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A Road from Coronary to Pulmonary: A Rare Imaging Presentation

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1. Abstract

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease. Its association with Pulmonary Atresia is considered to be the most severe form, the diagnosis of which plays an important role in determination of the treatment protocol. In such cases, systemic vascular channels called Major Aortopulmonary Collateral Arteries (MAPCA's) develop from aorta and its major branches to supply and maintain the pulmonary circulation. Such patients commonly undergo a Cardiac CT as an imperative pre-operative investigation for detailed information of these collaterals which helps plan further management. Here, we present a Case Report of an adult female patient with Pentalogy of Fallot wherein, a Cardiac CT showed the presence of dilated coronary-to-pulmonary collateral circulation. i.e. CAPA apart from the normally visualized MAPCA's, an extremely rare occurrence.

2. Introduction

Tetralogy of Fallot (TOF) with pulmonary atresia is a complex congenital heart disease with an incidence of 4.2-7 per 100,000 live births [1-3]. When TOF is associated with presence of an Atrial Septal Defect (ASD) or Patent Ductus Arteriosus (PDA), it is classified as a Pentalogy of Fallot.

In these patients, usually, Major Aorto-Pulmonary Collateral Arteries (MAPCA's) develop from descending thoracic aorta at the level of carina and abdominal aorta and its branches to help maintain the pulmonary blood flow and decrease the risk of pulmonary infarction. The presence of such collaterals is routinely detected on a Cardiac CT which forms the main pre-operative investigation in these patients. However, the presence of a collateral from the coronary artery to the pulmonary artery referred to as Coronary Artery–Pulmonary Artery Collateral (CAPA) or Coronary–Pulmonary Artery Fistula (CPAF) is an extremely rare occurrence. There is a dearth of literature regarding when to suspect and how to adequately demonstrate this anomaly.

Here, we present a case of Pentalogy of Fallot in an adult female patient with a CAPA arising from left main coronary artery to right main pulmonary artery and a PDA supplying the left main pulmonary artery with complete atresia of main pulmonary artery, this being the first such case in literature. This case also highlights the role of Cardiac CT in the diagnosis and management of this rare entity.

3. Case Report

A 21-year-old female patient was referred to our institute for evaluation and treatment of cyanotic congenital heart disease. She currently complained of long-standing effort intolerance, palpitations and dyspnoea (New York Heart Association Class III).

On examination, she was of average build with grade III clubbing and no cyanosis at present. Her pulse rate was 92/min with blood pressure of 120/76 mm Hg. The cardiovascular system examination revealed normal S1/S2 and a continuous murmur best heard in the right parasternal area over the third intercostal space. The baseline systemic arterial oxygen saturation (SaO2) was 87%. She had a past history of cyanotic spells at birth with multiple similar on and off episodes upto 6 months of age.

Her chest topograph showed mild cardiomegaly with signs of dilated right atrium, boot shaped heart, large aorta, and oligemic lung fields (Figure 1).

An evaluation with transthoracic echocardiography showed evidence of a large subaortic VSD and over-riding of aorta and pulmonary atresia. No collateral vessels or fistula could be appreciated on colour Doppler imaging. Mild tricuspid regurgitation with

severe pulmonary arterial hypertension was present.

A Cardiac CT was advised as a routine pre-operative investigation to evaluate the pulmonary vasculature and ascertain the presence of MAPCA's.

A retrospective ECG-gated Cardiac CT was performed on a 64 slice Multidetector Computed Tomography (MDCT) scanner (Brilliance Philips) in a helical mode followed by retrospective 2D and 3D reconstruction (MPR &MIP). The contrast images were obtained after injection of 90 ml of iodinated non-ionic contrast by bolus triggering. A venographic phase was also obtained including the upper abdomen to assess concomitant venous anomalies.

The Cardiac CT confirmed the echocardiographic findings of a large sub-aortic VSD with over-riding of aorta by approximately > 50% (Figure 2A and 2B) and complete atresia of pulmonary valve and main pulmonary artery (Figure 2C).

