Original Research Article

Role of MDCT in evaluation of total anomalous pulmonary venous connection (TAPVC)

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Abstract

Background: Pulmonary venous abnormalities are frequent in paediatric population with congenital heart disease and are particularly common in patients with abnormalities of atrial and visceral situs such as heterotaxy. These findings are treatable if indentified earlier. Accurate nature and extent of abnormalities is important for decision making about treatment. Chest radiograph and Echocardiography are earlier techniques used in suspected cases, but due to their limitations, definitive diagnosis is not made. Computed tomography angiography is a non-invasive and sensitive choice for mapping the pulmonary veins without the need for invasive cardiac catheterization. Complementary imaging in the form of cardiac magnetic resonance (MR) imaging is frequently used now-a days. **Materials and Methods:** A retrospective study was done at a tertiary care hospital between May 2018 to December 2019 in which 10 patients who presented with recurrent LRTI, breathlessness and failure to thrive were referred to radiology department for evaluation using CT. Males:female ratio was 1:1 in this 10 patients. Age group of this study was 6 days to 16 years. The MDCT examination was done using Siemens SOMATOM Definition AS. Cardiac CT was performed on ultra fast scanner with 1 mm slice thickness and 0.75 mm collimation after intravenous administration of non-ionic contrast agent at the rate of 4.5cc/sec with trigger of 180 HU in descending aorta. Retrospective 3D reconstruction of data was performed to obtain volume rendered images. **Results:** A retrospective study was done at a tertiary care hospital in which 10 patients within age group of 6 days to 16 years were studied. Mostly presented symptoms were recurrent LRTI, breathlessness and failure to thrive . In the early neonatal period, affected infants developed cyanosis and congestive heart failure. Males: female ratio was 1:1 in this 10 patients. Age group of this study was 6 days to 16 years. Out of 10 cases studied, 5 cases (50 %) were found to be supracardiac, 3 cases (30 %) were cardiac and 2 cases were infracardiac . In all cases, complete drainage of pulmonary venous blood is directed to the right heart. On chest radiograph correlation, most common finding was prominent right heart. It is because of volume overload. As required blood does not reach to the left atrium, it appears normal or hypoplastic in size. **Conclusion:** Using MDCT, proper pulmonary venous anatomy and abnormalities can be demonstrated, which helps for earlier treatment.
Keywords: cardiac CT, congenital heart disease, TAPVC, radiology.

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INTRODUCTION

Part of Foregut give rise to lung buds which have systemic arterial supply and venous drainage to the cardinal system. The common pulmonary vein (CPV) develops from the primitive left atrium. This CPV grows toward the lungs and establishes connections with the pulmonary venous system. With time, this primitive lung connection with cardinal venous system regresses, and pulmonary veins starts draining into the left atrium. When left atrium undergoes expansion over time, both CPVs and four connecting pulmonary vessels incorporates in it. Failure of this CPV to connect to the left atrium, with persistence of the primitive splanchnic connections of the pulmonary veins to the cardinal systemic veins and thence to the right atrium gives rise to TAPVC. Usually, this is isolated entity but can be seen as a component of other complex heart disease such as heterotaxy[4,5]. According to anomalous venous drainage, four types are seen.[6,7,8] Supracardiac,

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cardiac, infracardiac, and mixed. Chest radiography and Echocardiography as the first and safest imaging modality for cardiovascular abnormalities. But they may fail in complete depiction of some complex feature of TAPVC. Contrast cardiac MDCT, has ability to provide good anatomical and abnormality demonstration.

MATERIALS AND METHODS

A retrospective study was done at a tertiary care hospital between May 2018 to December 2019 in which 10 patients who presented with recurrent LRTI, breathlessness and failure to thrive were referred to radiology department for evaluation using CT. Males:female ratio was 1:1 in this 10 patients. Age group of this study was 6 days to 16 years. The MDCT examination was done using Siemens SOMATOM Definition AS. Cardiac CT was performed on ultra fast scanner with 1 mm slice thickness and 0.75 mm collimation after intravenous administration of non-ionic contrast agent at the rate of 4.5cc/sec with trigger of 180 HU in descending aorta. Retrospective 3D reconstruction of data was performed to obtain volume rendered images.

