upon these findings, the patient was referred to our clinic for operation. Physical examinations showed that his pulse was 96/min, blood pressure was 100/70 mmHg. Other physical examination signs were unremarkable. Patient's electrocardiography (ECG) was sinus rhythm. Pre-operative control computed tomography (CT) imaging showed that the infarct area was markedly reduced and the contrast difference was reduced (Figure 1C, 1D).

Written informed consent was obtained from patient. Under general anesthesia, sternotomy was done. Then cardiopulmonary bypass was institu-

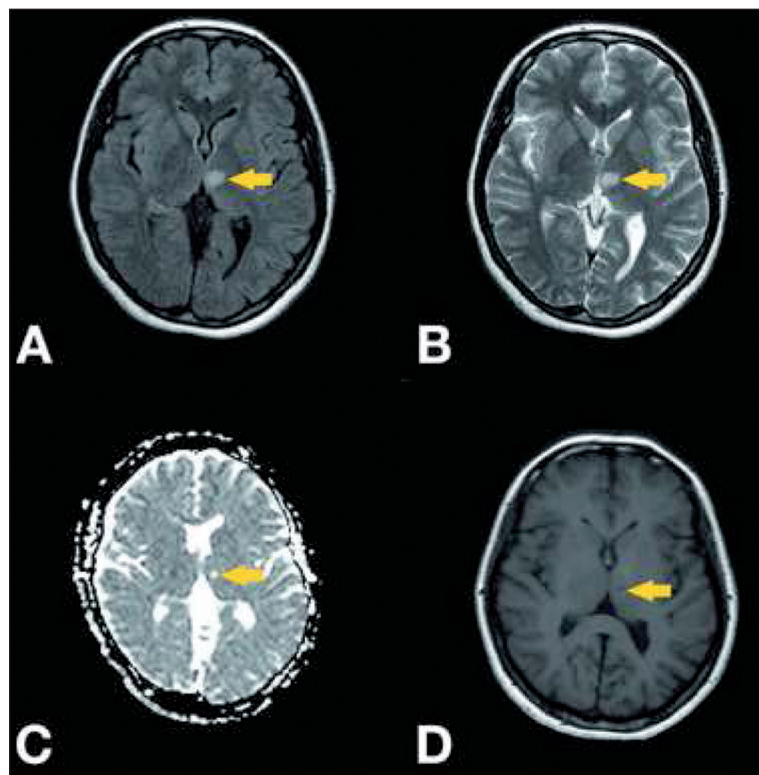


FIGURE 1: A, B) Six months prior computed tomography images of the anterior half of the left thalamus infarct area (yellow arrow), C, D) Pre-operative control computed tomography imaging (yellow arrow).

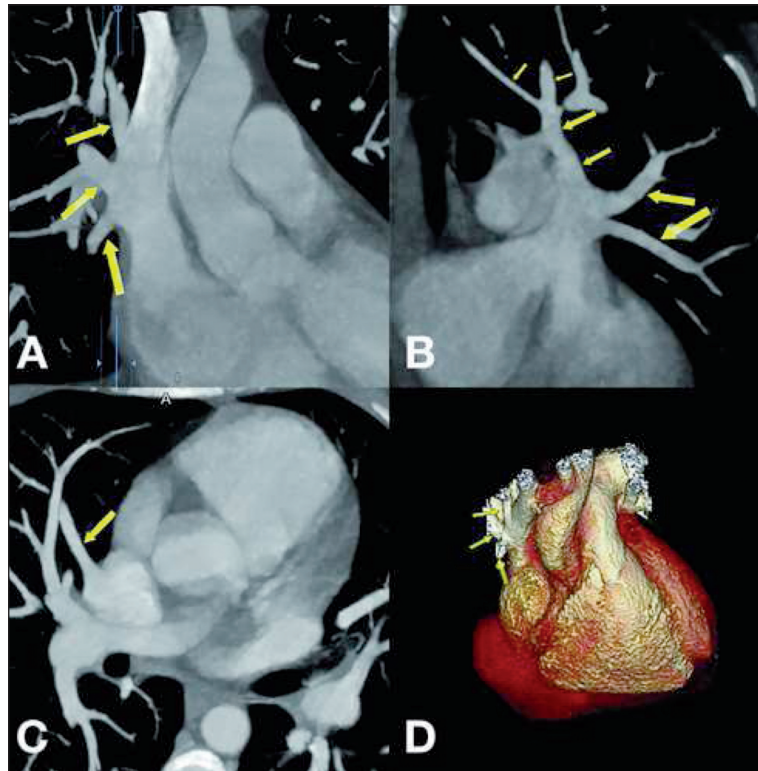


FIGURE 2: Computed tomography angiography images of different sections of abnormal pulmonary venules (yellow arrows).

