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Case Report

MDCT Imaging of Stenotic Total Anomalous Pulmonary Venous Connection to the Right Superior Vena Cava in a 4.5 Months Infant

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Abstract

Total anomalous pulmonary venous connection (TAPVC) is an uncommon anomaly classified according to the site where the drainage to the systemic circulation occurs: supracardiac, cardiac, and infracardiac drainage. In superior TAPVC, the pulmonary veins from each lung join to form a venous collector with an ascending common vessel, and opens into the posterior aspect of the superior vena cava. We present a case of stenotic supracardiac total anomalous pulmonary venous connection, that is one of the rarest forms of presentation. This is clinically serious and needs urgent surgical correction. MDCT allows to perform an adequate diagnosis of anomalous drainage, and an adequate surgical approach with very little exploration time. In our opinion, CT MD is the technique of choice in the diagnosis of this entity.

Keywords: *Congenital Heart diseases; Total Anomalous Pulmonary Venous Connection, Multiple Detector Computed Tomography (MDCT)*

Introduction

Total anomalous pulmonary venous connection (TAPVC) defines the anomaly in which the pulmonary veins have no direct connection with the left atrium and, drain into one of the systemic veins or in the right atrium. Total anomalous pulmonary venous connection (TAPVC) is an uncommon anomaly accounting for 1.5 - 3% of congenital heart diseases and classified according to the site where the drainage to the systemic circulation occurs: supracardiac, cardiac, and infracardiac drainage. In superior TAPVC, the pulmonary veins from each lung join to form a venous collector located in the right or left hemithorax with an ascending common vessel ascends, passes anterior to the hilum, and opens into the posterior aspect of the superior vena cava (SVC). In rare cases, the ascending pulmonary venous vessel connects to the azygos vein [1,2]. The obstructed form of supracardiac drainage is an unusual and severe supracardiac variant [3].

Case Report

We present a case of a baby girl, with 4.5 months of age and 6 kg. She was referred to the hospital because of poor feeding and poor weight gain during last month. Physical examination disclosed tachycardia, hyperactive precordium and very mild liver enlargement. Cyanosis was not evident but pulse oximetry revealed mild desaturation (O_2 Sat: 88%) unresponsive to supplementary oxygen. Patient did not need ventilatory or inotropic support at any time before admission. Plain chest radiograph on admission showed cardiomegaly and pulmonary plethora. Color Doppler 2D echocardiography showed anomalous drainage of pulmonary veins to a common collector that coursed behind the atria to the SVC (Figure 1). A contrast enhanced Computed Tomography (MDCT) of the heart showed all four pulmonary veins joining in a large unobstructed common collector that coursed behind the atria and superiorly to join the SVC next to the drainage of azygous vein (Figure 2). It courses cranially and passes between the right pulmonary artery (anterior) and the right main

stem bronchus (posterior). These structures produce an extrinsic obstruction to pulmonary venous flow. The blood drained with difficulty to the SVC and pooled in the pulmonary veins causing a sub-acute pulmonary edema. Right SVC was largely dilated and there was a normal course of the innominate vein. The MDCT showed a significant decrease in caliber of the main bronchi, given the situation of the dilated collector between the bronchus and the pulmonary arteries. Drainage of inferior cava vein and suprahepatic veins were normal. Interatrial septum was shifted to the left and there was an atrial septal defect shunting from right to left. Intracardiac anatomy was otherwise normal. Surgery was uneventfully performed on cardiopulmonary bypass and moderate hypothermia: a wide anastomosis was created between the common collector and the left atrium, the vertical vein was ligated close to its connection to the SVC and the atrial septal defect was closed. Postoperative echocardiogram showed right pulmonary veins draining into the left atrium without significant stenosis or residual shunt. There was good biventricular contractility, and mild to moderate tricuspid regurgitation. Postoperative plain chest radiograph showed plethora resolution (Figure 3).

