

Cor Triatriatum Sinister Associated with Partial Anomalous Pulmonary Venous Return Mimicking Total Anomalous Pulmonary Venous Connection

Total Anormal Pulmoner Venöz Dönüş Anomalisini Taklit Eden Parsiyel Anormal Pulmoner Venöz Dönüş ve Cor Triatriatum Sinister

© Gökmen Akkaya¹, © Meltem Çakmak², © Muhammet Akyüz¹, © Onur Işık¹

¹University of Health Sciences Turkey, İzmir Tepecik Education and Research Hospital, Clinic of Pediatric Cardiovascular Surgery, İzmir, Turkey

²University of Health Sciences Turkey, İzmir Tepecik Education and Research Hospital, Clinic of Anesthesiology and Reanimation, İzmir, Turkey

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Abstract

One year old patient was referred to our clinic with the diagnosis of total anomalous venous connection. Despite tachycardia and pulmonary plethora, the patient was acyanotic and the transthoracic echocardiography detected pulmonary venous baffle or vertical vein. Considering these doubtful findings, computed tomography angiography was performed and revealed that right pulmonary veins was draining towards an enlarged vena cava superior while right pulmonary veins into the left atrium. Furthermore, a fibrotic tissue that encurtains the mitral orifice was observed. Consequently, the patient underwent surgery with the diagnoses of cor triatriatum sinistrum and partial anomalous pulmonary connection. The exact diagnosis was confirmed after a profound surgical inspection; afterwards, then the whole process was accomplished smoothly and the patient discharged in good condition.

Keywords: Cor triatriatum sinistrum, partial anomalous pulmonary venous return, total anomalous pulmonary venous connection

Öz

Bir yaşındaki hasta total anormal venöz dönüş ön tanısıyla kliniğimize yönlendirildi. Taşikardi ve pulmoner konjesyona rağmen hasta asiyantikti ve transtorasik ekokardiyografide pulmoner venöz kese veya vertikal ven saptanmadı. Bu şüpheli bulgular göz önünde bulundurularak yapılan bilgisayarlı tomografi anjiyografide sağ pulmoner venlerin genişlemiş vena kava superiora, sol pulmoner venlerin ise sol atriyuma boşaldığı görüldü. Ayrıca mitral kapağı çevreleyen fibrotik zar da gözlemlendi. Bunun üzerine hasta kor triatriatum sinister ve parsiyel anormal pulmoner dönüş tanılarıyla ameliyata alındı. Kesin tanı, kapsamlı bir cerrahi incelemenin ardından doğrulandı; sonrasında tüm süreç sorunsuz bir şekilde tamamlandı ve hasta sağlıklı bir şekilde taburcu edildi.

Anahtar Kelimeler: Cor triatriatum sinistrum, parsiyel anormal pulmoner venöz dönüş, total anormal pulmoner venöz dönüş



Address for Correspondence/Yazışma Adresi: Gökmen Akkaya MD, University of Health Sciences Turkey, İzmir Tepecik Education and Research Hospital, Clinic of Pediatric Cardiovascular Surgery, İzmir, Turkey

Phone: +90 506 861 00 84 **E-mail:** akkayagokmen@gmail.com

ORCID ID: orcid.org/0000-0002-0509-1971

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Introduction

Cor triatriatum sinister (CTS) is identified as an anatomical division of the left atrium by a fibromuscular membrane that separates the pulmonary venous drainage from the mitral valve orifice. Various theories have been assessed to determine the reason for CTS such as fault in mal-septation, mal-incorporation, or entrapment during cardiac development so far. Nevertheless, the underlying embryological mechanism is yet unclear and remains controversial. A similar mechanism also cause pulmonary venous connection anomalies if pulmonary venous drainage is interrupted at earlier gestational age in reference to CTS⁽¹⁾. Consequently, a combination of CTS and partial anomalous pulmonary venous return (PAPVR) or total anomalous pulmonary venous connection (TAPVC) are not exceptional. On the other hand, manifestations and severity of symptoms among patients considerably vary, thus often lead to misdiagnosis. Such patients, therefore, require further examination to conclude the optimal surgery type and operative timing.

Herein, we present a patient who was referred to our clinic as late-diagnosed TAPVC and subsequently got definitive diagnosis as CTS and PAPVR peroperatively.

Case Report

One year old boy weighing 8.8 kg's was referred to our clinic with the diagnosis of TAPVC. Dyspnea and tachycardia

were determined in the physical examination, however, there was no cyanosis (SATO₂ 98%). The heartbeat was 132 bpm and the rhythm was sinus. The chest graphy was not significant except for pulmonary plethora. The transthoracic echocardiography (TTE) performed in our center revealed doubtful findings. No pulmonary venous baffle and atrial septal defect (ASD) were detected. Enlarged right atrium, vena cava superior (VCS), and an apparent azygous vein were observed; and increased pulmonary venous blood pressure was measured. Considering the suspicious diagnosis and questionable TTE examinations, computed tomography (CT) angiography was performed to define the precise anatomy.

