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Case Report

Infracardiac Type Total Anomalous Pulmonary Venous Connection. A Clinical Case Presentation and Literature Review

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Abstract

The Total anomalous connection of pulmonary veins (TAPVC) of the infracardiac type is a rare pathology whose diagnosis is achieved by transthoracic echocardiography (TTE). However, due to its limitations, thorax angiotomography (TAT) emerged as the preferred complementary modality due to its high resolution, as well as multiplanar and 3D reconstruction capabilities, which are essential for correct anatomical evaluation and surgical planning.

We present the clinical case of a pediatric patient in whom TAT was performed in addition to a TTE study to determine the size, number, and connection of the pulmonary veins.

Keywords: Total Anomalous Connection of Infracardiac type Pulmonary Veins; Transthoracic Echocardiography; Congenital Heart Disease; Computed Tomography Angiography of the Chest; Thorax Angiotomography (TAT).

Abbreviations

TAPVC: Total Anomalous Connection of Pulmonary Veins; TAT: Thorax Angiotomography; TTE: Transthoracic Echocardiography; MIP: Maximum Intensity Projection; PDA: Patent Ductus Arteriosus; ASD: Atrial Septal Defect

Introduction

Total anomalous connection of pulmonary veins (TAPVC) is a congenital heart disease characterized by the absence of union between the pulmonary veins and the left atrium of the heart [1-5]. It consists of an anomalous connection between the pulmonary veins with the right atrium, or with one or more systemic veins [6-8].

Darling., *et al*. [7] classified TAPVC into 4 types: supracardiac, cardiac, infracardiac, and mixed, depending on whether the connection site(s) of the draining vein is with the systemic veins, the right atrium, or a combination [6,9,10].

In infracardiac type TAPVC there is a retro pericardial venous collector in which the pulmonary veins of both lungs converge and drain through a "vertical vein" that travels with the esophagus, crosses the diaphragm, and joins a suprahepatic vein, venous, inferior vena cava, azygos system, splenic vein, superior mesenteric vein or portal vein. In this pathology, the body of the left atrium is hypoplastic, with a morphologically normal mitral valve and left ventricle or with a certain degree of hypoplasia. This type of TAPVC has a high incidence of pulmonary venous obstruction that can lead to premature death if a correct diagnosis and early surgical intervention are not performed [2,8].

The purpose of this study is to present the case of a two-month-old boy diagnosed with infracardiac type TAPVC since it is a rare congenital heart disease with a high percentage of complications and mortality, which makes it important to have extensive knowledge to interpret the findings accurately. It is also intended to highlight the usefulness and importance of thorax angiotomography (TAT), as a complementary imaging study to transthoracic echocardiography (TTE), in which a specific evaluation of the anatomical details of this pathology can be carried out since they are crucial to guide decision-making and surgical planning.

Case Presentation

A 2-month-old patient, who was born from a normal pregnancy at 40 weeks of gestation by eutocic delivery with a weight of 3050 g, breathed and cried at birth.

The symptoms begin one month after birth with failure to thrive, respiratory distress, progressive central cyanosis, and worsening alertness, which was the reason for medical attention.

Physical examination at the emergency department revealed respiratory distress, hypoactivity, poor tissue perfusion, and central cyanosis, requiring ventilatory mechanical support. The precordium is rhythmic, and hypodynamic, with a systolic murmur in the fourth intercostal space, grade III/VI, with a reinforced second heart sound (S2). Presence of hepatomegaly, thready peripheral pulses, and delayed capillary refill.

It is assessed by the pediatric cardiology service finding infracardiac type TAPVC in the transthoracic echocardiography, for which TAT was requested to establish the size, number, and connection of pulmonary veins, where positive findings were reported: infracardiac type TAPVC with portal connection, patent ductus arteriosus (PDA), atrial septal defect (ASD) and bilateral lower lobe atelectasis (Figures 1-4).

Figure 1: 3D reconstruction. Posteroanterior view of the confluence of the four pulmonary veins (white arrowheads) to the collector (white arrow), which drains into the portal vein (gray arrowhead) through the vertical vein (grey arrow).

