Anesthetic and Intensive Care Management of Left Main Coronary Artery to Main Pulmonary Artery Fistula Diagnosed in Postoperative Case of Tetralogy of Fallot

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ABSTRACT

Cases of coronary to pulmonary artery fistula are seen in patients of pulmonary atresia with ventricular septal defect (VSD). These fistulas are rarely seen in patients of Tetralogy of Fallot (TOF). In this case report, we have presented ICU management of a postoperative case of TOF, with missed diagnosis of left main coronary artery (LMCA) to main pulmonary artery (MPA) fistula.

Keywords: Left main coronary artery, main pulmonary artery, Tetralogy of Fallot, ventricular septal defect

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Submitted: 02-May-2020 Revised: 21-Jun-2020 Accepted: 06-Aug-2020 Published: 19-Apr-2021

INTRODUCTION

Cases of coronary to pulmonary artery fistula are seen in patients of pulmonary atresia with ventricular septal defect (VSD).^[1] These fistulas are rarely seen in patients of Tetralogy of Fallot (TOF). In this case report, we have presented ICU management of a postoperative case of TOF, with missed diagnosis of left main coronary artery (LMCA) to main pulmonary artery (MPA) fistula.

CASE REPORT

A 6-year-old male child presented with history of bluish discoloration of lips and nails since birth and easy fatigability since two years. His clinical examination, chest X-ray and echocardiogram were consistent with diagnosis of TOF.

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	DOI: 10.4103/aca.ACA_87_20

CT Pulmonary Angiography (CTPA) done 4 years back suggest of a severely stenosed collateral from descending thoracic aorta (DTA) to pulmonary artery. Present echocardiography did not suggest of any collaterals. He underwent intracardiac repair (ICR) surgery and was shifted to Intensive Care Unit (ICU). In ICU on 2nd postoperative day (POD) as patient was stable, he was extubated and the drains were removed on 3rd POD. On 4th POD patient was having shortness of breath, arterial blood gas (ABG) analysis showed respiratory acidosis and chest X-ray showed right side pleural effusion. Patient was reintubated and right side drain was inserted, which had a collection of 300 ml serosanguneous discharge. Echocardiography was done, but the report was inconclusive. So CTPA was done to rule out any missed MAPCAs, we found LMCA to MPA

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How to cite this article: Panda S, Gandhi H, Surti J, Mishra A, Champaneri B. Anesthetic and intensive care management of left main coronary artery to main pulmonary artery fistula diagnosed in postoperative case of tetralogy of fallot. Ann Card Anaesth 2021;24:272-4.



Figure 1: (a) Left main coronary artery to Main pulmonary artery fistula and (b) Major aorto pulmonary collateral arteries from descending thoracic aorta supplying both lower lungs

fistula [Figure 1] and a MAPCA from descending thoracic aorta (DTA) supplying both lower lungs [Figure 1]. Patient was then shifted to cath lab, where coiling of MAPCAs was done [Figure 2] and device closure of fistula was done [Figure 2]. On 5th POD, vitals of the patient was stable and there was no further drain, thus patient was extubated and the drains were removed on 6th POD. On 7th POD patient was shifted to postoperative ward and discharge from hospital on 10th POD.

DISCUSSION

The congenital coronary artery to pulmonary artery fistula is rare anomaly with reported prevalence of 1 in 50,000.^[2] The cause of coronary arteriovenous fistula is due to persistence of sinusoidal connections between the lumens of primitive tubular heart that supply myocardial blood flow in early embryologic period.^[3] As there is reduced pulmonary blood flow, collaterals between systemic and pulmonary circulations develop over period of time to increase pulmonary arterial flow. Most of coronary fistula although remain asymptomatic, there can be complains such as myocardial ischemia due to steal phenomenon, thrombosis, embolism, rupture, endocarditis, and arrythemia.^[4]

Accurate delineation of the anatomy prior to surgery helps to plan management. In our case CTPA was done 4 years back, which suggested of severely stenosed collateral from DTA to pulmonary artery and no coronary to pulmonary fistula. Also preoperative echocardiography findings also missed it. Intraoperatively surgeons noticed return from pulmonary veins during on pump, but it was manageable with suction and left arterial vent, they thought it may be due to small collaterals which generally becomes atretic over period of time. Dilated LMCA was seen, but since it



- 2. Major Aortopulmonary Collateral (MAPCA) suppling both lower lungs
- 3. Coiling of MAPCA

Figure 2: (a) Coiling of Major aorto pulmonary collateral artery (b) Device closure of Left main coronary artery to Main pulmonary artery fistula

can be also normally found in cyanotic patients and echo report did not suggest of coronary to pulmonary fistulas they did not trace it further.

In the postoperative period, since the patient was reintubated due to dyspnoea and respiratory acidosis. Chest X-ray suggest of significant pleural effusion, so we suspect the cause may be due to significant MAPCAs. To rule out CTPA was done, which suggested of LMCA to MPA fistula and one MAPCA from DTA supplying both lower lungs. As surgical closure of the fistula and the MAPCA was high risk procedure and in our institute we routinely do coiling procedures,^[5] so we decided to shift patient to cath lab for further management after taking opinion of pediatric cardiologist and the operating surgeon. The above finding was then confirmed by angiography in cath lab, following which coiling of MAPCA and device closure of the coronary fistula was performed.

CONCLUSION

Preoperative identification of collaterals is important by various investigations such as cineangiography, CTPA or magnetic resonance angiography in patients of TOF, when echocardiography suggest severe infundibular stenosis, but patient is not so cyanosed especially in older children. In cases like us, a team approach of pediatric cardiologist, pediatric cardiac surgeon, cardiac anesthetist and pediatric intensivist is required for early diagnosis and successful management of the patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship Nil.

Conflicts of interest

There are no conflicts of interest.

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