ted ascending aorta and bicaval cannulation. With rigorous dissection, the SVC and the five anomalous pulmonary veins were identified. The aorta was cross-clamped and cardioplegia was given from the aortic root. Right atriotomy was performed and cardiac evaluation revealed sinus venosus type ASD. Five right pulmonary venous branches were opened to the SVC. At the same time, the orifices of these pulmonary veins located in the upper part of the superior vena cava were also seen. A pericardial patch was used to close the cardiac stump of the SVC in order to avoid stenosis or obstruction of the pulmonary venous return. The pericardial patch was sutured to the edge of the ASD over the SVC orifice. Then we used Warden Procedure that baffling this orifice to re-direct the blood flow from these anomalous pulmonary veins into the left atrium (Figure 3A, 3B, 3C, 3D). The postoperative course was uneventful. Postoperative day of sixth, the patient was discharged. In the follow-up of second months, the good result of surgical repair

was confirmed by with echocardiographic assessment.

DISCUSSION

Diagnosing of PPVRA is often clinical challenging. Clinical findings and course of the disease vary depending on the degree of left-right shunt. The amount of shunt from left to right, symptoms and presence of pulmonary arterial hypertension (PAH) affect medical or surgical treatment decisions. Signs or symptoms may include dyspnea, exercise intolerance, lower extremity edema, palpitation, atrial fibrillation/flutter, right ventricular dilatation or heart failure and pulmonary hypertension, transient ischemic attack (TIA), or stroke.^{1,4} Cerebral embolization and associated TIA and stroke development probably seem to be related to ASD being very close to the SCV ostium. Most of these patients are misdiagnosed or incidentally diagnosed as primary pulmonary hypertension. In addition, these patients may remain asymptomatic for years. In our case, there was a

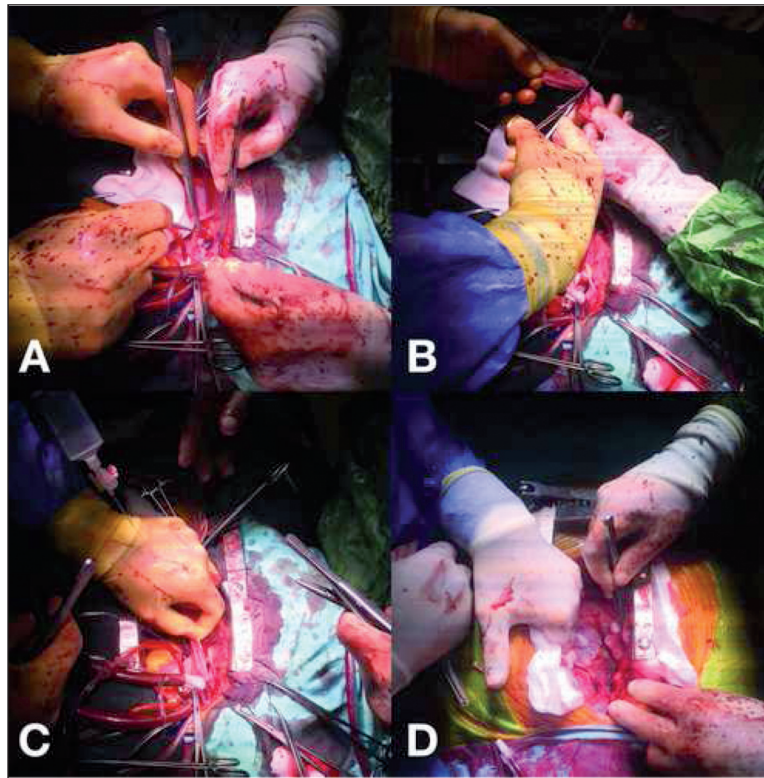


FIGURE 3: Images during surgery. **A)** sinus venosus type ASD, **B)** preparation of pericardial patch, **C)** repair of ASD with pericardial patch and tunnel formation, **D)** expansion of SVC incision using pericardial patch and repair.

history of thalamic infarction six months ago. The cause of the diagnosis was already the research done after thalamic infarction.

While right anomalous pulmonary veins are usually connect vena cava and right atrium, left anomalous pulmonary veins connect coronary sinus and left innominate vein.⁵ In our case, all anomalous pulmonary veins were associated with SVC.

This abnormal systemic venous return and shunt from left to right cause a volume overload in the lung during a long period. Because oxygen rich blood returns to the right heart again without joining the systemic circulation. With time, constantly increasing pulmonary blood flow can lead to progressive remodeling of the pulmonary circulation and escalate pulmonary arterial resistance. Furthermore, PAH and right ventricular volume overload resulting in right ventricle (RV) failure occur.⁶ This may be confused with heart failure with preserved ejection fraction (HFpEF). Patients with PPVRA can be mistakenly identified as HFpEF.⁴ Our patient had not yet developed PAH or right

heart failure. We thought this was related to the patient's young age.

