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Aortopulmonary Septal Defect

A rare congenital cardiac anomaly

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We report a case of a 12-years-old girl child who presented with palpitation. Patient was able to do her regular activities, attending school. However, she used to become dyspnoeic on exertion.

The patient was referred to us for CT angiography. CT angiography was done in a 256 CT scanner (SIEMENS), that showed cardiomegaly with dilated left atrium and left ventricle. A defect of size 28 mm was noted between the ascending aorta and the pulmonary artery connecting the two vessels, about 16 mm cranial to the aortic root (figure 1). Pulmonary artery and its branches were also dilated. Main pulmonary artery measured 29 mm, its right branch 22mm and left branch 19mm. Ascending aorta was dilated near the defect. Aorta was arising from the left ventricle and pulmonary artery from right ventricle. Pulmonary veins were showing normal drainage into the left atrium. The branches of pulmonary artery were appearing prominent near the hilum. Lung window was showing mosaic attenuation in both lung fields. Diagnosis of aortopulmonary septal defect (APSD) with pulmonary hypertension was made.

Informed consent was obtained from the patient and from her father to use his medical data for publication.

Discussion

APSD also known as aortopulmonary window is one of the rare congenital cardiac abnormalities seen in 0.5% of all congenital heart defect.[1] It develops due to embryological incomplete separation of the common arterial trunk. This results in an abnormal connection between ascending aorta and main pulmonary artery.

APSD are classified into three types based on their location. Type I is the most common where the defect lies between the posteromedial wall of the ascending aorta and the main pulmonary artery just cephalad to the sinus of Valsalva. Type II defects occur in the distal part of the aortopulmonary septum adjacent to the right pulmonary artery. Type III defects are total defects involving the entire length of the aortopulmonary septum.[2,3] The aortic valve and right ventricular outflow tract remain normal. Truncus arteriosus may be confused with APSD, however truncus arteriosus has a truncal valve instead of two semilunar valves in APSD.

APSD can occur in isolation or in up to 50% of the cases are associated with other congenital heart defects such as interrupted aortic arch, transposition of great vessels, coarctation of aorta, tetralogy of Fallot, ventricular septal defects, atrial septal defect, and tricuspid atresia.[4] Variation of pulmonary arteries, and coronary arteries have also been described in association with this defect. There was no other associated abnormality in our case.

Chest x-ray shows cardiomegaly and increased pulmonary vascular markings as a result of left to right shunt. 2D echocardiography may demonstrate the defect, aortic root and chamber enlargements. CT angiography can accurately delineate the location of defect, its size, distance from aortic root, size of cardiac chambers, other associated anomalies, status of branches of pulmonary arteries and the lung fields.

Treatment of an AP window depends on its size where small defects can be closed by suture closure while larger defects may require a surgical patch closure. Catheterization can be considered when a defect is small enough to allow for device closure without causing stenosis of the great arteries or interference with the semilunar valves. Medical management includes anti-congestive medications such as diuretics and digoxin that can provide symptomatic relief without alteration of the disease course.

Authors' Contribution

SN did the concept, literature search, data acquisition, manuscript preparation, manuscript editing and manuscript review. SVR, SM and TT were involved in the literature search, manuscript editing and manuscript review. All authors approved the final version of the manuscript.

