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Case Report

A rare case of pulmonary atresia with ventricular septal defect with right-sided aortic arch ☆

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ABSTRACT

Pulmonary atresia with ventricular septal defect is a congenital heart malformation in which a lack of continuity between ventricles and the pulmonary artery is accompanied by ventricular septal defect. We report a 3-year-old female patient, who presented with a history of shortness of breath and bluish lips. Computed tomography pulmonary angiography showed an absent main pulmonary artery, right pulmonary artery, left pulmonary artery and showed a defect that connects the right ventricle and left ventricle, which concluded a VSD (ventricular septal defect).

Introduction

Pulmonary atresia is a congenital cardiovascular anomaly in which there is a complete disruption of the right ventricular outflow tract (RVOT) and the pulmonary trunk [1]. Pulmonary atresia is estimated to occur in 1 in every 10,000 live births. Pulmonary atresia with ventricular septal defect (PA-VSD) is a group of malformations characterized by a lack of continuity between the right ventricle and the pulmonary artery and associated with a VSD, with an incidence of 4-10 per 100,000 live births. In these cases, right ventricular hypertrophy usually occurs due to increased pressure from obstructed RV outflow. There is a genetic factor contribution, with increasing prevalence among those with Tetralogy of Fallot (TOF). Infants with PA-VSD usually present with cyanosis and hypoxia or heart failure [2,3].

In pulmonary atresia, the main pulmonary arteries and their branches are usually atretic or hypoplastic. Pulmonary atresia causes no blood supply to the lungs from the right ventricle, so it requires an extracardiac source of pulmonary blood supply, namely the formation of the main aortopulmonary collateral artery (MAPCA). The MAPCA is believed to be a persistent fetal primitive intersegmental artery originating from the descending aorta. They connect to the original pulmonary artery at the lobar or subsegmental level to provide pulmonary blood flow. Unlike typical bronchial arteries, they have elastic walls like those of the pulmonary artery and the aorta, provide no mediastinal or spinal cord branches, do not connect to the intercostal arteries, and anastomose with the pulmonary arteries near the hilum, not at the periphery of the lung. Here, we present a rare case of pulmonary atresia with a ventricular septal defect with a right-sided aortic arch as well as the role of imaging modality in the diagnosis.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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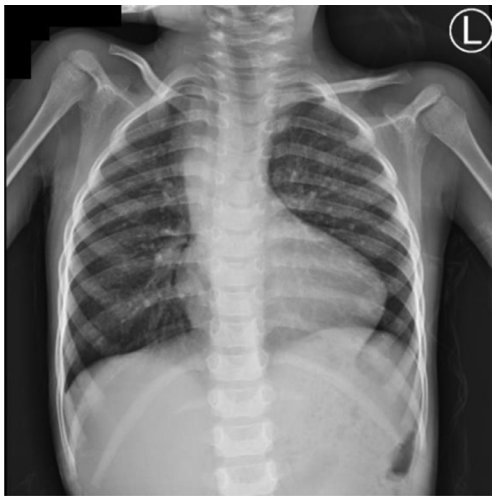

Fig. 1 – Chest radiograph revealed cardiomegaly, right side aortic arch, and absent pulmonary conus.

Case presentation

A 3-year-old female patient presented to us with a history of shortness of breath which presented intermittently, especially when the child has physically active. Complaints were accompanied by bluish lips. Her chest X-ray examination showed an enlarged heart with an aortic arch on the right side and the absence of pulmonary cones (Fig. 1). On computed tomography pulmonary angiography (CTPA), there was a defect that connects the right ventricle and left ventricle which concluded a VSD (ventricular septal defect) (Fig. 3).

Computed tomography pulmonary angiography (CTPA) also revealed absent main pulmonary artery, right pulmonary artery, and left pulmonary artery. The MAPCAs were seen to arise from the descending aorta which supplies the right and left lung lobes. The absence of a native pulmonary artery with MAPCA supplying the lungs directly indicates type C PA-VSD. In addition, there is a right-sided aortic arch with dilatation of the ascending aorta, the aortic arch, and the descending thoracic aorta Figure 2.

Discussion

PA-VSD is recognized as a group of congenital cardiac malformations in which there is a lack of continuity between the ventricle and the pulmonary arteries associated with ventricular septal defect. These patients may have atresia of part or all of the central pulmonary arteries. The blood supply to the lungs originates from extracardiac sources, most commonly from a patent ductus arteriosus (PDA) or from major aortopulmonary collateral arteries (MAPCAs), which connect the systemic circulation with the pulmonary arteries [1,2] Figs. 4 and 5.

This anomaly has also been referred to as “pseudotruncus arteriosus,” and “truncus arteriosus type 4.” In a subset of tetralogy of Fallot (TOF) with marked infundibular stenosis, there may be complete obstruction of pulmonary blood flow. This subset has been described as TOF with pulmonary atresia and forms a specific type of PA-VSD. Total disconnection of the right ventricle from the pulmonary arteries and the obligatory presence of extracardiac sources of pulmonary arterial blood flow differentiate PA-VSD from the TOF [1,2].

