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CASE REPORT

Tetralogy of Fallot with an anomalous origin of right coronary artery from the pulmonary artery in a case of type A aortopulmonary window—A diagnostic challenge

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ABSTRACT

Aortopulmonary (AP) window is a conotruncal defect with an incidence of < 1% amongst congenital heart defects. It often occurs in association with other congenital cardiac defects. Coronary artery anomalies in association with the AP window are infrequent. An anomalous right coronary artery (RCA) from the pulmonary artery (ARCAPA) is an extremely rare entity with 0.002% incidence in the general population. To the best of our knowledge, the triad of Tetralogy of Fallot (TOF), ARCAPA and AP window is not yet reported in the literature.

Keywords: tetralogy; AP window; anomalous right coronary artery from the pulmonary artery

1. Introduction

Aortopulmonary window (AP window) is a rare conotruncal defect with an incidence of < 1% amongst congenital heart defects[1]. Its association with coronary artery anomalies or Tetralogy of Fallot (TOF) is rare[2]. The presence of an AP window masks the clinical features of co-existent TOF or coronary anomalies thus posing a diagnostic challenge[3].

We present the diagnostic challenges faced in an eight-month-old infant having AP window, TOF and anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA).

2. Case history

An eight-month-old infant weighing 5.2 kg, a diagnosed case of ventricular septal defect (VSD) was refereed to out institute for the surgical closure of the VSD due to failure to thrive and recurrent respiratory tract infections.

Clinical examination showed, a saturation (SpO2) of 95% with a heart rate of 140/min. The child was tachypneic with chest indrawing. Auscultation showed a continuous murmur in the left second intercostal space and bilateral equal air entry. The chest X-ray illustrated cardiomegaly with RV (Right Ventricle) apex and bilateral plethoric lung fields (**Figure 1**). ECG showed biventricular hypertrophy. 2D echocardiography demonstrated a large anterior malaligned subaortic VSD with > 50% aortic override, proximal AP window,

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and good size confluent branch pulmonary artery anatomy with mild flow acceleration across the pulmonary valve (Gmax 25 mm Hg), unobstructed right ventricular outflow tract (RVOT) (**Figure 2**). Hence, the patient was planned for VSD and AP window closure.

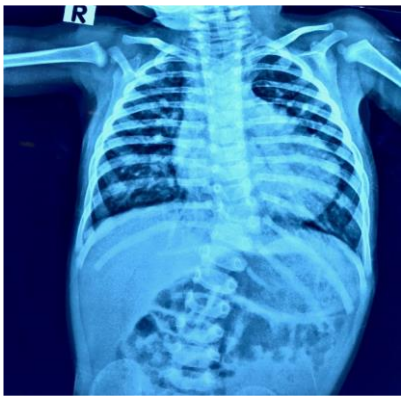

Figure 1. Chest X-ray.

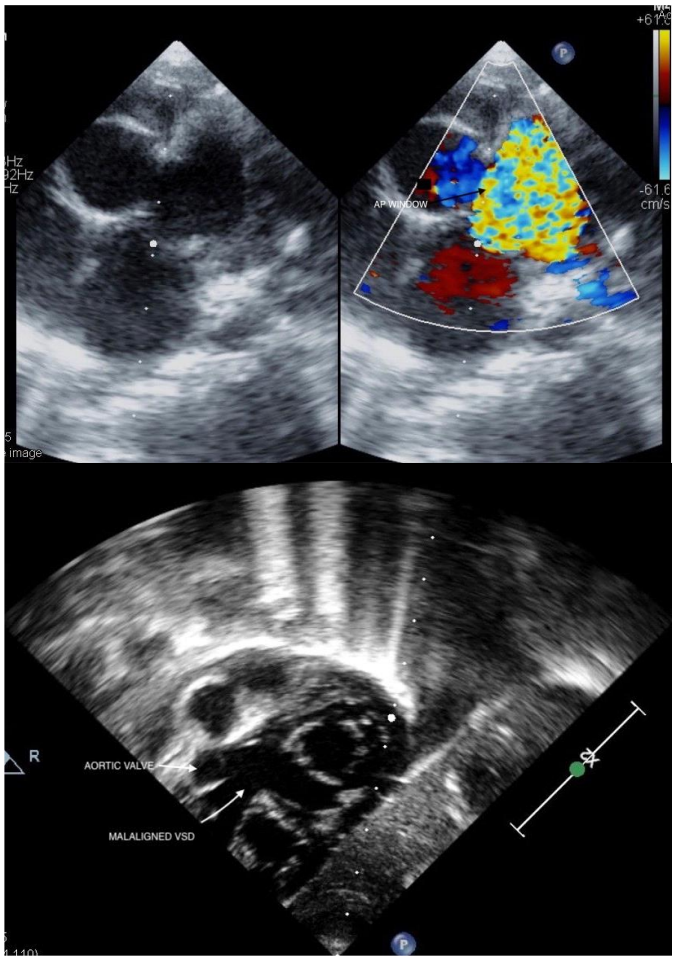

Figure 2. Preoperative 2D echocardiography.

APW—Aortopulmonary window.