RESULTS AND ANALYSIS

A retrospective study was done at a tertiary care hospital in which 10 patients within age group of 6 days to 16 years were studied. Mostly presented symptoms were recurrent LRTI, breathlessness and failure to thrive . In the early neonatal period, affected infants developed cyanosis and congestive heart failure. Males: female ratio was 1:1 in this 10 patients. Age group of this study was 6 days to 16 years. Out of 10 cases studied, 5 cases (50 %) were found to be supracardiac, 3 cases (30%) were cardiac and 2 cases were infracardiac . In all cases, complete drainage of pulmonary venous blood is directed to the right heart. On chest radiograph correlation, most common finding was prominent right heart. It is because of increased volume. However, required blood does not reach to the left atrium, it appears normal or hypoplastic in size. Typical snowman appearance , also known as figure of 8 heart or cottage loaf heart 2-3 are seen in 1-2 classical cases of supracardiac type. On echocardiography which was available for few cases shown TAPVC with ASD or patent foramen ovale. Hypoplastic left atrium and dilated right atrium. After comparing available chest radiograph and echocardiography of patients, CT gave definitive abnormality type which helped clinician for further management of patient.

Most commonly seen cases were showing Supracardiac type followed by cardiac and infracardiac.

Rare type of mixed type was not found. MRI gives good tissue characterisation for accurate diagnosis. But due to cost issues and availability, MDCT was preferred.

DISCUSSION

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital anomaly of the pulmonary veins drainage[9]. In TAPVC , the pulmonary veins connect abnormally to the systemic venous circulation, instead of draining to left atrium. It is a cause of neonatal cyanosis . For survival, a right-to-left shunt is obligatory[10]. Depending upon the type and degree of pulmonary venous obstruction, it shows different presentations like 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[11]. Unobstructed type I TAPVC has very similar pathophysiology to a large ASD, so lesions not detected in infancy may be discovered later. Several cases of adult untreated TAPVC have been reported, even few cases were diagnosed after 60 years of age.

CLASSIFICATION:

It has four types-

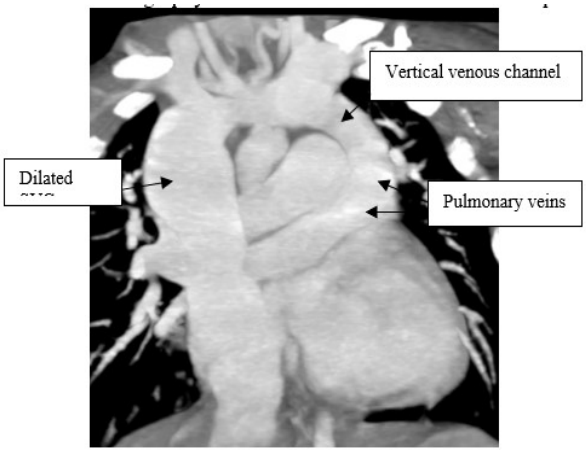
Type I: Supracardiac (most common)
Type II: Cardiac
Type III: Infracardiac
Type IV: Mixed.

In all 4 types, complete drainage of pulmonary venous blood is directed to the right heart. Here, we will go through few of the above cases.

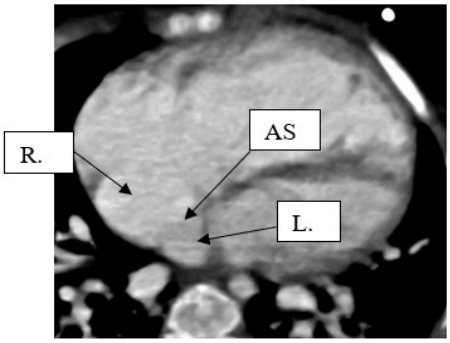
Case 1-

A 2 years old female child complaining of fever since 20 days.

2D echocardiography reveals - coarctation with anomalous pulmonary venous connection and large ASD.



