

Case Report



Pulmonary Atresia with Anomalous Origin of the Pulmonary Artery

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Abstract

Pulmonary atresia (PA) with ventricular septal defect is a severe form of congenital cyanotic heart disease (TOF) but its not very common form. Here main pulmonary artery does not arises from common arterial trunk. In pulmonary atresia there is no proper formation of the pulmonary valve which allows blood from the heart to the lung of patient. Instead of opening and closing of the valve, a solid sheet of tissue develops. Therefore, blood cannot flow by its normal path to take oxygen from the lung. Instead, inadequate blood travels to the lung through other natural routes within the heart and its arteries and presenting degree of pulmonary atresia or discontinuity in arteries. MDCT gave structural details in this regard. So, role of MDCT play an important role to evaluate the details important information needed in preoperative evaluation for surgical policy in atretic patients. Here we discuss a case of pulmonary atresia (PA) with anomalous origin of the pulmonary artery.

Keywords: Pulmonary Atresia, Ventricular Septal Defect, MDCT.

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Introduction

A group of cardiac malformations known as pulmonary atresia with ventricular septal defect (PA VSD) are characterized by the absence of luminal continuity and blood flow between ventricle and pulmonary artery, with the right ventricle's (RV) blood egress occurring through the ventricular septal defect.¹⁻³ Pulmonary valve atresia with a VSD is a severe form of tetralogy of Fallot (TOF).⁴ Due to pulmonary valve atresia, the pulmonary vascular bed may receive blood from a number of places, including the patent ductus arteriosus (PDA) and aortopulmonary collateral arteries (APCAs).⁵ Anomalous origin of either pulmonary artery branch from the aorta has been reported. The association of pulmonary atresia (PA) with anomalous origin of the pulmonary artery, however, a very rare occurrence.^{4,7} In order to properly manage patients, it is crucial to completely define the global cardiovascular anatomy, including the native pulmonary artery, APCAs, PDA, and intracardiac malformation.^{8,9}

Case presentation

A 2-year-old girl was referred to our cardiology department with history of fever, several time of breathlessness and bluish

discolouration of finger for 5-6 months which exaggerated for last 4 days. Clinical examination revealed mild central cyanosis with oxygen saturation of 86% and digital clubbing were evident, respiratory rate 42 beats/min, pulse rate was 95-100 beats/min. All her pulses were equally palpable but bounding in character. Blood pressure was 140/60 mmHg. A loud heart murmur was present. Both lung fields were clear. Chest radiography revealed cardiomegaly (boot-shaped) with a cardiothoracic index (CTI) >60%, absent pulmonary bay with associated oligoemia and right sided aortic arch. Electrocardiography showed abscesce of pulmonary valve with atretic pulmonary artery, a mal-aligned VSD with 35% overriding of aorta and multiple MAPCA. Computed tomographic angiogram of pulmonary artery demonstrated a prominent overriding aortic outflow tract with a mal-aligned VSD, non-visualization of proximal part of pulmonary trunk, hypoplastic distal part of MPA communicating with distal part of ascending aorta which divided into right-left branches supplying lungs and thrombus in both pulmonary arteries. Right sided arch of aorta and descending thoracic aorta (Figure 1 to 4). There were right ventricular hypertrophy and three MAPCA's.

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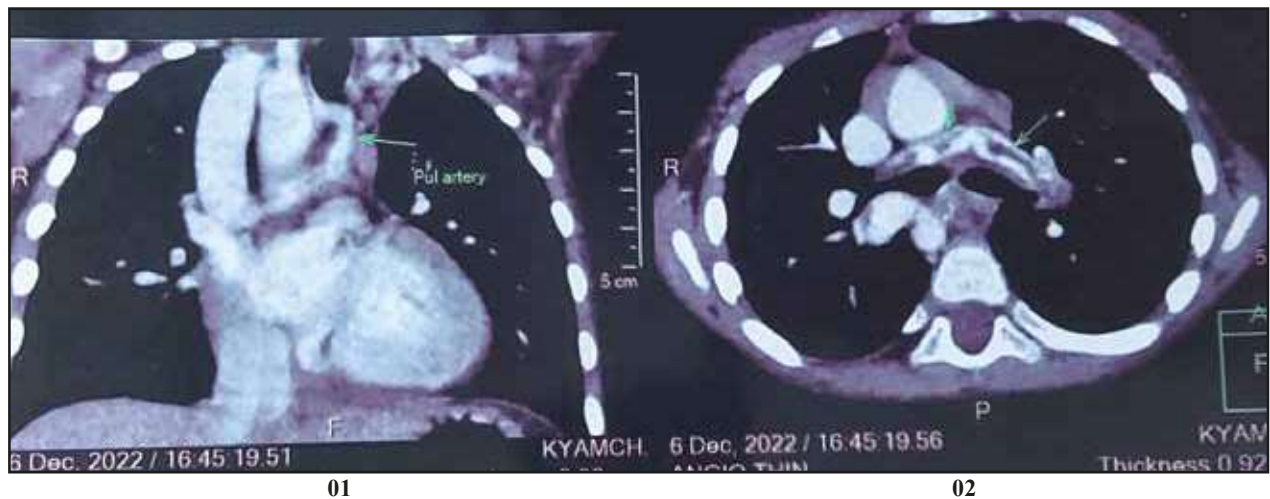


Figure 1 and 2: Coronal reformatted image of CT angiogram demonstrates the hypoplastic distal main pulmonary artery arising from distal part of ascending aorta (Fig 1) and presence of thrombi within right and left branches (Fig 2).

