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Case Report

Computed tomography features of supracardiac total anomalous pulmonary venous connection in an infant

Tariq Alam MMBS, FCPS[a], *Hidayatullah Hamidi MD*[b,*], *Mer Mahmood Shah Hoshang MD*[b]

[a] *Department of Radiology, Northwest General Hospital Peshawar, KPK, Pakistan*
[b] *Department of Radiology, French Medical University for Children (FMIC), Kabul, Fifth district, 1006 Kabul, Afghanistan*

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ABSTRACT

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital anomaly of the pulmonary veins drainage. In this entity, the pulmonary veins, instead of draining to left atrium, connect abnormally to the systemic venous circulation. A right-to-left shunt is obligatory for survival. Based on its type and degree of pulmonary venous obstruction, TAPVC may result in pulmonary hypertension and congestive heart failure. In severe cases, urgent diagnosis and surgical correction is essential to reduce morbidity and mortality. Echocardiography as the first and safest imaging modality for cardiovascular abnormalities may fail in complete depiction of some complex feature of TAPVC. Computed tomography angiography is then a noninvasive and sensitive choice for mapping the pulmonary veins without the need for invasive cardiac catheterization. Contrast-enhanced MR angiography can be a radiation-free alternative. Authors present a computed tomography–detected supracardiac TAPVC with small patent ductus arteriosus in a 2 months cyanotic infant.

Case presentation

A 2-month-old female infant was sent to radiology department to undergo chest computed tomography (CT) angiography for evaluation of pulmonary veins (PVs). The child developed cyanosis in her first week of life. Unfortunately, neither prenatal ultrasound nor postnatal echocardiography reports were available.

Chest CT angiography was performed with 128-slice Siemens scanner after administration of 8 mL of nonionic water-soluble contrast material.

The CT findings were as follows:

All 4 PVs were joining together behind the small left atrium (LA; Fig. 1) and their confluence converging to a large vertical vein (VV) that was ascending lateral to the pulmonary trunk. LA was smaller in size and no PV entrance was noted into it (Fig. 2). The VV was draining into the dilated left innominate vein and subsequently to enlarged superior vena cava (SVC) and right atrium (RA; Fig. 3). The RA and right ventricle (RV) were also dilated (Fig. 3).

A small (approximately 6.5 mm) atrial septal defect (ASD) was detected (Fig. 2). The small ASD was functioning as right-

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* *Corresponding author.*
E-mail address: Hidayatullah.hamidi@gmail.com (H. Hamidi).

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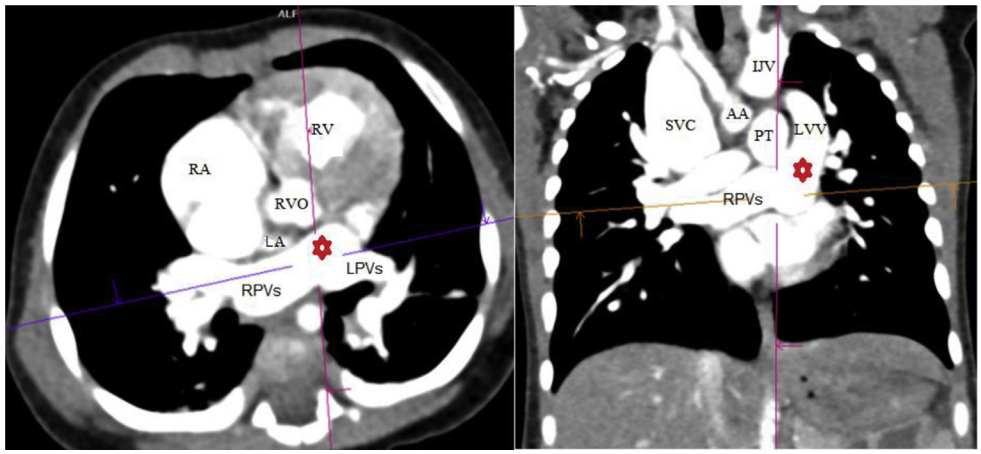

Fig. 1 – Axial (right) and coronal (left) postcontrast computed tomography sections: the right pulmonary veins (RPVs) and left pulmonary veins (LPVs) join all together posterior to the left atrium (LA) and left ventricular outlet (LVO). The confluence of PVs (red star) converges into a large left vertical vein (LVV) that is ascending lateral to the pulmonary trunk (PT). The superior vena cava (SVC), right atrium (RA), right ventricle (RV), and even left internal jugular vein (IJV) are dilated. The pulmonary truck (PT) is prominent compared to the ascending aorta (AA) at the same level.

to-left shunt and made this supracardiac total anomalous pulmonary venous connection (TAPVC) compatible with life.

No evidence of patent ductus arteriosus (PDA) or ventricular septal defect (VSD) was observed.

Discussion

Background

TAPVC is a rare congenital anomaly of the PVs drainage that connects abnormally to the systemic venous circulation instead of LA. [1] The incidence of this rare entity is approximately 1 of 17,000 live births [2] or 1%-5% of all cardiovascular congenital anomalies [3].

TAPVC is caused by nonfusion of pulmonary venous confluence with LA, thus, the oxygenated pulmonary blood is redirected to right heart and pumped once again to the lungs [4]. A right-to-left shunt is obligatory for survival that typically occurs at atrial level through either an ASD, patent foramen ovale [3] complete absence of the atrial septum [4] or less commonly a PDA [3]. Some of the mixed blood of the RA crosses through this shunt to the left heart to supply the systemic circulation [4].

Cases of tetralogy of Fallot and double-outlet RV are reported in association with TAPVC [5]. Higher frequency of TAPVC is observed in patients with heterotaxy syndromes especially asplenia [6].

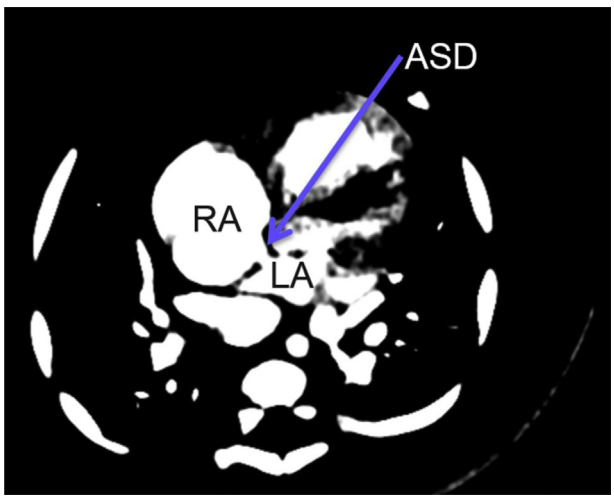

Fig. 2 – Reformatted axial image at atrial level: the right atrium (RA) is dilated, whereas the left atrium (LA) has smaller size. The small atrial septal defect (ASD) is well depicted. The congested lung vessels can also be seen.

Clinical perspective

TAPVC is a cause of neonatal cyanosis, and if right-to-left shunt is not present, it may rapidly result in death [3]; hence, being a surgical emergency in neonates [2].

In severe cases, urgent diagnosis and surgical correction is essential to reduce morbidity and mortality [2]. However, if there is no obstruction and a sufficient shunt is present, patients may not present to clinic urgently [7].

TAPVC can occasionally be misdiagnosed as persistent pulmonary hypertension of the newborn and even echocardiographic findings may overlap [7].

Classification

Based on the location of pulmonary venous drainage, TAPVC is classified into 4 major types [1,3,4]. In all 4 types, complete drainage of pulmonary venous blood is directed to the right heart [4].