A small PDA was seen arising from inferior surface of arch of aorta and communicating with the left main pulmonary artery with severe junctional stenosis (Figure 3A, 3B and 3C).

Additionally, it was observed that the left main coronary artery (LMCA) was dilated with a large tortuous collateral (CAPA/CFAP) arising from it after the origin of LCx and LAD, further communicating with right main pulmonary artery, thus providing the major source of supply to the right pulmonary circulation (Figure 4A,4B,4C and 4D). The distal pulmonary arteries were moderately narrowed in calibre.

Apart from this, multiple dilated and tortuous collaterals (MAPCA's) were also noted in the mediastinum arising from the arch and descending aorta and coursing into bilateral lungs via lung hilum (Figure 5A). The largest of these collaterals (9.2 mm in diameter) was seen arising from the right lateral wall of the descending thoracic aorta at the D8 vertebral level to the right ascending pulmonary artery (Figure 5B).

The LMCA showed normal division into the Left Anterior Descending artery (LAD) and Left Circumflex artery (LCX) which had normal course and calibre. The Right Coronary Artery (RCA) was seen to arise from the right coronary cusp and showed normal calibre and course.

An evaluation of the cardiac chambers showed mild concentric hypertrophy of right ventricle with dilated right atrium.

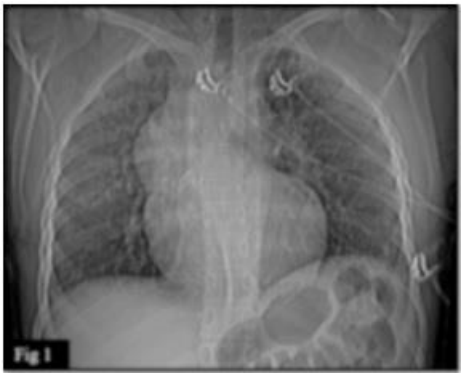

Figure 1: Frontal topograph demonstrates a boot-shaped heart with an uplifted apex secondary to RV hypertrophy and dilated ascending aorta causing bulge of right heart border above right atrium.

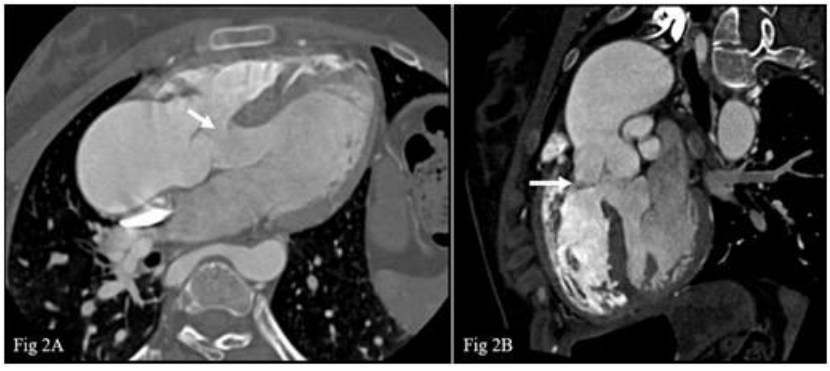

Figure 2A and 2B: Cardiac CT images showing a sub-aortic VSD with over-riding of aorta by approximately > 50% (white arrows).

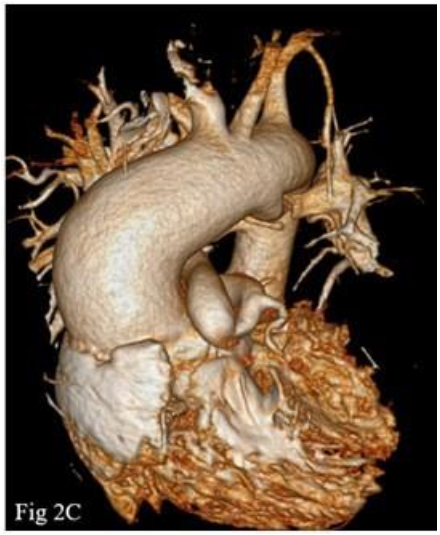

Figure 2C: Reconstructed VRT images showing complete pulmonary atresia.

