Major Aorto Pulmonary Collateral Arteries (MAPCA) with Tetralogy of Fallot and Pulmonary Artesia in Middle Age Adult: a Rare Finding

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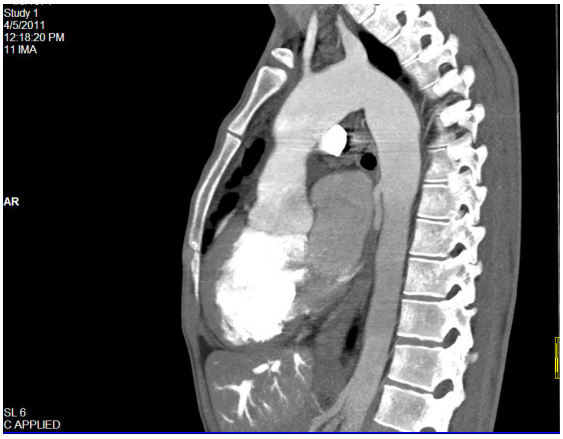

Figure 1. Sagittal MIP CT image of pulmonary angiography is showing origin of lower lobe aortopulmonary collateral from descending aorta.

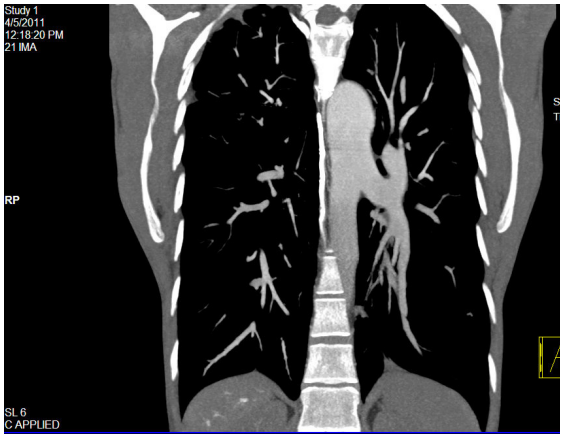

Figure 2. Coronal MIP CT is showing left pulmonary artery from descending aorta

A forty four years old male patient presented with headache and dizziness in our outpatient department. On examination, he had pulse rate of 96 /min, blood pressure of 122/80 mmHg, JVP was not raised. He had clubbing and cyanosis on General physical examination. On auscultation pansystolic murmur was heard all over the chest. His respiratory and abdominal examination was normal. There were no significant findings on electrocardiography and skiagram chest. Echocardiography revealed ventricular septal defect with overriding of aorta, pulmonary artery could not be visualised. On computrized tomography pulmonary angiography revealed Ventricular septal defect with overriding of aorta. The main and normally confluent right and left pulmonary arteries were not visualized. On left side there was large MAPCA arising from descending thoracic aorta at D8 level and dividing into upper and lower lobar arteries and segmental branches were seen. On right side there were three large MAPCAS seen arising from (i) proximal right subclavian artery (supplying the rt upper lobe), (ii) arch of aorta (at D5 level supplying right middle and lower lobes), (iii) descending thoracic aorta (at D7, D8 level supplying right lower lobe) respectively were seen. Inferior vena cava and pulmonary veins were normal. (**Figure 1 and 2**) Rest of examination was normal.

Another significant finding in other investigations were Polycythemia (Hb-18 gm%), PCV 60. In view of above findings he was diagnosed a case of Cyanotic congenital heart disease-tetralogy of fallot (TOF) with pulmonary atresia with MAPCAs with polycythemia (secondary to cyanotic congenital heart disease).

In view of symptomatic polycythemia in the form of headache and dizziness, he was managed with phelebotomies On and Off depending on his consent. He was also given conservative treatment as and when required for other symptomatology. He had marked improvement with treatment. He was further advised to have regular follow up and to maintain his medical records.

Tetralogy of fallot is a common form of cyanotic congenital heart disease found in the early phase of life. Among patients of TOF, pulmonary atresia occur only in 12% of cases.[1] In these cases native pulmonary artery may be of normal size but is hypoplastic. In remaining cases of hypoplastic pulmonary artery, pulmonary blood supply is from MAPCAs.[2] In our patient, CT pulmonary angiography showed absence of main, right and left pulmonary arteries, and pulmonary circulation was from MAPCAS only. In literature, there are four type of aortopulmonary circulation in patients of TOF with pulmonary atresia as described by Castenda[3] and colleagues as shown in **Table 1**. Our patient comes under type 4 as per this classification. Patients with large MAPCAS and unrestricted pulmonary blood flow are more prone to develop pulmonary arterial hypertension (PAH) and congestive cardiac failure (CCF). Survival of patients with TOF and pulmonary atresia depends upon adequacy of pulmonary blood flow.[4] In our patient, blood flow was adequate in both lung fields. Our patient had TOF with pulmonary atresia and pulmonary circulation was through MAPACAs only. So, there was less chance of developing PAH and CCF and that is why he did not develop the same till the age of forty four years. Survival in such cases has not being widely studied in literature. Survival is prolonged because they are diagnosed at early stage and surgery is done if needed.

Table 1. Four type of aortopulmonary circulation in patients of TOF with pulmonary atresia[3]

	Non-arteric MPA	PDA	RPA-LPA confluence	MAPCA
Type 1	+	+	+	-
Type 2	-	+	+	-
Type 3	-	-	+	+
Type 4	-	-	-	+

In one study, the patient who survived without surgery was 54 years old. The average survival is not beyond third decade. Our patient had survived till the age of 44 years and without PAH and CCF. The prolonged survival in such cases depend on adequate but not excessive aorto pulmonary circulation. Long term consequences of this pathology are chronic volume overload of left ventricle resulting in cardiac failure. This case was reported because of rarity of condition and the survival of patient beyond forth decade of life without surgery.

REFERENCES

1. Garne E, Nielsen G, Hansen O, Emmertsen K. Tetralogy of fallot. A population-based study of epidemiology, associated malformations and survival in western Denmark 1984-1992. Scand Cardiovasc J. 1999;33(1):45-8.
2. Asija R, Hanley FL, Roth SJ. Postoerative respiratory failure in children with tetralogy of fallot, pulmonary atresia, and major aortopulmonary collaterals: a pilot study. Pediatr Crit Care Med. 2013;14:384.
3. Rome JJ, Mayer JE, Castaneda AR, Lock JE. Tetralogy of fallot with pulmonary atresia. Rehabilitation of diminutive pulmonary arteries. Cardiol. 1993;88(4 Pt 1):1691-8.
4. Bertranou EG, Blackstone EH, Hazelrig JB, Turner ME, Kirklin JW. Life expectancy without surgery in tetralogy of fallot. Am J Cardiol. 1998;42(3):458-66.