Figure 2: TAT with MIP. ASD (green arrow) left retro atrial collector (white arrow) and atelectasis in the lower lobes of both lungs (red arrowheads) are observed. Central venous catheter beam hardening artifact (asterisk). TAT: thorax angiotomography (TAT). MIP: Maximum Intensity Projection. ASD: atrial septal defect.

Figure 3: TAT, oblique coronal reconstruction with MIP. Anteroposterior view of the confluence of the four pulmonary veins (red arrowheads) to the left retro atrial collector (blue arrow), which drains into the portal vein (black arrow) through the vertical vein (white arrow). TAT: thorax angiotomography (TAT). MIP: Maximum Intensity Projection.

Figure 4: TAT, oblique sagittal reconstruction with MIP. Aortic arch (white arrow), PDA (green arrow), and left pulmonary artery (red arrow) are identified. The left retro atrial collector (blue arrow) and the vertical vein (black arrow) are observed. ATCT: chest computed tomography angiography. MIP: Maximum Intensity Projection. PDA: patent ductus arteriosus.

The suggested management plan was surgical correction as soon as possible.

Discussion

TAPVC results from failure of the common pulmonary vein to fuse with the left atrium or atresia of the common pulmonary vein, with the persistence of the primitive splanchnic vein connections of the pulmonary veins to the systemic cardinal and/or umbilical vitelline veins, reaching flow that communicate to the right atrium. Infracardiac-type TAPVC (as in the case of our patient) is the result of a persistent connection with the umbilical vitelline veins [4,5,11-14].

TAPVC is a heart disease with mixed shunt, which in hearts with atrioventricular and ventriculoarterial concordance represents 1-3% of congenital heart diseases [2], with a reported incidence of 7 per 100,000 live births [5]. It is a cyanotic condition with the lowest prenatal detection rate [5,11], as in our patient, in whom the diagnosis was made two months after birth. In 40% of patients, it is usually associated with heterotaxy syndrome, right isomerism, and polysplenia or asplenia [7].

In general, TAPVC can occur in isolation (with ASD, PCA, or both, as in the case of our patient) or associated with other cardiac malformations [15]. For the postpartum survival of patients, the presence of a right-left shunt is necessary, which can be through an atrial septal defect (ASD) of variable size. When the atrial septum is rarely intact, there is no shunting of blood from the right to the left side of the heart, which causes instant death [1,3,7,13].

Of the four types of TAPVC, infracardiac (as in the case presented) represents 25% [1,2]. The four pulmonary veins drain into a confluence posterior to the left atrium and a vertical vein, originating from this site, descends anterior to the esophagus and passes through the esophageal hiatus of the diaphragm, to drain into the portal vein, which is the most frequent site, as in this patient (Figures 3 and 4); to a lesser extent, it can drain into the suprahepatic veins, ductus venosus, inferior vena cava, azygos system, splenic vein, or superior mesenteric vein [3,6,12,13]. In this type, an inverted Christmas tree pattern is described, formed by the vertical orientation of the confluence of the superior and inferior pulmonary veins, which join separately along the vertical vein (Figure 3) [5]. Venous obstruction occurs with an incidence of approximately 95-100% [8,16]. Obstruction of the vertical vein can

be anatomical, extrinsic (passing through the esophageal hiatus of the diaphragm), or intrinsic (due to collector stenosis). Functional-type obstruction occurs when the anomalous connection is in the portal vein and the high resistance of the intrahepatic blood flow prevents the passage of venous blood flow from the pulmonary drainage to the heart [3,11,12,14], as it happened with our patient. Neonates with pulmonary venous outflow obstruction usually present with respiratory distress and cyanosis, as well as congestive heart failure, which are predictive findings for mortality [1], such as what happened in our case.

The gold standard for the evaluation of TAPVC has been cardiac angiography for many years [17]. However, although a hemodynamic and anatomical evaluation can be combined during the procedure, it has disadvantages such as the dose of ionizing radiation, the difficulty in observing the systemic and pulmonary vascular system at the same time, the overlapping of adjacent vascular structures, the not being able to assess associated airway diseases and complications such as vascular dissection [6,9,15,17]. Therefore, it is necessary that for the evaluation of infracardiac type TAPVC, another effective and non-invasive imaging method should be used.