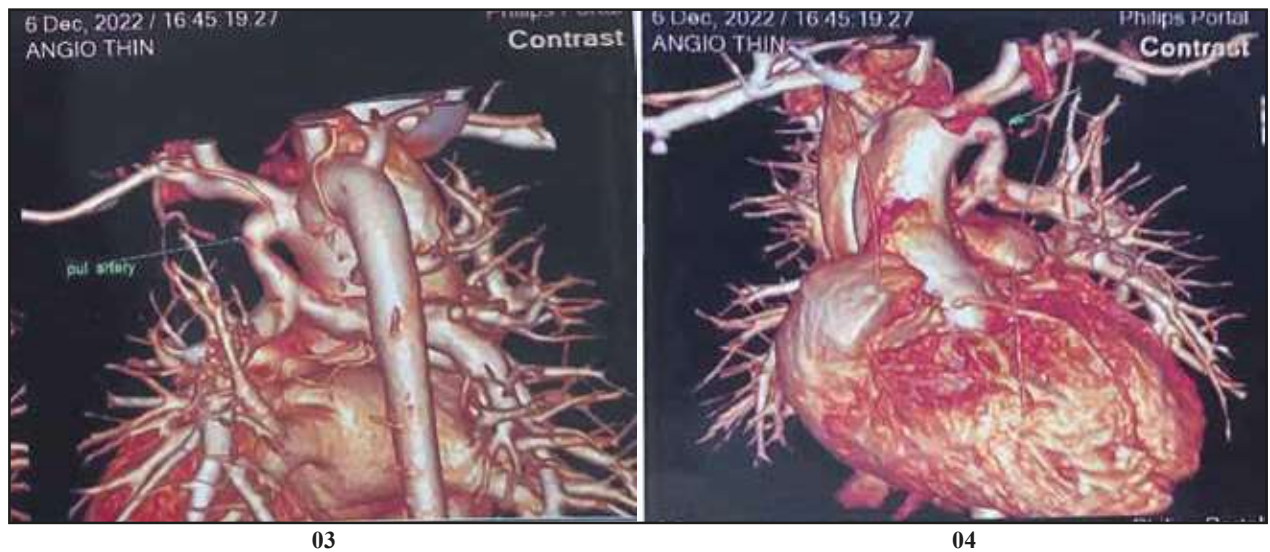


Figure 3 and 4: 3D reconstructive volumetric images demonstrate the hypoplastic distal main pulmonary artery, right and left branch.

Discussion

Tetralogy of fallot is the most common cyanotic heart condition along with pulmonary atresia, which aggravates the situation in such a way that a solid tissue sheet is developed instead of pulmonary valve, blood cannot pass through its natural path to the lung and different pathways form that carry oxygen-poor blood towards the lungs.^{1,2} The pulmonary arteries of patients with the typical form of TOF with pulmonary atresia are typically normal in size with normal peripheral pulmonary arborization. Additionally, compared to patients with PA-VSD, patients with TOF with pulmonary atresia have less established systemic-to-pulmonary collateral vessels.^{5,9} Our patient has a congenital cyanotic heart defect which is characterised by hypoplastic distal part of MPA arises from terminal part of ascending aorta and divided into right-left branches and supplying lung with thrombus in both pulmonary artery causing under-perfusion of the lungs. This patient presented with an

overriding of aorta with right sided aortic arch, descending thoracic aorta, VSD, pulmonary valve atresia, absent proximal main pulmonary artery (MPA). Collett and Edwards classified it first, According to the classification- in type I, aorta and main pulmonary artery share a common arterial trunk. Type II, right and left pulmonary arteries arise separately from the posterior part of truncus. Type III, separate origins of the pulmonary arteries from the lateral aspect of the truncus and at type IV, neither pulmonary arterial branch arising from the common trunk with the lungs supplied by collaterals (pseudotruncus). Van Praagh reclassified this as A1; aorta and main pulmonary artery share a common arterial trunk, in type A2 separate origins of the branch pulmonary arteries from the left and right lateral aspects of the common trunk; type A3, origin of one branch pulmonary artery (usually the right) from the common trunk, with other lung supplied either by collaterals or a pulmo

nary artery arising from the aortic arch and type A4, coexistence of an interrupted aortic arch. But our case did not match with any of the type of above mention classification. In our case hypoplastic distal MPA arises from distal part of ascending aorta just proximal to the origin of arch of aorta. Children with atratic and anomalous pulmonary artery are always at high risk of death if the disease is not treated, even after treatment, children are vulnerable to multiple heart problems.^{2,5} To resolve this, a proper technique for determining cardiopulmonary status and their correct management is vital, so it is important to provide pulmonary arterial measurement information for the surgical procedure of pulmonary atresia. Imaging purpose was to determine the extent and nature of pulmonary outflow reduction, to demonstrate associated anomalies and to present degree of pulmonary atresia or dis- continuity in the arteries.^{4,6,8} In this regard the MDCT gave structural descriptions.

Conclusion

MDCT is a reliable exploratory test for non-invasive assessment of global cardiovascular anatomy in patients with PA-VSD not only for preoperative assessment for surgical policy but also use full in postoperative further assessment.

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