Partial anomalous pulmonary venous connection or Cor-triatriatum - A confliction between preoperative versus intraoperative diagnosis

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Abstract

Both Cor-triatriatum and partial anomalous pulmonary venous connection (PAPVC) come under the rare spectrum of congenital heart diseases. These entities may co-exist in a patient or may appear independent of each other. Since their clinical presentation remains the same, it comes down to the cardiac imaging modality used. Here we describe a case of a 45-year female presenting with exertional dyspnea and palpitations who was diagnosed with cor-triatriatum in the preoperative echocardiography which intraoperatively was identified to be PAPVC. This case report highlights the peculiarities of echocardiographic findings in this patient and the importance of multimodal imaging techniques in such cases.
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Keywords: Cor-triatriatum, Partial anomalous pulmonary venous connection, transthoracic echocardiography, transesophageal echocardiography, diagnostic dilemma

Introduction

The morphological resemblance of cor-triatriatum and partial anomalous pulmonary venous connection (PAPVC) make their diagnosis at times difficult. A fibromuscular membrane segregates the left atrium (LA) into two chambers in cor-triatriatum. The membrane may be fenestrated or complete leading to reduced blood flow across the mitral valve (MV). Cor-triatriatum may associated with ostium secundum type of atrial septal defect (OS-ASD), left superior vena cava (LSVC), and PAPVC. It presents during infancy, however, may be diagnosed incidentally during adulthood.

Similarly, PAPVC goes undiagnosed due to the lack of symptoms. Clinical suspicion is the key to early detection for preventing complications like heart failure, pulmonary hypertension, and arrhythmias. Therefore, cardiovascular imaging plays a crucial role in confirming the diagnosis.

Here we present a rare case of a 45-year-old female who was incidentally diagnosed with cor-triatriatum preoperatively which, however, intraoperatively was found to be PAPVC. We also emphasize the importance of different imaging modalities for making a definitive diagnosis.

Case report

A 45-year-old female presented with a history of exertional dyspnea. She was evaluated for post-partum hemorrhage during her last pregnancy and incidentally diagnosed to have congenital heart disease. On prompt questioning, she gave a history of mild exertional dyspnea associated with palpitations and pedal edema.

On examination, she was afebrile and normotensive. Her pulse was irregular, and she was found to be in atrial flutter (AF). Her jugular venous pressure was not raised. Bilateral pedal edema was noted. The cardiovascular examination was unremarkable. She was receiving tablet metoprolol and torsemide as treatment.

Her blood investigations were normal. Electrocardiogram showed a right axis deviation with P-pulmonale and a right ventricular (RV) hypertrophy. Her chest X-ray revealed cardiomegaly with right atrial (RA) and LA enlargement. Both lung fields were congested. The pulmonary bay was full and pulmonary arteries were prominent [Figure 1]. Preoperative transthoracic echocardiography (TTE) diagnosed the patient with a large OS-ASD and a non-obstructive fibromuscular membrane in the LA dividing the chamber in two.
Mild mitral stenosis (MS) with a MV area of 1.8 cm² and moderate tricuspid regurgitation (TR) with a right ventricular systolic pressure (RVSP) of 65 mmHg, dilated LA, RA, and RV were also documented. Her left ventricle (LV) function was normal. A cardiac catheterization revealed RA pressures of 22/5 mmHg, RV of 62/15 mmHg, and PAP of 62/15 mmHg. A pulmonary to systemic blood flow ratio (QP/QS) of 5.6 and a pulmonary vascular resistance index (PVRI) of 1.6 WU/m² was recorded. Saturation in SVC and RA was 64% and 94% respectively. A diagnosis of acyanotic congenital heart disease with increased pulmonary blood flow, OS-ASD, cor-triatriatum with mild MS and moderate pulmonary artery hypertension (PAH) with AF was made.

The patient was planned for ASD closure with resection of membrane and MV repair. Intraoperative transesophageal echocardiography (TEE) was performed, showing an OS-ASD of 2.04 cm with a left-right shunting and a fenestrated membrane in the LA [Figure 2A, B]. Inter atrial septum (IAS) was bulging towards LA, signifying elevated right sided pressures. The LA also demonstrated blood flow from two pulmonary veins (PV). The anterior and posterior MV leaflet tips were thickened but the gradient across it was only 6/2 mmHg [Figure 2C]. The tricuspid valve morphology was normal, but a mild TR was present. However, the RVSP was 60 mmHg and the main pulmonary artery was dilated to 5.4 cm size. LV systolic function was normal.