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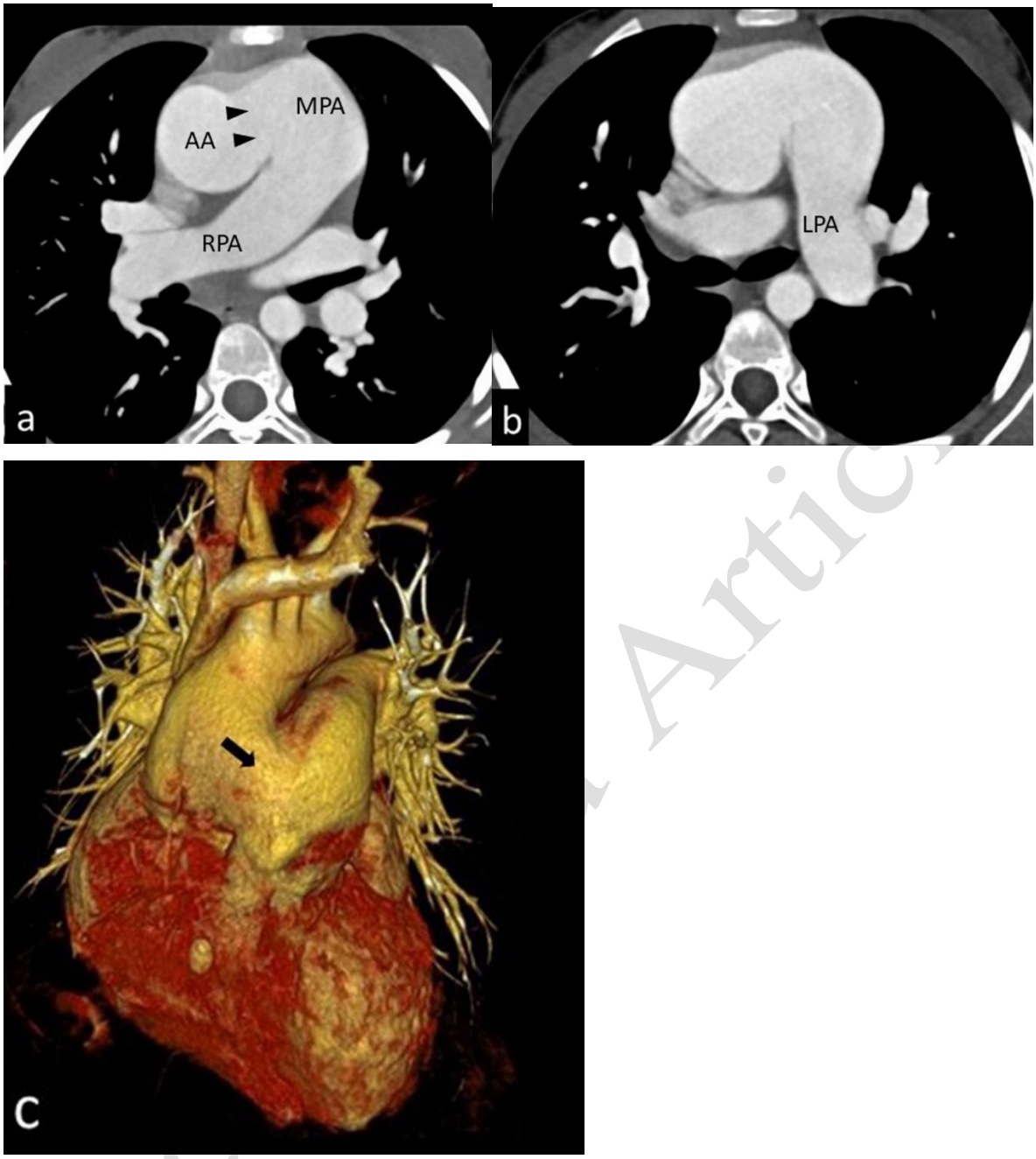

Figure 1: Axial sections of CT angiography at the level of ascending aorta **(A,B)** shows a large defect (arrowheads) between the ascending aorta and the proximal main pulmonary artery. There is dilated main pulmonary artery, its right (RPA) and left branch (LPA). Volume rendered image **(C)** shows communication between the ascending aorta and proximal main pulmonary artery (arrow) about 16 mm from aortic root. Left subclavian, left common carotid and innominate artery are arising from the arch of aorta.

MPA = main pulmonary artery, RPA = right pulmonary artery, LPA = left pulmonary artery.