The pulmonary circulation in PA-VSD is extremely variable and depends on the presence or absence of native pulmonary arteries, PDA, and MAPCAs. The specific intracardiac defect in PA-VSD may be TOF, double-outlet right ventricle, or transposition of the great arteries [1,2].

On the basis of the characterization of pulmonary circulation, PA-VSD is classified into 3 types. In type A, the native pulmonary arteries are present and are supplied by the PDA. In type B, pulmonary blood flow is provided by both native pulmonary arteries and by MAPCAs. In type C, native pulmonary

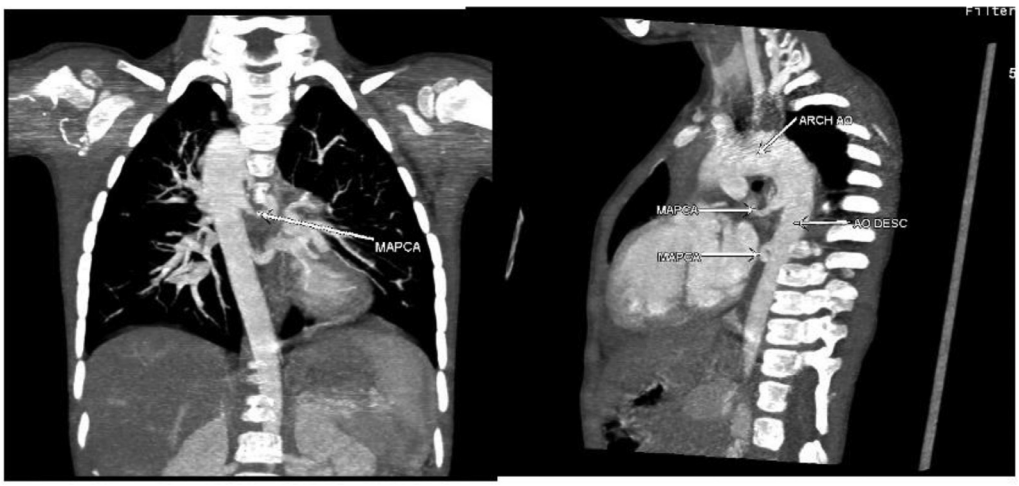

Fig. 2 – Computed tomography pulmonary angiography (CTPA), coronal and sagittal sections: MIP (maximum intensity projection) images reveal a right sided aortic arch with the arterial tract emerging from the left lateral aspect of the descending thoracic aorta which is a MAPCAs.

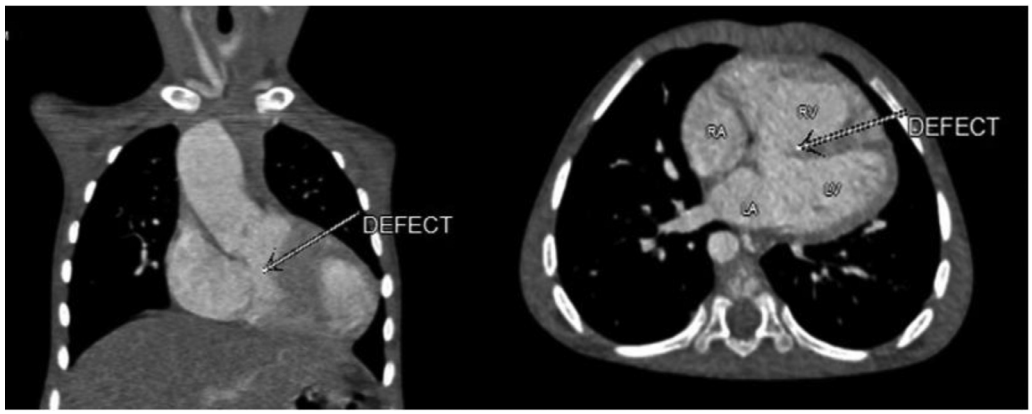

Fig. 3 – Computed tomography pulmonary angiography (CTPA), coronal and axial sections reveal a defect that connects the right ventricle and left ventricle which is a VSD (ventricular septal defect).