Definitive treatment of PPVRA with SVD is surgical repair. Various surgical techniques have been identified. The most common of these is the Warden Procedure.⁷ This procedure includes the complete closure of the sinus venosus type ASD and the redirection of pulmonary veins to left atrium (LA) without making obstruction in the pulmonary veins and the SVC (Warden Procedure).⁷ It should also be ensured that the sinus node is not injured.⁸ The advantages of the Warden Procedure include the prevention of sinus node dysfunction and pulmonary/systemic venous obstruction.⁷

Newly developed rhythm disorders have been reported in 20-40% after surgical repair. Supraventricular tachycardias are frequently reported in patients who need to be enlarged, especially in the vena cava. Transient dysrhythmias may occur in the early period due to the intensity of interventions performed around the sinus node.⁵ In our patient we also expanded with an autologous pericardial patch

to avoid narrowing the SVC-right atrium junction. She also did not develop any rhythm disturbances in postoperative period. Surgical repair of pulmonary veins should be done in patients with high level SVC drain rather than direct RA. If abnormal pulmonary veins bind to higher levels of SVC, the cephalic segment of the SVC may move further away from the RA and cause stretching or stenosis of cavo-atrial anastomosis. Therefore, extensive dissection of the brachycephalic vein and SVC is very important to prevent possible tension in the cavo-atrial anastomosis.⁷ Otolog pericardial patch is tunneled and the pulmonary venous circulation is directed to the LA. In doing so, cavo-atrial anastomosis should be performed without strain/injury to prevent SVC-RA stenosis, and this requires very rigorous dissection of the brachiocephalic vein and SVC.^{5,7,8} In a patient with single right SVC, clamping of this vessel may cause neurological damage as the brain will expose it to high venous pressures.⁷ If the patient has persistent left SVC, it provides adequate cerebral venous drainage and prevents adverse neurological complications.⁷ In our case, there was no persistent left SVC, but no neurological damage occurred in the preoperative or postoperative period.

In conclusion it should be known that in a patient with diplopia, there may be paradoxical em-

bolization of PPVRA and accompanying sinus venosus type ASD. The Warden Procedure is an option that is safe in these patients and that is gratifying results. We think that our case is the first case report of cerebral (thalamic) infarction after subclinical DVT in a patient with PPVRA and sinus venosus type ASD.

Source of Finance

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: İhsan Alur; **Design:** İhsan Alur; **Data Collection and/or Processing:** İhsan Alur, Ali Fedakar; **Analysis and/or Interpretation:** Ali Fedakar, İhsan Alur; **Literature Review:** Mehmet Şirin Yıldırım; **Writing the Article:** İhsan Alur, Mehmet Şirin Yıldırım; **Critical Review:** Ali Fedakar.

REFERENCES

1. Wang H, Guan H, Wang DW. Partial anomalous pulmonary venous connection to superior vena cava that overrides across the intact atrial septum and has bi-atrial connection in a 75-year-old female presenting with pulmonary hypertension. *BMC Cardiovasc Disord.* 2014;14:149. [Crossref] [PubMed] [PMC]
2. Ho ML, Bhalla S, Bierhals A, Gutierrez F. MDCT of partial anomalous pulmonary venous return (PAPVR) in adults. *J Thorac Imaging.* 2009;24(2):89-95. [Crossref] [PubMed]
3. Sahay S, Krasuski RA, Tonelli AR. Partial anomalous pulmonary venous connection and pulmonary arterial hypertension. *Respirology.* 2012;17(6):957-63. [Crossref] [PubMed] [PMC]
4. Sharma RK, Houston BA, Lima JA, Cameron DE, Tedford RJ. Never too old for congenital heart disease: sinus venosus atrial septal defect with anomalous pulmonary venous return in an octogenarian. *Pulm Circ.* 2015;5(3):587-9. [Crossref] [PubMed] [PMC]
5. Ömeroğlu SN, Mansuroğlu D, Kırılı K, Kayalar N, Mutlu B, Ardal H, et al. [Surgical repair of partial anomalous pulmonary venous connection and long-term results]. *Türk Göğüs Kalp Damar Cer Derg.* 2005;13:103-6.
6. Sears EH, Aliotta JM, Klinger JR. Partial anomalous pulmonary venous return presenting with adult-onset pulmonary hypertension. *Pulm Circ.* 2012;2(2):250-5. [Crossref] [PubMed] [PMC]
7. Ramanan S, Sasikumar N, Manohar SR, Cherian KM. The warden procedure through a posterolateral thoracotomy approach. *Tex Heart Inst J.* 2014;41(5):499-501. [Crossref] [PubMed] [PMC]
8. Perri G, Graziani F, Bruno P, Grandinetti M, Lanzillo C, Marziali M, et al. Modified Warden procedure in adult with partial anomalous pulmonary venous connection after previous atrial septal defect repair. *Cor et Vasa.* 2016;58:e501-4. [Crossref]