ROLE OF COMPUTED TOMOGRAPHY (CT) ANGIOGRAPHY IN DETECTION OF AORTOPULMONARY WINDOW (APW): REPORT OF A RARE ENTITY

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ABSTRACT

A 3 months old female child referred for scans at the department of radiology with suspicion of a congenital heart disease. A CT angiography was performed, which revealed an abnormal communication between the proximal aorta and the pulmonary trunk in the presence of separate aortic and pulmonary valves representing aortopulmonary window. This case shows the importance of CT angiography in the diagnosis and classification of aortopulmonary window as it gives a complete overview of the anatomy which can subsequently make planning for surgery easier.
Keywords: Congenital, Computed Tomography (CT) angiography, Aortopulmonary window, Radiology

Introduction

Aortopulmonary window is an abnormal communication of the pulmonary trunk with the ascending aorta with separate pulmonary and aortic valves.[1] The failure of fusion between the embryonic conotruncal ridges leads to the defect between the right and left circulation. The window begins above the semilunar valves with extension distally to the ascending aorta and the main pulmonary artery. It is a rare anomaly representing 0.2% to 0.6% of all the congenital heart diseases.[2]

Case Report

We report the case of a 3 months old female child referred for scans at the department of radiology. The patient had presented with increasing shortness of breath, poor feeding and irritability with a strong suspicion of a congenital heart disease. A CT angiography was performed with multiple axial sections from the aortic arch to the iliac crests after administration of IV contrast with early arterial phase. In addition, reformatted sagittal and coronal sections were obtained.

It revealed a situs solitus with levocardia; however, there was an abnormal communication between the proximal aorta and the pulmonary trunk in the presence of separate aortic and pulmonary valves representing aortopulmonary window as shown in (Fig. 1, 2 and 3). There were no signs of other congenital heart defects and the rest of the examination was within normal limits.

Discussion

The first to describe aortopulmonary window as a communication of the pulmonary trunk with the ascending aorta with separate pulmonary and aortic valves was Elliotson in 1830 while the first successful surgical

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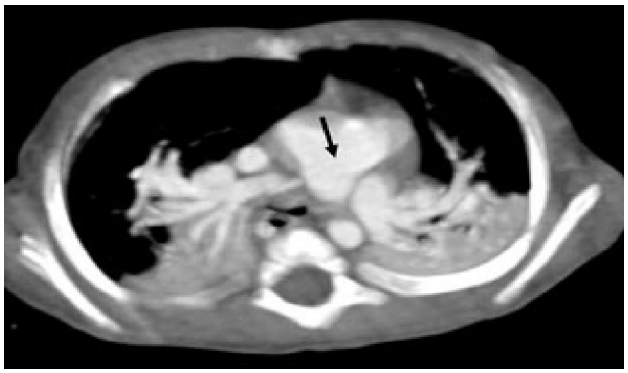

Figure 1: Computed Tomography (CT) axial section showing an abnormal communication between the proximal aorta and the pulmonary trunk

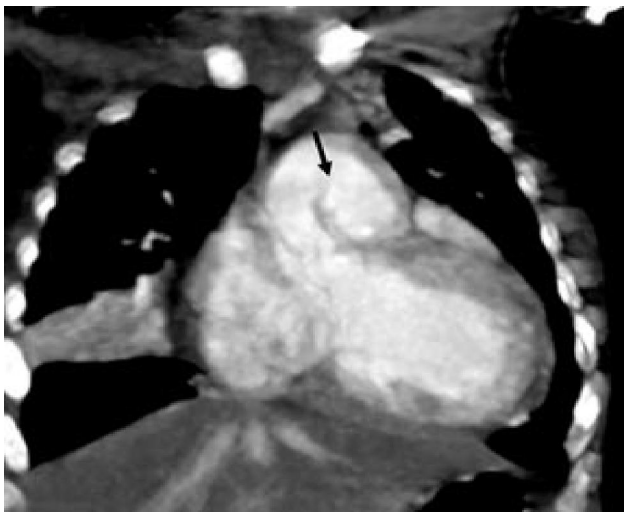

Figure 2: Computed Tomography (CT) coronal section showing an abnormal communication between the proximal aorta and the pulmonary trunk