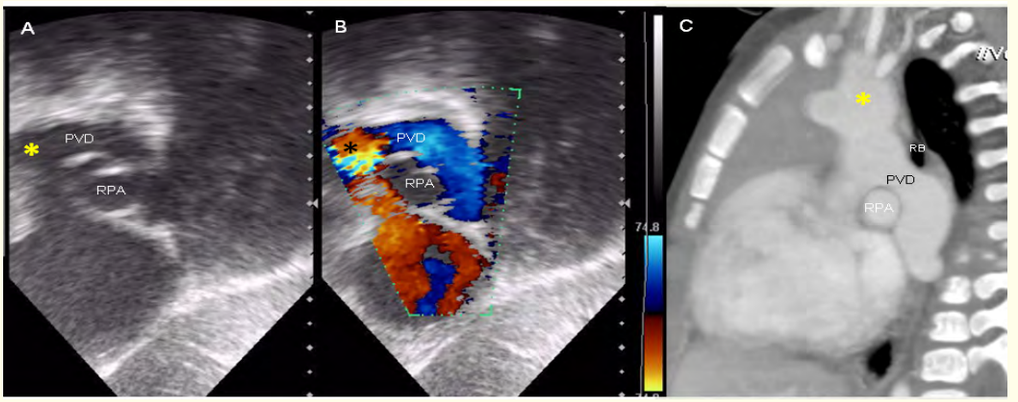

Figure 1: *Images of echocardiography 2D (A), and Doppler color (B). Contrast enhanced MDCT reformatted sagittal MIP image (C). The venous collector courses cranially, and passes between the right pulmonary artery and the right main stem bronchus. Remember that it is customary to follow the course of the azygous over the right main bronchus. PVD (pulmonary venous drainage), RPA (right pulmonary artery), RB (right bronchus), * (superior vena cava).*

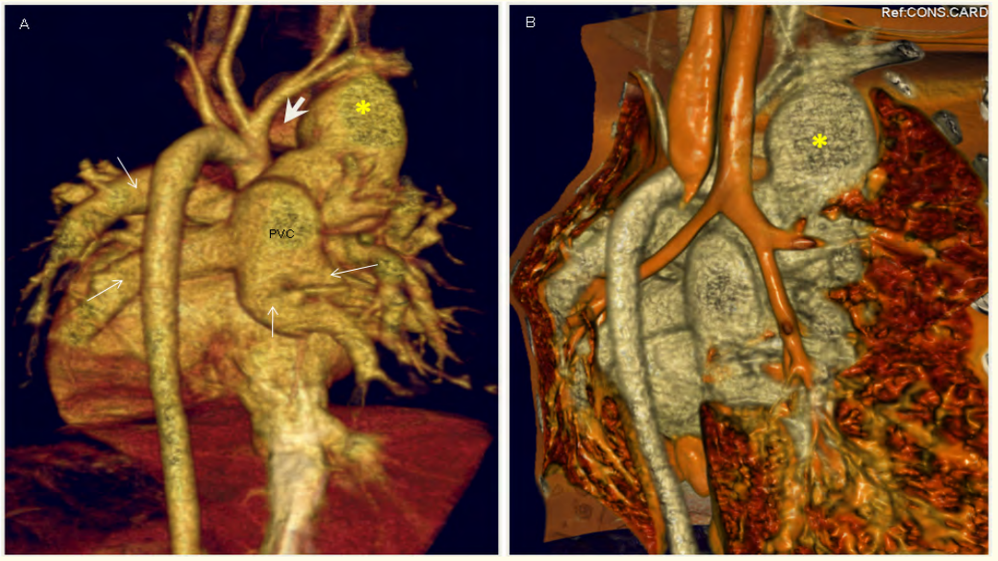

Figure 2: *Contrast enhanced MDCT. VR-3D, posterolateral views (A and B). A. The study shows a total anomalous pulmonary venous drainage with retrocardiac collector. Four pulmonary veins drain in this collector (white arrows) which courses cranially to join the posterior aspect of the right superior vena cava (*), below the arch of the azygous vein. There is massive dilatation of the superior cava vein. The innominate vein maintains a conserved gauge (arrow head). Azygous system dilatation is not observed. B. The strange thing in this case is the crossing of the collector behind and below the right main bronchus, opening the angle of the carina and reducing the anteroposterior diameter of the airway. PVC (pulmonary venous collector), RB (right bronchus), PVD (pulmonary venous drainage), RPA (right pulmonary artery).*