All four pulmonary veins are draining into a common vertical venous channel which is situated in the left anterior mediastinum, inferiorly extending in the retrocardiac region posterior to left atrium . It is communicating with the right sided SVC. No pulmonary veins are draining into left atrium.



Large ostium secundum type of atrial septal defect.
CT of this patient also revealed-

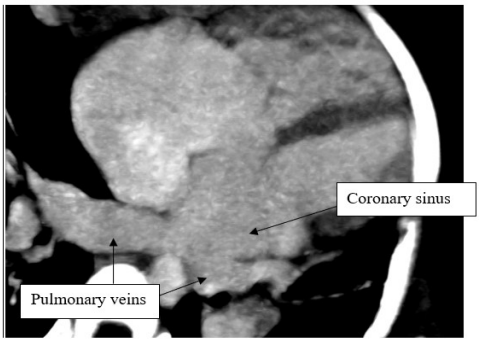
- Sub-aortic membranous ventricular septal defect.
- Cor triatriatum sinister.
- Double chambered right ventricle.
- Patent ductus arteriosus (Krichenko type E).
- Pulmonary arterial hypertension with dilated right heart chambers and right heart strain.

As seen above , supracardiac is the most common type of TAPVC. In this, the pulmonary veins drain in the confluence which is usually oriented horizontally. This confluence comes posterior to left atrium in location. An ascending vertical vein originates from this confluence(12,13). This vertical vein passes posterior to the left atrial appendages. Sometimes, it can get trapped between the dilated artery and the left bronchus, leading to pulmonary venous obstruction. The vertical vein finally drains into the innominate vein. The innominate vein and SVC are dilated. Dilatation of right atrium is noted due to volume overload. There has to be another way of blood flow to the left heart, it is always seen in the form of either an atrial septal defect or patent foramen ovale.

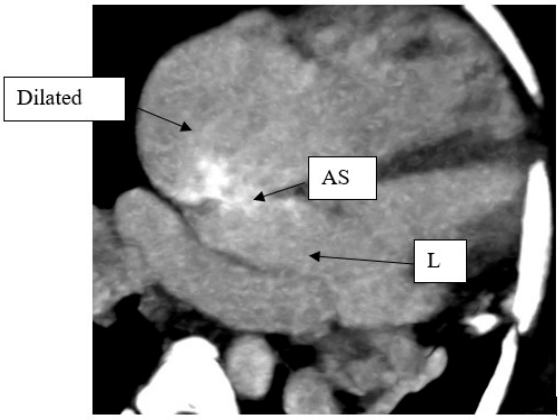
Case 2-

A 8 years old female child complaining of breathlessness with cyanosis and clubbing.

2D echocardiography suggestive of intracardiac TAPVC with all 4 pulmonary veins draining into coronary sinus, moderate sized ostium secondum ASD, dilated right atrium and ventricle.



Four pulmonary veins draining into the right atrium through the dilated coronary sinus.



A large ostium secundum ASD.
CT of this patient also revealed-

- Pulmonary arterial hypertension with bilateral pulmonary plethora.

Second most common type of TAPVC cardiac type, in this pulmonary venous confluence connects directly to the right atrium, usually through the coronary sinus. Significant dilatation is seen in the pulmonary veins, coronary sinus and right atrium. Obstruction is rarely noted in this type[14]. Pulmonary venous confluence drains to systemic veins below diaphragm in the infracardiac type of TAPVC. Usually the confluence is vertically oriented and runs posterior to the left atrium. Most commonly, infradiaphragmatic connection is\seen with the portal vein (at the confluence of the splenic and superior mesenteric veins) . Other venous connection is rarely seen[15]. It is the most common type to show the obstruction. Therefore, these neonates presents with severe respiratory distress.

CONCLUSION

TAVPR is an uncommon congenital cardiac anomaly. Contrast-enhanced MDCT and MRA are useful in the diagnosis as they provide accurate anatomic information used for presurgical planning. As CT is less time consuming, easily available, it became useful in critically ill neonates and infants with TAPVR. As surgery is the

mainstay of treatment, improvements in diagnostic imaging resulted in accurate depiction of this complex entity and advanced surgical techniques decreased the postoperative mortality rates.[16]

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