Journal of Cardiovascular and Thoracic Research, 2012, 4(4), 123-124
doi: 10.5681/jcvtr.2012.029
http://jcvtr.tbzmed.ac.ir

Isolated Right Sided Anomalous Pulmonary Venous Connection Associated with Significant Right Ventricular Enlargement and Intact Interatrial Septum

Zahra Ojaghi Haghigh[1], Anita Sadeghpour[1], Azin Alizadehasl[2*]

[1]*Rajaie Cardiovascular Medical and Research Center, Tehran University of Medical Sciences, Tehran, Iran*
[2]*Cardiovascular Research Center, Tabriz University of Medical Sciences, Tabriz, Iran*

ARTICLE INFO

Article Type:
Case Report

Article History:
Received: 2 Sep 2012
Accepted: 21 Oct 2012
ePublished: 30 Oct 2012

Keywords:
PAPVC
Right Ventricular
Atrial Septal Defect

ABSTRACT

Partial anomalous pulmonary venous connection (PAPVC) is a very rare congenital heart disease where one or more of the pulmonary veins are connected to the venous circulation. Although initially suspected with inexplicable right ventricular enlargement on transthoracic echocardiography, other modalities such as transesophageal echocardiography, CT angiography or cardiac Magnetic resonance (CMR) imaging are able to diagnosis the anatomical abnormalities. We present a 29-year-old female with moderate right ventricular enlargement and isolated right upper and middle pulmonary vein anomalous return to superior vena cava.

Case Report

A 29-year-old female with history of palpitation was evaluated. On physical examination the jugular venous pressure and carotid contour were normal. There was evidence of mild parasternal lift consistent with right ventricular (RV) enlargement with no evidence of audible cardiac murmurs. Transthoracic echocardiography (TTE) confirmed moderate RV enlargement with preserved systolic function. Transesophageal echocardiography (TEE) demonstrated the left-to-right shunt across the isolated right sided Partial Anomalous Pulmonary Venous Connection (PAPVC) to superior vena cava (SVC) by color Doppler and with following administration of agitated saline contrast, with Qp/Qs of 1.25:1. However, in our opinion the degree of RV enlargement on both TTE and TEE was disproportionate to the approved shunt, so Cardiac computer tomography (CT) confirmed the TEE diagnosis; a congenital partial anomalous connection of right upper and middle pulmonary veins to the SVC as the etiology of RV enlargement.

Discussion

PAPVC is a very rare congenital heart disease where one or more of the pulmonary veins are connected to the venous circulation. Its incidence within the general population is 0.4–0.7%. Approximately 90% of all PAPVC's arise from the right lung, 7% from the left lung, and 3% of patients have bilateral PAPVCs emanating from both lungs connecting to the SVC, the inferior vena cava, the right atrium (RA) or the innominate vein. Common congenital heart diseases associated with PAPVC include sinus venosus and secundum type atrial septal defects while PAPVC is rarely seen alone. On physical examination, patients with PAPVC may present with an elevated JVP, parasternal lift due to RV enlargement, a right sided S3 and even pulmonary artery hypertension.[1-3]

Echocardiography is the best initial modality of choice for the noninvasive diagnosis of PAPVC. TTE recognizes RA and RV enlargement as flattening of the interventricular septum in systole and diastole due to RV pressure overload, elevated pulmonary artery pressure and the presence or absence of an interatrial shunt on color Doppler and contrast study. In addition to PAPVC, other cardiac diseases that should be considered in the differential diagnosis of unexplained RV enlargement include sinus venosus defect, ostium primum or secundum ASD, or arrhythmogenic RV dysplasia. Although TTE is unable to clearly characterize the anatomy of the pulmonary veins, TEE is able to certify the presence or absence of PAPVC. If all four PVs are not identified emptying into the left atrium on TEE, CMR may provide complementary data.[3-5]

The interesting finding in our case is the significant RV enlargement with only right upper and middle pulmonary veins anomalous return to SVC; therefore, TEE and complementary CT angiography confirmed this diagnosis. Thus, as previously-mentioned[5], even one pulmonary vein anomalous return can dilate the right heart chambers. In our case, the patient with Qp/Qs of 1.25:1 on TTE, TEE

***Corresponding author:** Azin Alizadehasl, Email: alizadeasl@yahoo.com

and CT angiography were performed conservatively on an annual basis.

In conclusion a patient with unexplained RV enlargement PAPVC (one or more pulmonary veins) should be considered in the differential diagnosis. Multimodality cardiac imaging using echocardiography, CT angiography and CMR may provide a comprehensive noninvasive evaluation of PAPVC.

References

1. Okada T, Yahagi T, Miura T, Araki T, Goto T, Kawashima S, et al. [Partial anomalous pulmonary venous connection to inferior vena cava (incomplete type of scimitar syndrome) in an elderly patient]. **Kokyu To Junkan** 1993;41:297-301.

2. Al-Ahmari S, Chandrasekaran K, Brilakas E, Tahlil W, Dearani J, Malouf J, et al. Isolated partial anomalous pulmonary venous connection: Diagnostic value of suprasternal color flow imaging and contrast echocardiography. **J Am Soc Echocardiogr** 2003;16:884-9.

3. Menzel T, Lambertz H. [Partial anomalous pulmonary venous return--detection of an isolated aberrant right upper pulmonary vein into the superior vena cava with biplanar transesophageal echocardiography]. **Z Kardiol** 1994;83:306-10.

4. Kafka H, Mohiaddin RH. Cardiac MRI and pulmonary MR angiography of sinus venosus defect and partial anomalous pulmonary venous connection in cause of right undiagnosed ventricular enlargement. **AJR Am J Roentgenol** 2009;192:259-66.

5. Otto CM, Schwaegler RG. The practice of clinical echocardiography. 3rd ed. Philadelphia: Saunders, Elsevier; 2007.