After systemic heparinization and an ACT of >480s, aorto-bicaval cannulation was established. Cardiopulmonary bypass was initiated and cardioplegia was administered. On opening the RA, it was found that right upper and lower PVs were draining into the RA while both the left PVs opened into the LA confirming the diagnosis of PAPVC. An OS-ASD was present. An incomplete membrane was also visible in the LA. However, this diagnosis of PAPVC was missed in the preoperative TTE. The ASD was closed with autologous pericardium and the right PVs diverted into the LA. After coming off CPB, TEE examination confirmed successful closure of ASD but a membrane in the LA was still visible [Figure 3A and B]. The patient was weaned from CPB successfully and the postoperative period was uneventful.

Discussion

Cor-triatriatum is a rare anomaly with a 0.1-0.4% incidence. A fibromuscular membrane divides the LA into proximal and distal chambers. This membrane is usually fenestrated, and the severity of symptoms depends on the size of the fenestration. Various classification systems have been proposed for cor-triatriatum, the simplest being the one by Loeffler. Symptoms presenting in adults are dyspnea (68%) and palpitations (27%). Patients may develop pulmonary hypertension and right ventricular failure. Differential diagnosis includes MS, LA thrombus or pulmonary venous stenosis. Definitive diagnosis is established with 2D echocardiography, contrast echocardiography, 3D echocardiography, TEE, cardiac catheterization, computerized tomography (CT), or magnetic resonance imaging (MRI).

On the other hand, PAPVC may have either one or more PVs draining into the RA. The incidence remains 0.7%. Often patients are asymptomatic, and diagnosis is made incidentally. However, if symptoms occur, they present with dyspnea on exertion, arrhythmias, and right heart failure. Echocardiography and cardiac CT are used to confirm the diagnosis. Cardiac MRI can further elucidate anatomy. Muthialu et al described a case report of a 3-month child with a cardiac variant of total anomalous pulmonary venous return and highlighted its morphological resemblance with cor-triatriatum. He stated that although these two congenital heart diseases are different, they will look similar and involve a similar surgical strategy to repair.

Since the presentation remains same, it comes down to the imaging investigations to differentiate between the two. In our patient, the age of presentation was 45 years with an accidental detection of a cardiac lesion highlighting the uniqueness of the case. Both TTE and TEE showed a membrane in the LA with a fenestration and an ASD which led to the diagnosis of cor-triatriatum preoperatively. Two PVs opened in the upper LA chamber and the rest two could not be visualized. Thus, emphasizing the importance of a detailed examination during echocardiography to identify all four pulmonary veins. Cases have been described in literature showing association of cor-triatriatum with PAPVC which could be a plausible reason for the
discrepancy in the diagnosis made.[7] The cardiac catheterisation data demonstrated a higher saturation in RA of 94% in our patient that could be explained either by a left-right shunting across ASD or by the opening of pulmonary veins in the RA due to PAPVC. The shunting across ASD could also interfere with PV flow in the RA resulting in the inability to diagnose PAPVC. Further in favor of PAPVC was the RVSP of 60mmHg with only a mild TR implying a significant component of pulmonary hypertension. Since only TTE was used before surgery, a correct definitive diagnosis might have been missed. The presence of a poor far-field resolution and overlapping lung tissue obstructs adequate pulmonary vein evaluation during TTE.[8,9]

Thus in conclusion, if all PVs drainage is not visible on echocardiography of patients with cor-triatriatum then a high level of suspicion is required to rule out associated anomalous pulmonary venous connection using different further cardiac imaging like 3D TEE, high-resolution CT, or MRI which would provide excellent diagnostic information.

References

5. Slight RD, Nzewi OC, Buell R, Mankad PS. Cortriatriatum sinister presenting in the adult as mitral stenosis: An analysis of factors which may be relevant in late presentation. Heart Lung Circ. 2005;14:8–12.

Figures and legends:
Figure 1: Preoperative Chest Xray of the patient showing cardiomegaly, prominent pulmonary vessels, and pulmonary plethora.

Figure 2: (A)-TEE image ME 4C color compare view showing the incomplete membrane (2 arrows) in the left atrium dividing it into two chambers. (B)- TEE image showing 2D deep transgastric view with an ASD and a complete membrane like structure visible in LA. (C)TEE image ME 4C color compare view showing thickened AML and PML and blood flow across mitral valve .IAS is bulging towards to left atrium which signifies elevated Right sided pressures. Two left side pulmonary veins flow towards LA are also seen.
Figure 3: (A)-TEE image ME 4C color compare view showing an incomplete membrane still visible in the LA in the post-CPB period. (B)-TEE image showing a 2D Deep TG image of the LA with pulmonary veins giving a membrane like appearance even in the post CPB period. ASD is not seen anymore.