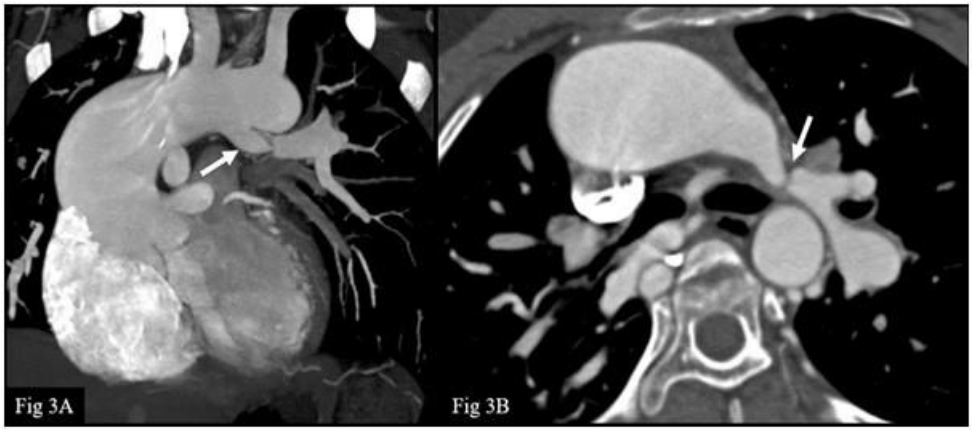

Figure 3: Reconstructed coronal MIP **(3A)** and axial images **(3B)** showing persistent PDA arising from arch of aorta communicating with LPA with severe junctional stenosis (white arrows).

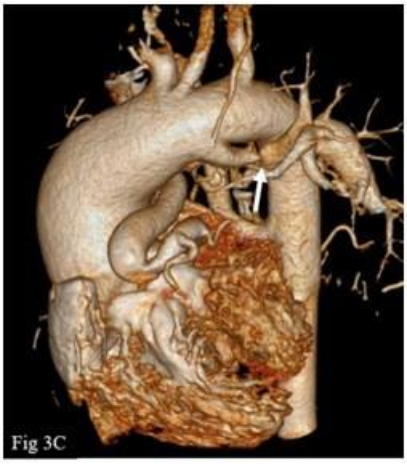

Figure 3C: Reconstructed VRT images showing the PDA communicating with LPA (white arrow).

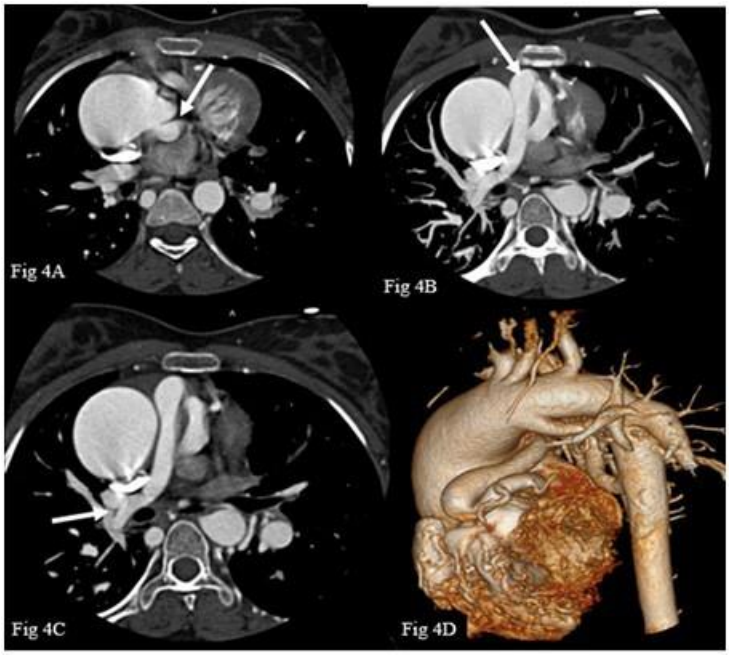

Figure 4: Sequential contrast axial images **(4A, 4B, 4C)** and reconstructed VRT **(4D)** images showing dilated LM with a large collateral communicating with RPA (white arrows). Reconstructed VRT **(4D)** images also shows LAD and LCx arising from LM (blue arrows).

