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Research Article

A CASE OF PINK TETRALOGY OF FALLOT

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A R T I C L E I N F O

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A B S T R A C T

Tetralogy of Fallot is a well known congenital cardiac defect, has four major components and various minor associations. The four major components being overriding of aorta, membranous ventricular septal defect, right ventricular hypertrophy and right ventricular outflow tract obstruction. Survival of the patient depends on the degree of right ventricular outflow tract obstruction. In literature there are reports of survival confirmed beyond fifth decade with or without correction. If not present at birth, cyanosis develops in the first year of life. If the right ventricular outflow tract obstruction is mild and ventricular septal defect is in balance the patient might not have cyanotic spells is referred as having ''Pink Tetralogy of Fallot.'' Absence of Pulmonary valve is seen in only 2-6 % of TOF cases whose documented survival was not beyond 22years.

INTRODUCTION

A 24 years old male, school dropout, resident of Karaikal, Pondicherry has presented with sudden onset weakness and sensory loss of left upper and lower limb for 1 day while he was in his home which was static in nature. Patient was having a history of a Fever, Headache and Vomiting for the past 1 week. Fever was of high grade, continuous type and associated with chills and rigors. Headache was generalised, and throbbing in character associated with vomitings which were projectile. No History of loss of consciousness, seizure episode or double Vision. Patient was diagnosed to have 'Hole in the Heart' at 16 days of age on a routine checkup after which there was no follow up as he was completely asymptomatic. No history of any cyanotic spells or repeated respiratory tract infections or developmental delay in the childhood or seizure attacks. Patient was born to mother with an unremarkable marital and antenatal history and was not having history of delayed cry or difficulty during breast feeding. Patient who was physically fit during his schooling had to be dropped from school in view of his lack of interest to studies.

General Physical examination was unremarkable except he was febrile with 101^0F. Pulse was 102 per minute, regular and large volume pulse with no radio-radial or radio- femoral delay. Blood Pressure was 110/70 mm Hg and respiratory rate was 26 per minute. Systemic examination showed Higher Mental Functions and all Cranial Nerves intact. And motor system examination revealed normal bulk with hypertonia and brisk reflexes on left side with a complete loss of power.

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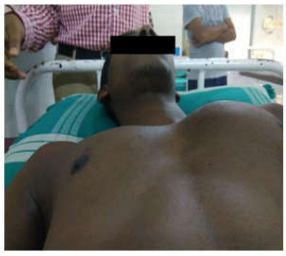
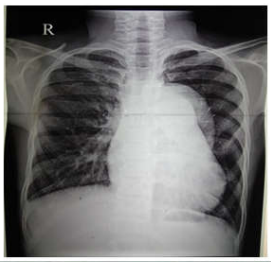
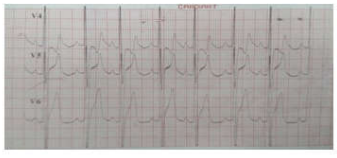
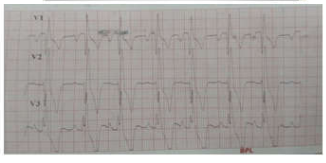
Sensory system showed loss of all modalities of sensation on left side. Cerebellar system, spine and cranium were normal on examination. No signs of meningeal irritation were present. Cardiovascular System has showed precordial bulge which was active on inspection with apical Impulse felt in 6th Intercostal Space ½ inch medial to Mid-clavicular line and hyperdynamic in character. Parasternal Impulse Present Grade III- AIIMS Grading was felt. Systolic Thrill is present at 4th ICS on left sternal edge. On Auscultation, first and second heart sounds were present with normal intensity. A harsh Pan-systolic Murmur of Grade 5, heard with diaphragm of stethoscope over left parasternal area was also heard.

Blood investigations of the patient were normal limits except for a Total WBC count of 12,100 cells per ml with an Hemoglobin of 13.8 gm/dl.



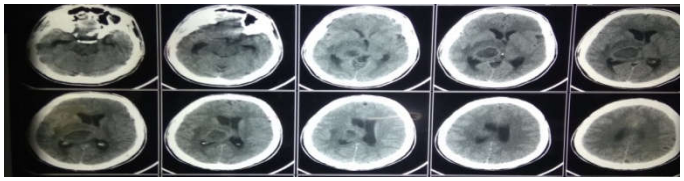






ECG showed sinus rhythm with right atrial enlargement and biventricular hypertrophy.

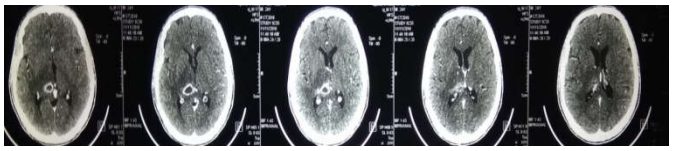
Chest X-ray showed lungs fields to be clear with Cardiomegaly and large homogenous curvilinear opacity on the left over the cardiac shadow mostly aneurysmal dilatation of pulmonary artery



Computed Tomographic imaging of Brain showed large lobulated capsulated hypodense or cystic lesion involving right thalamus, right capsular tracts with surrounding vasogenicedema effacing right lateral ventricle with midline shift towards left side and the diagnosis of Deep seated Cerebral Abscess was confirmed on CECT BRAIN.