CT angiography demonstrated that the right pulmonary veins drains towards the enlarged VCS whilst the left pulmonary veins towards the left atrium. Moreover, a cavernous obstructive fibrous tissue was observed in the left atrium that hindering the pulmonary venous flow towards the mitral orifice (Figure 1A, B). However, there was no apparent vertical vein in CT as well as in the TTE images.

Consequently, the certain diagnosis was established as CTS, PAPVR with intact interatrial septum. Then, considering the current hemodynamic situation, the patient underwent urgent surgery. The mediastinal access was achieved thru midline sternotomy. Following bi-caval cannulation, the cardiopulmonary bypass was initiated. The diameter of VCS was significantly enlarged and a right upper pulmonary vein adjacent to VCS was observed, however, no pulmonary

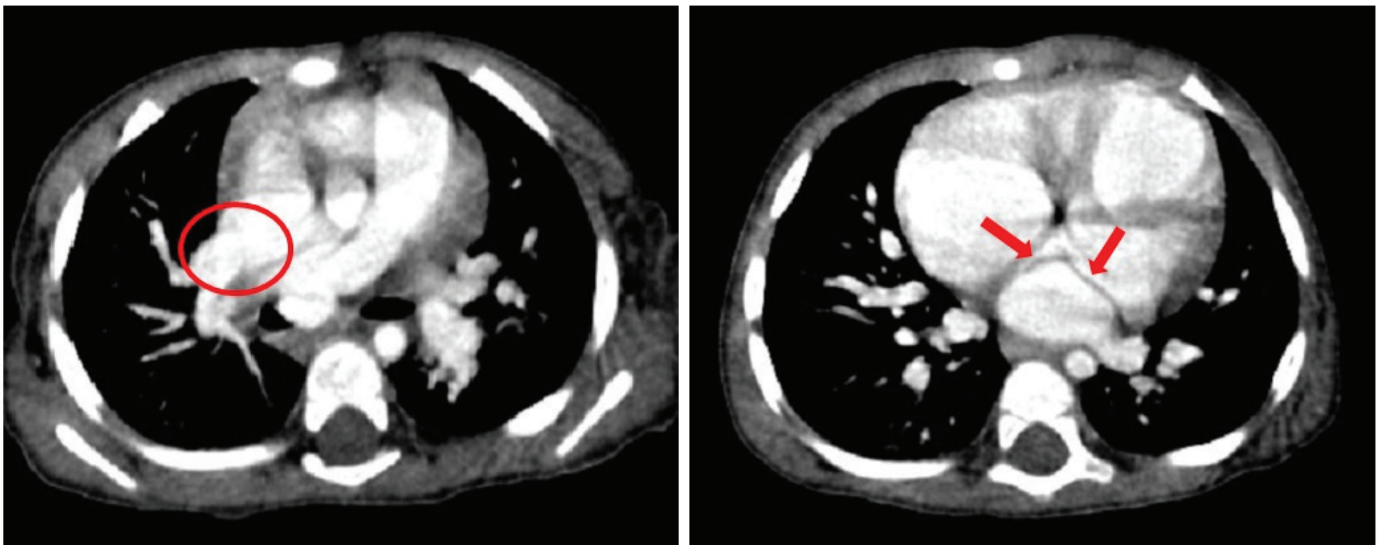


Figure 1. A, B. In the red circle right upper pulmonary vein and the superior vena cava connection is being shown. Red arrows marking the C shaped image of the fibrous membrane separating the pulmonary veins from the rest of the left atrium

venous baffle detected when the apex was removed. After cardioplegia administration and right atriotomy incision, the cardiac anatomy was precisely identified. CTS and right PAPVD and small secundum ASD were diagnosed by profound inspection (Figure 2). After septectomy, an occlusive fibrous membrane that encurtains the mitral valve was observed then resected. Afterwards the septectomy was closed via ePTFE graft with continuous stitches so as to reroute the right upper pulmonary venous flow into the left atrium. The whole surgical process was accomplished smoothly.

Afterward, the patient was transferred to the intensive care unit with negligible inotrope support and readily extubated at the sixth postoperative hour. However, pulmonary arterial hypertension led to prolonged non-invasive mechanical support and sildenafil 1 mg/kg/day was initiated. After the recovery period, the patient was transferred to ward on the POD 7 and then discharged on POD 13 in good condition with the daily medication of acetylsalicylic acid (5 mg/kg) and sildenafil (1 mg/kg).

The control TTE examination on the first month of follow-up was uneventful. The sildenafil treatment was gradually rarefied, then terminated in the control examinations.

Discussion

The development of the CTS begins with the appearance of the primitive pulmonary vein as an outpouching from the left atrium, subsequently connects with the primitive

pulmonary venous plexus. Then, the common pulmonary baffle is incorporated into the posterior left atrium, and the connection between the pulmonary venous plexus and other venous systems gradually peters out. In the lack of complete partitioning CTS occurs.

