Total anomalous pulmonary venous connection with descending vertical vein: Unusual drainage to azygos vein

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ABSTRACT

Most patients with total anomalous pulmonary venous connection have a set pattern of pulmonary venous drainage and predictable sites of obstruction. However, uncommon variations do exist and delineating the entire course is more important than just knowing the site of drainage. Azygos vein involvement in the circuit is nearly always associated with a complicated course, as was seen in our patient. This report reviews the drainage patterns when azygos vein forms a part of the circuit.

Keywords: Azygos vein, infracardiac, total anomalous pulmonary venous connection, vertical vein

INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) is an admixture lesion wherein whole of pulmonary venous return mixes with systemic venous return before getting into aorta. The site of anomalous connection to the systemic venous system varies and forms the basis of classification into supracardiac, cardiac and infracardiac variants. The presence or absence of obstruction in the circuit including at the site of atrial septal defect (ASD) further impacts the clinical presentation and outcome. The likelihood of obstruction varies with the site of drainage and forms the basis of timing of surgical correction. The importance of complete delineation of drainage path for surgical correction cannot be overemphasized. We hereby present a case of a neonate with clinical presentation consistent with obstructed TAPVC and an obstructed descending vertical vein. However, contrary to the expectation the drainage was into azygos vein, an uncommon site of drainage which is almost always obstructed.

CLINICAL PRESENTATION

An 8-day-old male neonate presented to us with rapid breathing and cyanosis. Tachypnea, tachycardia, cyanosis and heart failure were evident on examination. Chest radiograph showed a normal-sized heart with severe pulmonary venous hypertension. Clinical picture was suggestive of an obstructed TAPVC. A trans-thoracic echocardiogram confirmed absent pulmonary venous connections to left atrium, a non-restrictive ASD flowing right to left and severe pulmonary arterial hypertension (predicted right ventricular systolic pressure = 76 + right atrial mean). All four pulmonary veins joined to form a common chamber that drained inferiorly through a vertical vein showing low-velocity downward flow. Surprisingly, this vertical vein could not be imaged crossing the diaphragm and, instead, the superior vena cava (SVC) was found to be dilated. Turbulent flow (continuous gradient of 18 mmHg) could be visualized further downstream, but the exact site of obstruction was not clear on echocardiography. A subsequent computed tomography (CT) angiogram (CTA) confirmed the presence of a descending common chamber, lying to the left of the spine. This chamber formed a vertical vein that traversed across the spine to join the azygos vein just above the diaphragm [Figures 1 and 2]. The dilated azygos vein drained into the SVC at its usual position. There were two sites of severe stenoses, one within the vertical segment and other at the site of entry into azygos vein. Findings were confirmed at surgical repair, wherein the common chamber was opened into the left atrium and the vertical vein was ligated. The child had an uneventful post-operative course.

DISCUSSION

In early embryonic life, blood coming from the lung

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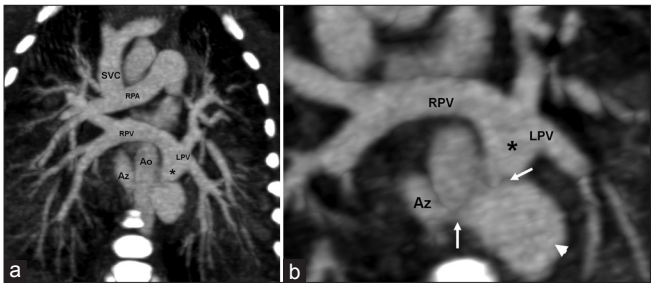

Figure 1: (a) Coronal oblique thick multi-planar reconstruction (MPR) of computed tomography angiogram (CTA). This reveals the right- and left-sided pulmonary veins (RPV and LPV, respectively) joining together to form a descending common chamber (*) to the left of the thoracic aorta (Ao). Increased attenuation along bronchovascular bundles in both lungs is consistent with pulmonary edema. (b) Zoomed-up view of a thin MPR in the same plane shows stenoses at two sites in the common chamber (*). The first obstruction (white arrow) is followed by a dilatation (white arrowhed). The second narrowing (white arrow) is at the site of drainage into azygos vein (Az)

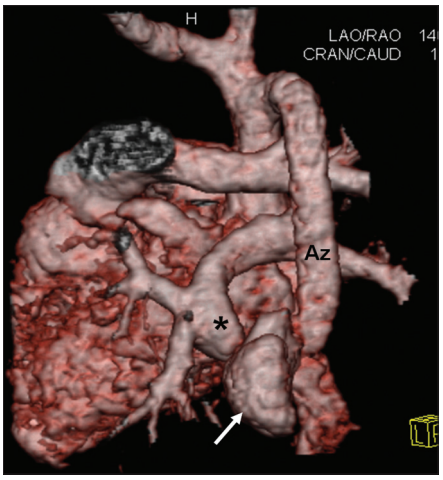

Figure 2: Volume-rendered technique of computed tomography angiogram in the left posterior oblique view. This provides an overview of the descending common chamber (*), dilatation (white arrow) and drainage into azygos vein (Az)

buds drains to the splanchnic plexus that connects to the paired common cardinal and umbilico-vitelline veins. Failure of the common pulmonary vein, arising from primitive lung buds, to connect to the pulmonary venous plexus leads to the persistence of one or more of these primitive venous connections.[1] Depending on the final drainage site of the pulmonary veins or the vertical vein formed by these veins, the defect is divided into four types: supracardiac (50%), intracardiac (25%), infracardiac (20%) and mixed type (5%). Unusual drainage patterns do exist, especially when the azygos vein gets involved anywhere along the course.[2]

TAPVC draining into the azygos vein was first reported by Edward *et al.*[3] in 1953. This variant accounts for 6.3% of the cases, and is almost always obstructive.[4] Entry of the vertical vein into the azygos vein can occur at any one of these sites: (a) arch of azygos vein, (b) junction of proximal and middle third of azygos vein, (c) middle third of azygos vein and (d) just above the diaphragm. A descending vertical vein is seen only when the vertical vein joins the azygos vein just above the diaphragm.[5] The descending vertical vein does not cross the diaphragm in contrast to infradiaphragmatic TAPVC; the only other similar anomaly is when the vertical vein drains directly into a supra-diaphragmatic inferior vena cava (IVC). However, drainage in supra-diaphragmatic IVC is usually unobstructed. This distinction is thus important for timing the surgical repair and prognosis.[6]

Differentiating azygos vein drainage of the pulmonary veins from a direct drainage into SVC is important because of a potential risk of injury to right pulmonary artery (RPA) and/or right bronchus during surgical repair of the latter. When the vertical vein joins the SVC directly, it opens into the lower part of SVC, coursing anterior to the RPA. In contrast, when drainage is through the azygos vein, it runs posterior and superior to the RPA, joining the SVC at a higher level. These details are not seen easily on echocardiography, and this is where a CTA is immensely useful. The main advantages of multi-slice CTA in congenital cardiac anomalies like TAPVC are the relative ease and accuracy with which the diagnosis can be made, as also the speed with which the procedure can be carried out. Unlike transesophageal and angiocardiographic images, which are difficult to provide three-dimensional orientation, the 3D rendering and the multiplanar images give a clear three-dimensional picture to the surgeon of what one is likely to find on the operating table.

Our case highlights the importance of delineating the complete anatomy of the pulmonary venous circuit in patients of TAPVC. Involvement of the azygos vein in the course should prompt a CTA unless the complete course is easily defined by echocardiography.

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