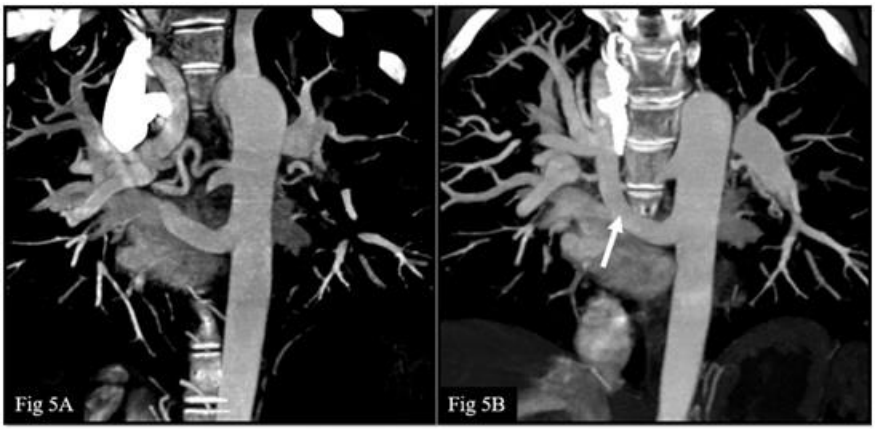

Figure 5A and 5B: Multiple dilated and tortuous MAPCA's with largest collateral (white arrow) arising from right lateral wall of aorta.

4. Discussion

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease with an estimated worldwide prevalence of 3 cases per 10,000 live births [4]. It is a conotruncal defect which occurs due to an anterior malalignment of the infundibular septum which is responsible for the four main components [5] (Figure 6).

a). Ventricular Septal Defect (VSD)

b). Pulmonary Infundibular Stenosis

c). Over-riding of aorta.

d). Right ventricular hypertrophy.

The association of TOF with an Atrial Septal Defect (ASD) or Patent Ductus Arteriosus (PDA) classifies it as a Pentalogy of Fallot, as was visualized in this case.

The clinical presentation of these patients as well as their prognosis predominantly depends on the degree of obstruction to pulmonary blood flow, whether hypoplastic, severely stenotic or completely atretic pulmonary valve and pulmonary arteries [6]. In such cases Multiple Abnormal Vascular Channels (MAPCA's) [7, 8] develop from the thoracic aorta, abdominal aorta, subclavian artery, bronchial and intercostal arteries which are connected to the pulmonary arterial vasculature distally, thereby maintaining the pulmonary blood flow and reducing the chances of pulmonary infarction.

However, in rare cases, coronary arteries, i.e. Right Coronary Artery (RCA), Left Main Coronary Artery (LMCA) or its branches Left Anterior Descending artery (LAD) and Left Circumflex artery (LCX) can be a source of pulmonary blood flow [9].

Amin et al. [10] hypothesized that a communication between the coronary and pulmonary artery is physiologically and embryologically more similar to a ductus arteriosus than to systemic–pulmonary collaterals as this communicates in an antegrade fashion with the central pulmonary arteries. Sometimes a coronary to pulmonary artery communication with sufficient calibre can become the primary source of pulmonary blood flow in patients with PA/VSD.

In this setting, whether such a communication should be referred to as a CAPA or fistula (CPAF) can be put up for debate. [11].