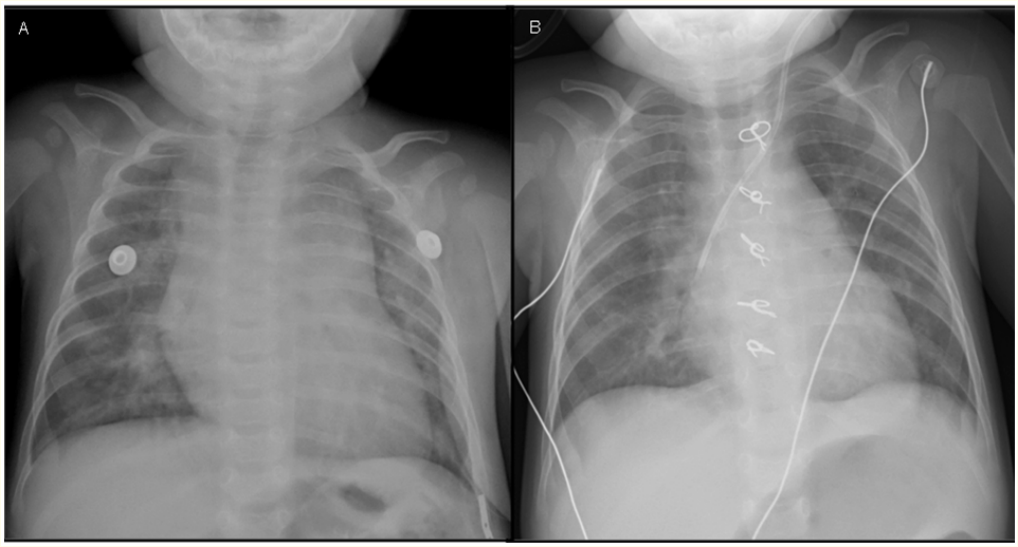

Figure 3: *Pre and postsurgical Plain Chest X-Rays. A. Marked cardiomegaly with enlargement of upper mediastinum secondary to increased pulmonary flow, and venous collector. B. Postoperative X-ray shows significant decrease of the size of the heart, and mediastinum shape.*

Discussion

The most frequent stenotic anomalous drainage is the infradiaphragmatic type, being the supracardiac case a very rare event [3]. MDCT is a powerful preoperative diagnostic tool for patients with TAPVC because it can provide excellent visualization of the venous anatomy and also the airways tree with reconstructed 3-D images from any angle, revealing information that may alter the surgical approach, like in our case. We could display the presence of TAPVC and evaluate the number of pulmonary veins and draining sites of the common pulmonary venous trunk on helical CT angiography. Other diagnostic modalities such as MR imaging and echocardiography are known to be useful for the evaluation of a TAPVC, but CT angiography may be the best option to perform the diagnosis and the appropriate surgical approach. In addition, it allows the study to be carried out in a very short time, which is essential in patients with an unstable clinical situation [4].

Conclusion

Total anomalous pulmonary venous connection is an uncommon anomaly and classified according to the site where the drainage to the systemic circulation occurs: supracardiac, cardiac, and infracardiac drainage. In superior TAPVC, the pulmonary veins from each lung join to form a venous collector with an ascending common vessel, and opens into the posterior aspect of the superior vena cava. Stenotic supracardiac total anomalous pulmonary venous connection is one of the rarest forms of presentation, like our case. This is clinically serious and needs urgent surgical correction. MDCT allows to perform an adequate diagnosis of anomalous drainage, and his anatomic relations. In addition, an adequate surgical approach can be performed with very little exploration time. In our opinion, MDCT is the technique of choice in the diagnosis of this entity.

Disclosure

The authors declare no conflict of interest.

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