Intraoperative inspection revealed higher origin of the right coronary artery (RCA). After a standard cardiopulmonary bypass (CPB) was established following the mobilization of the aorta and pulmonary arteries. Both the pulmonary arteries were snared, the aorta was cross clamped, and cardioplegia was delivered. The heart was vented through the patent foramen ovale (PFO). After the heart was arrested, a longitudinal

aortotomy was performed and the AP window was visualized from the aortotomy. The RCA origin was absent on the aortic side. Hence, the main pulmonary artery (MPA) was opened longitudinally, and we found that the RCA origin proximal to the type I AP window. The course of RCA was intramural in the aorta, which was unroofed. The AP window was closed from the MPA side, and the aortotomy was closed in 2 layers. Trans-atrial closure of the VSD was done used tanned autologous pericardium. The MPA was closed in 2 layers following de-airing and cross clamp removal. The heart resumed in sinus rhythm, CPB was terminated uneventfully with stable hemodynamics and minimal inotropic support (milrinone 0.5 mcg/kg/min and adrenaline 0.05 mcg/kg/min). However, the central venous pressure (CVP) remained slightly higher. Transesophageal echocardiography (TEE) showed no residual intracardiac lesions with good biventricular function.

The postoperative 2D Echocardiography showed hypertrophied infundibulum with severe subvalvular and valvar pulmonary stenosis, RVOT gradient of 90 mm Hg. Hence patient was taken back to the operating room and RVOT resection was performed. Though the pulmonary valve was slightly dysplastic, it admitted size adequate sized Hegar ($Z = 0$). Thus, it was left untouched. The CPB was terminated with a pRV/pLV of 0.6 and a RVOT gradient of 30 mm Hg with 2D echocardiography showing well opened RVOT (**Figure 3**). The postoperative period was uneventful, and the patient was extubated after two days of elective ventilation and discharged on postoperative day 10. The child is under follow up and doing well.

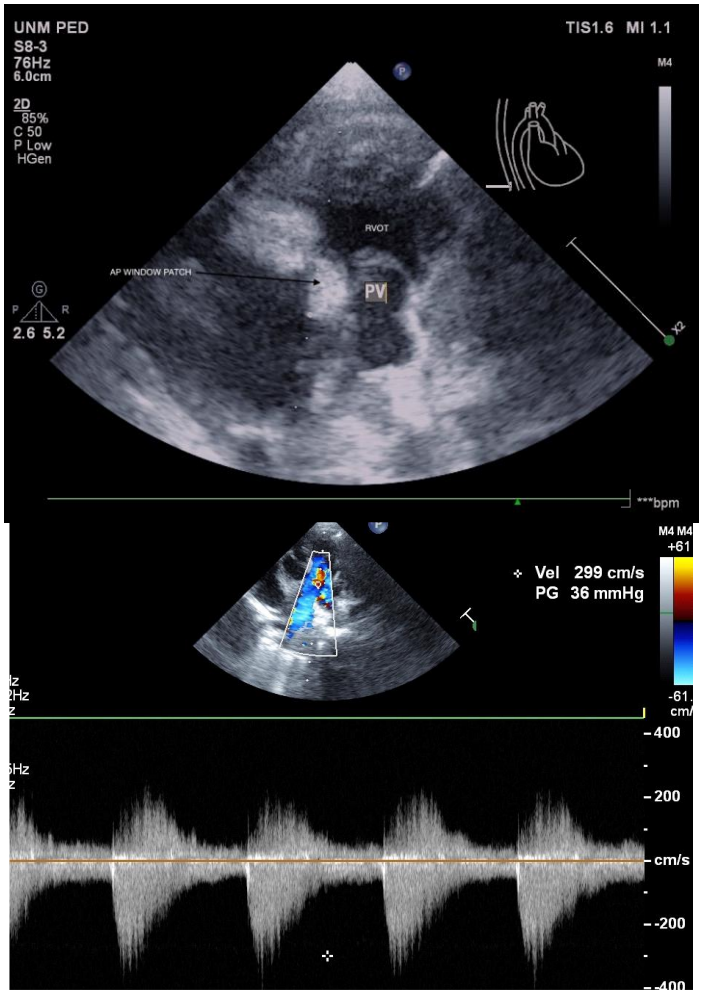

Figure 3. Postoperative 2D echocardiography.
PV—Pulmonary valve, RVOT—Right ventricular outflow tract.

3. Discussion

Aortopulmonary window is an uncommon congenital cardiac defect with an incidence of < 1%. It can occur in association with TOF and rarely can have coronary anomalies. This case is unique because of the co-existence of all three lesions in the same patient. The presence of an AP window masked the diagnosis of TOF and ARCAPA preoperatively owing to the large left to right shunt[3]. Because of large left to right shunt and severe pulmonary artery hypertension, there is hardly any antegrade flow across the RVOT which makes diagnosing TOF a challenge. Also, the features of ARCAPA may not be manifested due to high PA pressure[4].

The patient presented with history of repeated respiratory tract infections classical of a large left to right shunt, no history of cyanosis or cyanotic spells. There was no ejection systolic murmur auscultated in the pulmonary area. Pre-operative chest X-ray showed bilateral plethoric lung fields.

The only feature which could have raised the suspicion of TOF in this case was the anteriorly malaligned VSD. The presence of an anteriorly malaligned VSD requires further assessment for RVOT obstruction[5].

Thus, to conclude, the presence of an anteriorly malaligned VSD with an AP window warrants a thorough search for any other associated anomalies like Tetralogy of Fallot. Further, coronary anomalies although rare, can still occur in an AP window, and one should be prepared to tackle one if present.

Conflict of interest

The authors declare no conflict of interest.

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