

Right Sided Complex Partial Anomalous Pulmonary Venous Return Associated with Sinus Venosus Atrial Septal Defect

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Abstract

Partial anomalous pulmonary venous return is a rare congenital anomaly affects one or more right- or left-sided pulmonary veins. Sinus venosus atrial septal defect association of this anomaly may be also seen. In this paper, we have reported an unique partial anomalous pulmonary venous return case associated with sinus venosus atrial septal defect who shows segmental drainage difference in the same lobe.

Keywords: Computed tomography angiography; Magnetic resonance imaging; Partial anomalous pulmonary venous return; Sinus venosus atrial septal defect

Introduction

Partial Anomalous Pulmonary Venous Return (PAPVR) is a rare congenital anomaly described in 0.4-0.7% of autopsies [1]. Anomalous venous return can contain one or more right- or left-sided pulmonary veins draining into the coronary sinus, right atrium or systemic veins [2].

Although PAPVR and its Sinus Venosus Atrial Septal Defect (SVASD) association is a rare condition, there are cases reported in literature [3,4]. In this report, we have described a very rare PAPVR case different from the similar reports in the literature.

Case Report

A 30-year-old male visited the Cardiology Department of our hospital with dyspnea and palpitation complaints. 2/6 systolic murmur at the pulmonary focus and fixed split S2 were detected on auscultation. Right branch bundle block was observed on electrocardiography. On transthoracic echocardiography, the right heart cavities were dilated. SVASD was detected on transesophageal echocardiography. In the same examination, anomalous pulmonary venous return to Superior Vena Cava (SVC) was also suspected, and the patient was referred to Radiology Department for the pulmonary venous Computed Tomography Angiography (CTA) examination.

Pulmonary venous CTA demonstrated that combining apical and posterior segment pulmonary veins of the right upper lobe drain into SVC via one ostium. Anterior segment of the right upper lobe and the middle lobe pulmonary vein were draining into SVC-right atrium junction separately at the level of SVASD. The atrial septal defect size at the location of cavoatrial junction was 12 mm. Right lower lobe and left lung pulmonary veins were draining into the left atrium as usual. Inferior vena cava, SVC and coronary sinus were normal. Right atrium and ventricle were mildly large (Figure 1).

The surgery was planned by Cardiovascular Surgery department. But the patient did not accept either surgical or medical treatment and no further evaluation could be performed.

Discussion

PAPVR is a congenital anomaly which involves the drainage of one to three pulmonary veins into the right-sided circulation, creating a partial left-to-right shunt [3]. It represents persistence of embryonic anastomosis between the pulmonary veins and systemic venous

plexus resulting in one or more anomalous pulmonary veins [4]. In pediatric studies, PAPVR has been reported to be twice as common in males as females, and more frequently arising from the right (90%) than left upper lobe (10%). Additionally, the right upper lobe PAPVR is associated with SVASD in 80% to 90% and ostium primum atrial septal defect in 10% to 15% of cases. It is presumed that "unroofing" of the right superior pulmonary vein into the SVC during embryologic development produces a combination of anomalous return and ASD [5].

Depending on the severity of the left-to-right shunting, dyspnea, and palpitation associated with atrial arrhythmias, and the symptoms of right-sided heart failure and pulmonary hypertension may be seen in PAPVR patients [6]. The diagnosis cannot be possible until adulthood in many of the patients. The symptomatic patients at an early age tend to have more serious disease and more congenital malformations requiring surgery. In contrast, PAPVR recognised in adulthood emerges subclinical or mild disease with a few associated anomalies [3]. Our case has dyspnea, palpitation and arrhythmia, and his right heart cavities were mildly dilated.

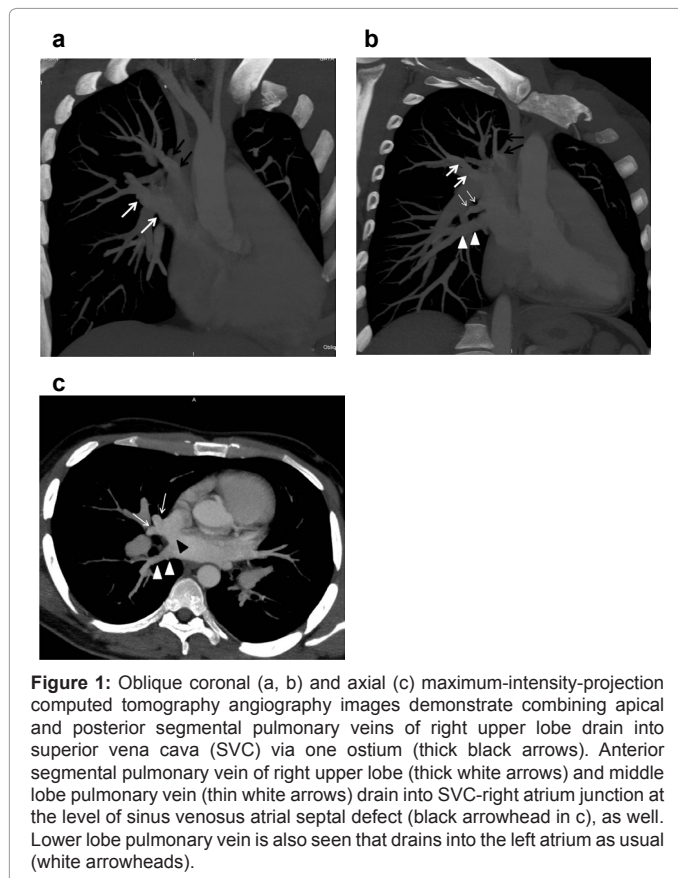
The important role of the transesophageal echocardiography in the diagnosis of PAPVR and ASD is well founded [4], and its diagnostic power is superior than transthoracic echocardiography. Because of its technical advantages such as the shorter distance between the probe and defect, longitudinal and short-axis images, and three-dimensional reconstruction capability that are essential for determining of the pulmonary-systemic venous connection, identifying the exact localization of the defect, transesophageal echocardiography currently is the most sophisticated diagnostic modality for the morphologic evaluation of ASDs. Additionally, intravascular ultrasonography has also similar advantages for diagnosis of ASDs. Even so, cross sectional imaging is the modality of choice for the diagnosis of the PAPVR with or without ASD. The presence, course, and the number of the anomalous pulmonary veins and probable associated cardiovascular defects can be