Depending on the gestational age at the interruption in the usual embryogenesis, various connections between the atrial chamber and the cardinal venous system may develop. For instance, early atresia in the common pulmonary vein results in TAPVC by canalizing the drainage through either the umbilicovitelline or cardinal venous system⁽²⁾. Although both CTS and PAPVR or TAPVC share the same hypothetical mechanism simultaneous cases are virtually rare.

The majority of CTS cases remain asymptomatic, however coexisting cardiac defects lead to various clinical manifestations and may be cloud certain diagnoses.

In occasional cases, CTS and TAPVC can be diagnosed by saline contrast echocardiography⁽³⁾. However, CT angiography mostly aids to determine certain diagnoses. However, challenging cases may require cardiac catheterization for confirmation⁽³⁾.

Although no vertical vein and ASD was observed and the patient was acyanotic; in the misguidance of TTE, our case was suspected of TAPVC. Hence, absolute exclusion of the CTS and associated intracardiac TAPVC was not possible. In a similar case, Kwak et al.⁽⁴⁾ highlight the importance and guidance of preoperative evaluation of the CT images.

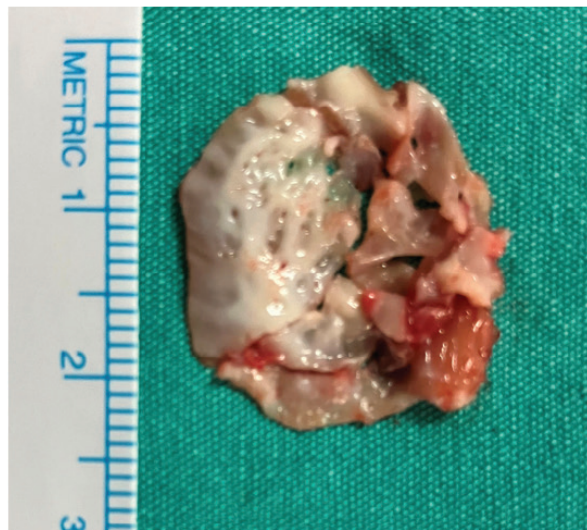
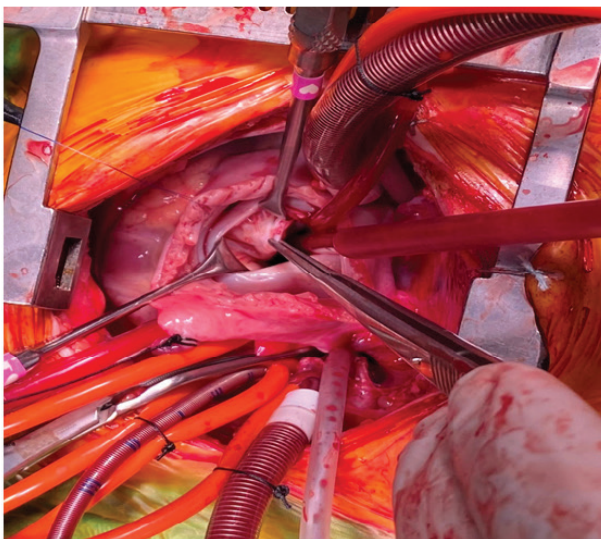


Figure 2. The surgical view through left atrial incision. The fibrous obstructive tissue in front of the mitral valve is visualized

Beyond that, CTS and PAPVD may develop rapid cardiogenic shock, especially in the presence of restrictive ASD. Such patients warrant immediate treatment. For instance, Schiller et al.⁽⁵⁾ prefer staged surgery in cardiogenic shock in infancy. Firstly, they maintained the ECMO support to establish hemodynamic stabilization. During this delay, catheterization was performed to confirm the diagnosis. Similarly, Moscoso et al.⁽⁶⁾ highlight that even the detailed anatomy and the exact diagnosis are highly required prior to the operation; nevertheless, in case of urgency, the confirmation of the diagnosis awaits the perioperative findings. For these reasons, the authors recommend a profound inspection and a careful anatomical analysis during surgery.

In conclusion, we state that CTS and PAPVD cases often manifest with severe symptoms and may easily be misdiagnosed with TAPVC. However, in experienced hands, CT angiography provides useful findings as becomes as sufficient as TTE images. Nevertheless, if urgent surgery is required in the lack of adequate sufficient time to determine the exact diagnosis, precautions must be taken and current anatomy should be inspected profoundly during surgery.

Ethics

Informed Consent: A written informed consent was obtained from both parents of the patient.

Authorship Contributions

Surgical and Medical Practices: G.A., M.Ç., M.A., O.I., Concept: G.A., Design: G.A., O.I., Data Collection or Processing: G.A., Analysis or Interpretation: M.A., O.I., Literature Search: M.Ç., M.A., Writing: G.A., O.I.

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