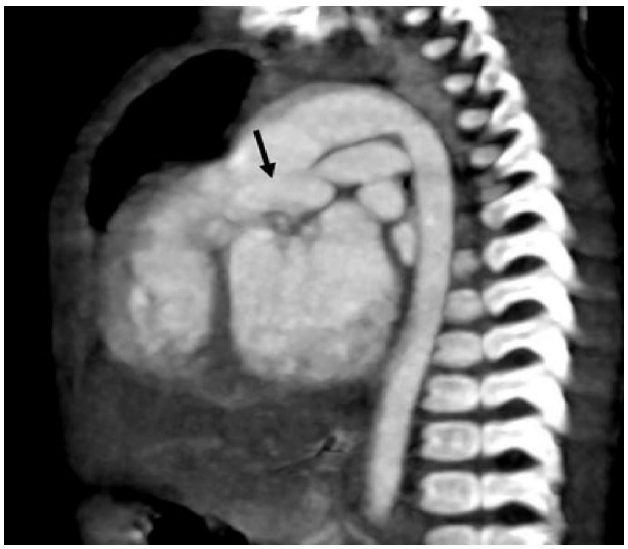

Figure 3: Computed Tomography (CT) sagittal section showing an abnormal communication between the proximal aorta and the pulmonary trunk

repair was done at Boston's Children Hospital in 1952.[1]

It is a rare anomaly representing 0.2% to 0.6% of all the congenital heart diseases.[2] It may occur by itself in 50% of the cases or be associated with simple repair anomalies (patent ductus arteriosis, atrial septal defect, patent foramen ovale) or larger complex anomalies (tetralogy of Fallot, pulmonary atresia, aortic arch interruption, aortic arc hypoplasia, coarctation of aorta, anomalous origin of coronary arteries).[3]

The failure of fusion between the embryonic conotruncal ridges leads to an aortopulmonary window. The defect starts just above the sinus of Valsalva and goes distally into the arch and can be of different sizes.[3] According to the Mori classification, it can be divided into three groups depending on the segment of the aortopulmonary septum involved: types I, II and III.[4] Type I involves the proximal part of the aortopulmonary septum while type II involves the distal. On the other hand, type III involves the complete length of the pulmonary trunk, starting above the semilunar valves and going to the level of pulmonary bifurcation. Ho et al. recently added a fourth group to this classification after the advent of transcatheter devices which are used to close the defect.[5] This new group, type IV, is reserved for intermediate defects with adequate superior and inferior rims.

Pathologically, aortopulmonary window resembles truncus arteriosis but can be differentiated by the fact that aortopulmonary window has essentially normal aortic and pulmonary valves.[1]

Clinically it is a left-to-right shunt that worsens during the newborn period as the pulmonary vascular resistance falls. This can be followed by a low cardiac output and congestive heart failure with an early onset of Eisenmenger's syndrome because of combined systolic and diastolic run-off into the pulmonary circulation.[6] The condition is thus fatal in most cases unless treated in infancy or early childhood.[7]

In addition to a clinical suspicion of congenital heart disease and ECG findings of biventricular hypertrophy, echocardiography helps in diagnosis by showing a "T-sign" with normal semilunar valves. Echocardiography also shows diastolic flow reversal in the aortic arch and downward flow in the main pulmonary artery.[6] The Angio-Computed Tomography scan is utilized to get additional anatomic details. It shows

the precise location of the defect along with any associated congenital heart defects. In our case, this was used and a defect was found in the proximal part of the aortopulmonary septum thus falling into the Mori classification of type I.

Conclusion

This case shows how important CT angiography is in the diagnosis of rare congenital disease such as aortopulmonary window as it gives a complete overview of the anatomy which can subsequently make planning for surgery easier.

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