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detected. Contrast-enhanced CT and CTA are optimum for this aim, allowing rapid data acquisition with high resolution and wide anatomic coverage [7]. Enhanced magnetic resonance angiography and cardiac magnetic resonance imaging also provide high spatial resolution for the evaluation of the cardiac anomalies and great vessels such as pulmonary veins and arteries. With magnetic resonance angiography, three-dimensional reconstructions which assure detailed anatomic information can be composed [8].

Surgical treatment may be considered in patients who have isolated PAPVR and signs of right ventricle overload. In the hands of the competent surgeons, this surgery can be performed with low morbidity and mortality, with improvement in the right ventricle size and pulmonary pressures in the majority [2]. The aim of surgical repair is to close the defect in the interatrial septum and to normalize the systemic and pulmonary venous drainage. There are various surgical techniques for treating PAPVR with or without ASD such as patch techniques, and Warden's technique [2,9,10]. Because of our patient did not accept the further evaluation or any treatment, we could not measure the pulmonary pressures and offer the postoperative information.

Normally, all of the pulmonary veins drain into the left atrium. In our case, combining apical and posterior segment pulmonary veins of the right upper lobe were draining into systemic venous circulation via SVC. Anterior segment of the right upper lobe and the middle lobe pulmonary veins were draining into SVC-right atrium junction separately at the level of SVASD. In medical literature there are case series [2,3] consist of the the right upper lobe PAPVR drain into SVC, but segmental drainage difference have not been reported before in medical literature. Majdalany et al. [2] reported their PAPVR series in 2010 that shows twenty year experience. In that report, there were

totally 43 PAPVR case of which 16 case had right upper lobe and/or right middle lobe pulmonary vein was draining into SVC. Also Ho et al. [3] reported their eight-year experience about this subject and there was also any case shows segmental drainage difference in the same lobe in their reports.

We have diagnosed a right sided PAPVR showing segmental venous drainage difference in the right upper lobe associated with SVASD. Normally, the expected pathology in PAPVR is the drainage difference between the lobar pulmonary veins in one lung. In the right upper lobe of our case, two of the three segmental pulmonary veins were draining into systemic venous system via SVC, the other was draining into right atrium. This segmental difference in the same lobe composes the uniqueness of the case and it is an extremely rare situation. The drainage of the other pulmonary veins were into the left atrium, as usual.

CTA provided full description of this complex anomaly, documenting the detailed anatomic information about the segmental and lobar pulmonary venous drainage, the location of the SVASD and the interatrial defect size. These kind of informations may guide the surgeon to choose the appropriate surgical technique, preoperatively and reduce the probable complications.

References

1. HEALEY JE Jr (1952) An anatomic survey of anomalous pulmonary veins: their clinical significance. *J Thorac Surg* 23: 433-444.
2. Majdalany DS, Phillips SD, Dearani JA, Connolly HM, Warnes CA (2010) Isolated partial anomalous pulmonary venous connections in adults: twenty-year experience. *Congenit Heart Dis* 5: 537-545.
3. Ho ML, Bhalla S, Bierhals A, Gutierrez F (2009) MDCT of partial anomalous pulmonary venous return (PAPVR) in adults. *J Thorac Imaging* 24: 89-95.
4. Ammash NM, Seward JB, Warnes CA, Connolly HM, O'Leary PW, et al. (1997) Partial anomalous pulmonary venous connection: diagnosis by transesophageal echocardiography. *J Am Coll Cardiol* 29: 1351-1358.
5. Fraser RS, Muller NL, Colman N. Developmental anomalies affecting the pulmonary vessels. In: Fraser RS, Muller NL, eds. *Diagnosis of Diseases of the Chest*. (4th edn). Philadelphia: Saunders, 1999.
6. Alsoufi B, Cai S, Van Arsdell GS, Williams WG, Caldarone CA, et al. (2007) Outcomes after surgical treatment of children with partial anomalous pulmonary venous connection. *Ann Thorac Surg* 84: 2020-2026.
7. Choe YH, Kang IS, Park SW, Lee HJ (2001) MR imaging of congenital heart diseases in adolescents and adults. *Korean J Radiol* 2: 121-131.
8. de Roos A, Roest AA (2000) Evaluation of congenital heart disease by magnetic resonance imaging. *Eur Radiol* 10: 2-6.
9. KIRKLIN JW, ELLIS FH Jr, WOOD EH (1956) Treatment of anomalous pulmonary venous connections in association with interatrial communications. *Surgery* 39: 389-398.
10. Warden HE, Gustafson RA, Tamay TJ, Neal WA (1984) An alternative method for repair of partial anomalous pulmonary venous connection to the superior vena cava. *Ann Thorac Surg* 38: 601-605.