The estimated incidence of such Coronary Artery to Pulmonary Artery Collaterals (CAPA) ranges from 1.3% to 10% [12]. To date, 52 cases of PA/VSD with CPAP have been reported in literature. The CPAP involved the LMCA in 34 cases, RCA in 12 cases, a single coronary artery in 5 cases, and both coronary arteries in 2 cases [11]. Mittal and colleagues [13] reported a case of a large fistula between the left anterior descending artery and the pulmonary artery in a patient with TOF. Another report describes a collateral communication between the left circumflex artery and the right pulmonary artery [14].

However, to the best of our knowledge, this is the first case with presence of multiple sources of pulmonary blood flow, i.e. LMCA to right pulmonary artery, PDA to left pulmonary artery and multiple dilated and tortuous MAPCAs from descending aorta and bilateral bronchial arteries.

The demonstration of such anomalous collateral pathways remains difficult and requires an invasive investigation such as Coronary Artery Angiography for their adequate detection. However, knowledge of existence of these variations can aid in their prompt detection on a simple non-invasive Cardiac CT as well, as was in our case. This can obviate the need for diagnostic invasive procedures, directly guiding the cardiac surgeons to plan surgical treatment of choice accordingly.

Determining the presence of collaterals has implications in management of such patients as well. Ever since the introduction of the concept of early one staged unifocalization in the surgical management of patients with PA/VSD, accurate delineation of all the sources of pulmonary blood flow has become the most critical step of the preoperative evaluation [15]. The main aim of treatment strategy is to recruit as many lung segments as possible into the pulmonary circulation. This case demonstrates the interdependence of size of CAPA and the number of MAPCA's supplying the pulmonary bed. The CAPA/CPAF is addressed surgically depending on the distal artery with which it connects. If the distal end of the CAPA is connected to the MPA, it is ligated and divided. If the distal end is connected to the right or left pulmonary arteries (as in our case) it is unifocalized.

The findings of Cardiac CT in our case thus helped guide the cardiac surgeon in planning a multistage treatment procedure.

The detection of coronary artery abnormalities has clinical implications as well. When the coronary artery drains into the capillary networks of the lungs, 'coronary artery steal' phenomenon can result which could cause myocardial ischaemia. However, this condition may also develop pulmonary hypertension and heart failure due to the haemodynamic effect of the left-to-right shunt. Hence, detection of the presence of these coronary artery to pulmonary artery connections has both prognostic as well as diagnostic implications.

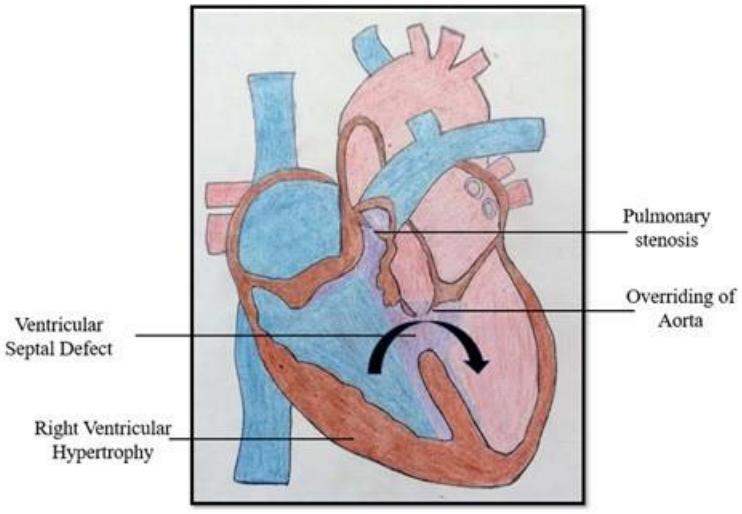

Figure 6: Schematic diagram showing components of Tetralogy of Fallot

5. Conclusion

Congenital cardiac diseases belong to a complex group of diseases and knowledge of their myriad of presentations has an important bearing on the overall physical and mental health of patients. With the advent of newer generation MDCT scanners, presence of various systemic–pulmonary as well as coronary-pulmonary collaterals can be delineated in a non-invasive manner with the added advantage of detection of extracardiac anomalies as well, thus obviating the need for invasive procedures such as catheter angiography.

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