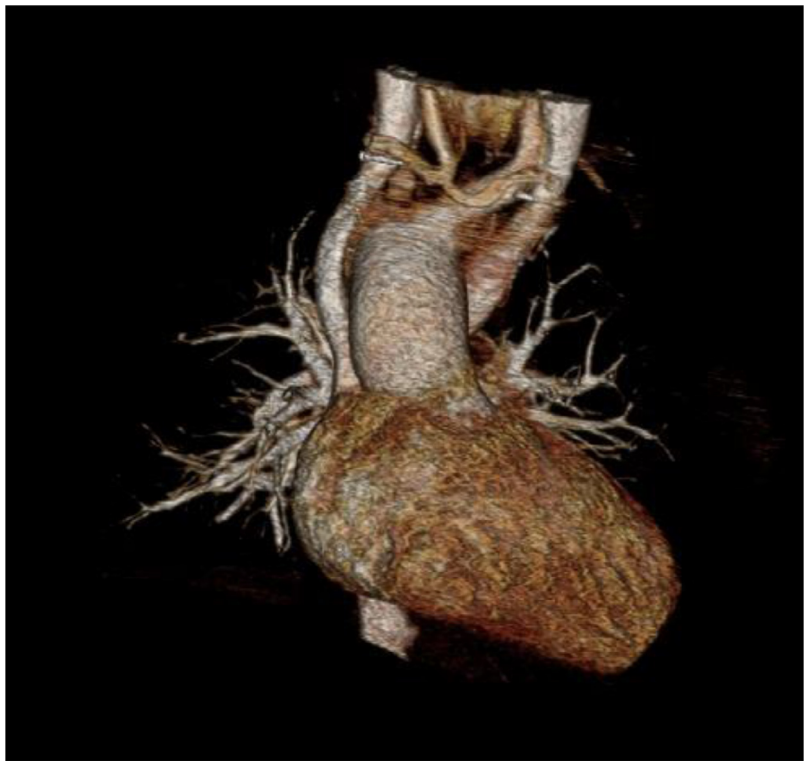

Fig. 4 – 3D Volume rendered image in frontal projections showing absent main pulmonary artery, right pulmonary artery, and left pulmonary artery.

arteries are absent and the blood supply is only through MAPCAs. MAPCAs are persistent tortuous fetal arteries that arise from the descending aorta and supply blood to pulmonary arteries in the lungs usually at the posterior aspect of the hilum [2,3].

In this case, chest X-ray examination, show cardiomegaly with the aortic arch on the right side and CTPA showed the pulmonary artery supply comes from several MAPCAs originating from the descending aorta which then divides to supply all pulmonary segments on both sides, but the blood supply from the MAPCAs is not sufficient to maintain oxygenation and is characterized by complaints of cyanosis. Thus a surgical approach was planned for this patient which consisted of uni focalization, which is a fusion of the MAPCAs followed by continuity of the right ventricle to the pulmonary artery and closure of the VSD [4,5].

CTPA allows direct visualization and accurate characterization of the branch pulmonary artery anatomy and depic-

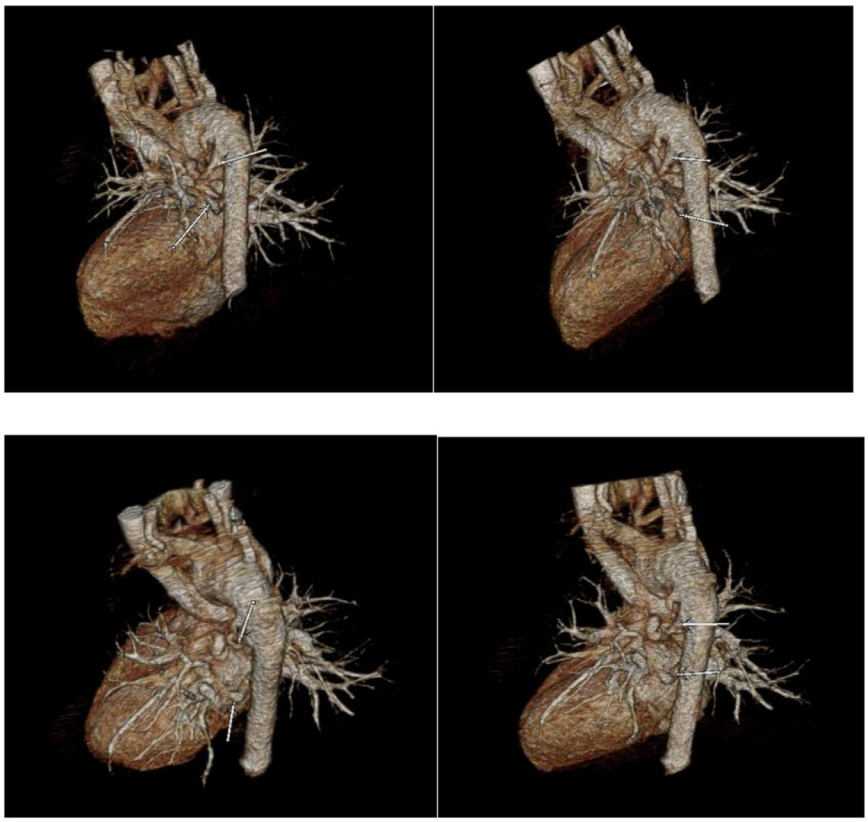

Fig. 5 – 3D volume rendered images showing an arterial channel emerging from the left lateral aspect of the descending thoracic aorta representing a MAPCAs.

tion of the pulmonary blood supply, which is necessary to determine the surgical approach describing the length of pulmonary atresia, presence of pulmonary artery branch junctions, size of the main, right and left pulmonary artery at the origin and hilum, and presence of pulmonary artery branch stenosis [4,5].

Conclusion

PA-VSD is a rare and complex congenital heart condition and requires the role of imaging modality for diagnosis and surgical planning.

Patient consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

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