TTE is considered the initial modality since it has a sensitivity and specificity of 97 and 99% for its detection and diagnosis, with a 61% concordance with surgery [4,6,8,12,15]. A complete and reliable anatomic evaluation of all pulmonary veins and heart defects has been demonstrated by this method [4,13]. In addition, it is possible to observe the type of TAPVC, the presence of vertical vein compression, the existence of a restrictive atrial septal defect, and the blood velocity of the stenotic pulmonary vein can be evaluated to determine the severity of the obstruction [2,8]. Among the disadvantages we find that it is a dependent operator mode, the acoustic window can be poor and the field of vision is small. These can condition poor visualization of extracardiac vascular structures such as the distal aorta, supra-aortic trunks, distal portion of pulmonary arteries, pulmonary veins, and in the special case of infracardiac-type TAPVC, the drainage site, and vertical vein obstruction [6,12,13,15-17].

TTE can allow for surgical planning by the surgeon. However, when this is not enough, as in the case of not finding the vertical vein drainage site in infracardiac type TAPVC, complementary imaging studies such as TAT or thorax angiography magnetic resonance imaging (MRI) are needed in some patients [5,8,13].

MRI has a great capacity to provide anatomical and physiological information, so in addition to the fact that it does not use ionizing radiation, it could be considered the best non-invasive technique for the diagnosis of infracardiac type TAPVC. However, due to the large percentage of patients who present with data of obstruction and poor clinical status in this pathology, the long duration of the study is one of the greatest drawbacks [4,8,15,16].

After performing the TTE on our patient and not being able to adequately observe the vertical vein connection site, an TAT was requested, which is the preferred complementary imaging modality in this type of pathology, since it is an imaging technique that is noninvasive with high spatial resolution, which allows the assessment of cardiovascular and extravascular structures, widely overcoming the limitations of TTE and providing surgeons with the most information required for surgical assessment [6,8,12,14-17]. TAT is available in many radiology departments across the country, the main advantages of which are multiplanar and 3D reconstruction capabilities. In addition, rapid acquisition times, allow the urgent definition of pulmonary venous anatomy in critically ill patients with obstruction and the use of short-term or no sedation [2,4-6,9,11,15-17]. Disadvantages include poor visualization of intracardiac structures, adverse reactions from using iodinated contrast media, and high radiation exposure [4,15,16], although dose reduction techniques have been introduced in recent years [17]. Sensitivity and specificity of 100% have been found in the TAT for the correct description of the stenosis, the course, and the drainage site of the vertical vein, which makes it superior to the TTE [8].

In our patient, 64-slice multidetector computed tomography was used, which allowed a high volume of coverage allowing a more precise and rapid evaluation of the anatomy as well as less radiation exposure. It was possible to locate the anomalous connection site, the obstruction of the collector, and the atrial septal defect, which are the three central aspects of clinical and surgical interest [3].

The goal of surgery is to integrate the collector with the left atrium, whose survival rate after the correction has improved. Early mortality of less than 10% has been reported and reported incidences of venous obstruction after repair are 5-10% [3,10].

Conclusion

Infracardiac-type TAPVC is a rare malformation with a high incidence of venous obstruction that can lead to critical

deterioration of the patient's clinical status and even death if immediate diagnosis and surgical correction are not achieved. Although this heart disease can be suspected in TTE, sometimes the lack of acoustic windows can limit the visualization of the pulmonary veins, especially the connection site of the vertical vein and the sites of obstruction. That is why TAT images should be used in a complementary way, since the short image acquisition time, the high resolution of anatomical details, and the multiplanar and 3D reconstruction capacity make it an essential imaging modality. Therefore, the radiologist must be familiar with the complex anatomy and morphology of this pathology, to provide a detailed description of the findings in the report, which will help plan an adequate surgical correction.

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