Case reported is a 9 year old male child, with history of cyanosis and dyspnoea on exertion and effort intolerance since 9 months of age. There was no history of squatting to relieve cyanosis and dyspnoea. No history of cough, leg swelling, oliguria, syncopal attack, any convulsions or neurological deficits. Birth history was normal, full term, 2700 grams, with an Apgar of 9. He had no family history of congenital heart disease, arrhythmias or other cardiac diseases. General examination revealed an asthenic plethoric patient, centrally cyanosed with grade 4 digital clubbing. His pulse was regular with a rate of 82 beats per minute, respiratory rate of 28 beats per minute, regular. Blood pressure was 100/62mmhg. Saturation was 68% on room air. On examination of the precordium, the apex beat was visible on the right side in the intercostal space 1.5 cm medial to the right mid-clavicular line with precordial bulge. First and second heart sounds were present with no murmur. Lungs were clear. Abdomen was normal without any organomegaly.

Chest radiograph showed dextrocardia with a cardiothoracic ratio of 0.6 suggesting no cardiomegaly according to his age.

Ultrasonography of abdomen showed liver on the right and spleen on left side with no organomegaly. 2DEcho was suggestive of dextrocardia with tetrology of Fallot. As we did not find the complete details about the diagnosis and the presence and extent of MAPCAS a Cardiac CT Angiography was done. The findings changed the entire picture which showed situs solitus with dextrocardia with large ASD, Large VSD, Patent ductus arteriosus, corrected transposition of great arteries, pulmonary atresia with Multiple aortopulmonary collateral arteries (MAPCAS) originating from arch of aorta and descending aorta measuring 2mm to 4mm. Atriventricular discordance was noted.

The packed cell volume was 62% with hemoglobin of 21gm/dl suggesting polycythemia. The final diagnosis of Complex congenital heart disease with MAPCAS with dextrocardia was made.

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ABSTRACT
Tetrology of Fallot is the most common form of cyanotic congenital heart disease. It is a combination of 4 different defects viz ventricular septal defect, obstructed outflow of blood from right ventricle to the lungs (pulmonic stenosis), displaced and over riddded aorta, and right ventricular hypertrophy. The blood flows into aorta from both right and left ventricles as a result of overriding and ventricular septal defect. Tetrology of Fallot with pulmonary atresia and major aortopulmonary collateral arteries (MAPCAS) is unique from most other types of complex congenital heart disease. Most of the cases of Tetrology of Fallot require surgical intervention and the mortality rate in untreated patients reaches 50% by age of 6 years. MAPCAS which are thought to derive from the embryonic splanchnic vascular plexus, contribute to the pulmonary blood supply in 30-65% of patients with Tetrology of Fallot and pulmonary atresia which prevents the patients from developing cyanotic spells. Our patient never had any cyanotic spells till nine years of age despite having marked cyanosis and grade IV digital clubbing. This is debatable whether it is advantageous or if it is harmful not to have cyanotic spells. Absence of cyanotic spells on one side had delayed his visit to hospital; on the other side having developed collaterals made him survive so long even without treatment. This was also evident that we were not dealing with a simple case of a cyanotic heart disease.

Keywords:
Tetrology of Fallot is the most common form of cyanotic congenital heart disease.

INTRODUCTION
Case reported is a 9 year old male child, with history of cyanosis and dyspnoea on exertion and effort intolerance since 9 months of age. There was no history of squatting to relieve cyanosis and dyspnoea. No history of cough, leg swelling, oliguria, syncopal attack, any convulsions or neurological deficits. Birth history was normal, full term, 2700 grams, with an Apgar of 9. He had no family history of congenital heart disease, arrhythmias or other cardiac diseases. General examination revealed an asthenic plethoric patient, centrally cyanosed with grade 4 digital clubbing. His pulse was regular with a rate of 82 beats per minute, respiratory rate of 28 beats per minute, regular. Blood pressure was 100/62mmhg. Saturation was 68% on room air. On examination of the precordium, the apex beat was visible on the right side in the 4th intercostal space 1.5 cm medial to the right mid-clavicular line with precordial bulge. First and second heart sounds were present with no murmur. Lungs were clear. Abdomen was normal without any organomegaly.
DISCUSSION

Tetrology of fallot makes about 10% of all congenital heart diseases. Pulmonary atresia with MAPCAs constitutes about 10% of patients with tetrology of fallot. The etiology is multifactorial, some of them being untreated maternal diabetes, phenylketonuria, and excess intake of retinoic acid. In patients with tetrology of fallot with pulmonary atresia, the pulmonary blood flow is mainly through a patent ductus arteriosus (70%) and less frequently via systemic collaterals (30%). PDA and collaterals may coexist as the source of pulmonary blood flow in such defects. The ductus is small and long and descends vertically from the transverse arch and connects to the pulmonary arteries which are confluent usually. Sometimes, a continuous murmur representing PDA shunt may be audible in a deeply cyanotic neonate who has TOF with pulmonary atresia. A right ventricular tap and a systolic ejection murmur at the mid left sternal border are usually heard on auscultation.

Transposition of great arteries, as the name implies, the two main arteries are transposed, with the aortic opening arising from the right ventricle, and the pulmonary artery arising from the left ventricle causing the hypoxemic blood circulating in the body and hyperoxemic blood in the pulmonary circulation. For the child to be able to survive with TGA, mixing of both the circulation is necessary. Hence, an ASD, VSD or a PDA should be present along with TGA.

Brain abscess, cerebrovascular accident, and subacute bacterial endocarditis are some of the complications associated with this disease.

Tetrology of fallot has a very classical presentation of cyanosis few weeks after birth and cyanotic spells with an audible murmur and a boot shaped heart with oligemia on chest radiograph. Unless the child’s heart is repaired surgically TOF is fatal.

The surgical repair can either be done as a primary surgical repair, staged repair or surgical unifocalization of pulmonary blood flow. The surgical mortality varies from 5% to 15%.

Here, we have a patient with central cyanosis without any cyanotic spells which delayed his first visit to the hospital and hence delay in the diagnosis of the complex heart disease. The age of presentation and symptoms were going against Fallot’s Tetrology. The case was made complicated due to associated dextrocardia, corrected transposition of great arteries, ASD, PDA and MAPCAs with atrioventricular discordance. There was thoracoabdominal discordance since...
liver was in the right hypochondrium and spleen in the left hypochondrium.

The late age of presentation and his survival can be attributed to the associated collaterals and TGA with left to right shunts. This atypical presentation of Tetrology of fallot with associated heart defects without any obvious complications make it a rare case and hence the need of reporting.

CONCLUSION

TOF with associated heart defects can be corrected surgically early with improved outcomes among patients. Efforts should be made on our part as health workers to identify these patients as early as possible so that they can benefit from corrective surgery to prevent late complications and poor quality of life.

References


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