Echocardiography showed large Sub-Aortic ventricular septal defect with Bi-directional shunt, Right Ventricular Hypertrophy, Mild Infundibular Pulmonary Stenosis, Absent Pulmonary Valve with dilated Pulmonary artery, Dilated Right Atrium and Right Ventricle, Normal Bi-ventricular function and contraction, Left Ventricular Ejection Fraction- 68%, Intact inter atrial septum without any collaterals. These findings were confirmative of Tetrology of Fallot with mild Right Ventricular Outflow Tract obstruction.

Immediate neurosurgeon opinion was asked for and patient was taken up for Abscess drainage through High Parietal Burr-Hole approach and as the post surgery CT brain has showed residual abscess, retapping was done.



And after which the CT brain has showed no significant changes when compared to previous CT brain. Pus swab sent from brain abscess has showed many pus cells, moderate gram positive cocci in pairs and chains and few gram negative bacilli. Organism was found to be Coagulase negative Staphylococcus sensitive to Amikacin, Clindamycin, Gentamycin, Oflaxacin, Teicoplanin, Vancomycin, Cefazolin. Patient recovered well post operatively and was advised physiotherapy and regular follow-up. No specific events were noted in two month follow up interval.

DISCUSSION

Tetralogyof Fallot with an incidence of 5.7% of all the congenital cardiac defects is one of the common congenital heart diseases seen. With advances in both palliative and curative surgery, the number of children with congenital heart diseases surviving to adulthood has increased dramatically. Despite these advances, congenital heart diseases remain the leading cause of death in children with congenital malformations.

The four components of TOF are ventricular septal defect (VSD), aortic override, right ventricular outflow tract obstruction and right ventricular hypertrophy. When obstruction of right ventricular outflow is mild to moderate and a fairly balanced shunt operates across VSD as a result of which patient may not be cyanotic i.e. Acyanotic or Pink Tetralogy of Fallot. When obstruction is severe, cyanosis will be present from birth and worsen when the ductus arteriosus begins to close.

Survival in patients with unoperated TOF is linked to degree of right ventricular outflow obstruction. Attrition is highest in severe pulmonary stenosis. With tetralogy of fallot of all degrees of severity, 11% are alive at age of 20 years, 6% at age of 30 years, 3% at age of 40 years. There are number of reports of patients surviving till seventh decade.

In 2% to 6% of patients with TOF, pulmonary valve tissue is either absent or consists of rudimentary remnants of cellular, vascular, myxomatous or primitive connective tissue. Usually marked by signs of upper airway obstructions, cyanosis may be absent or mild or moderate, heart is large and hyperdynamic with loud to and fro murmur. Marked aneurysmal dilatation of main and branch of pulmonary arteries results in compression of bronchi and produces recurrent pneumonia. Occasionally, such patients go through infancy with relatively few symptoms but respiratory distress due to trachea-bronchial obstruction and right ventricular failure conspire to limit longevity. One patient survived to age 22 years[1]. Other associated anomalies are absence of pulmonary artery branch, most often the left, usually suspected by difference in pulmonary vasculature on two sides. Tetrology of Fallot may be one of the spectrum of lesions like CATCH 22, DiGeorge syndrome or Shprintzenvelo - cardiofacial syndrome[2].

The evaluation of congenital heart disease has to be done in a three step approach. First, they have to be divided into two groups based on presence or absence of cyanosis. Second, using the increased or normal or decreased vascular markings over chest radiography and finally, using electrocardiography to identify presence of right, left or biventricular hypertrophy a diagnosis can be presumed. The character of heart sounds and the presence and character of any murmur further narrow the differential diagnosis. Using echocardiography, CT, MRI or cardiac catheterization, diagnosis can be confirmed. Echocardiography also identifies coexisting congenital malformations such as absent pulmonary valve, absent left pulmonary artery, endocardial cushion defects and ASD. The malaligned VSD and the biventricular aorta are well seen from parasternal and apical views. But for identifying and characterizing the levels of right ventricular outflow obstruction multiple subcostal and suprasternal views are usually needed. Multiplane Trans esophageal echocardiography (TEE) provides the most versatile angles to view the right ventricle outflow tract and the pulmonary artery segment. Although TEE can usually image the origins of aortopulmonary collaterals, standard selective angiography, magnetic resonance techniques and CT angiography are superior. 2D imaging with color flow can distinguish TOF with pulmonary atresia from truncus arteriosus[3]. Tissue Doppler imaging, a newer technique provides valuable information regarding systolic, diastolic and iso volumetric indices of ventricles has been used to study the diastolic and systolic parameters in TOF.

The physiologic consequences of TOF depends chiefly on the degree of obstruction to right ventricle outflow and to a lesser extent on systemic vascular resistance[4]. Right ventricular outflow obstruction is fixed but systemic resistance is variable[5].

Treatment of TOF depends on severity of Right ventricular outflow tract obstruction. The modified Blalock-Taussig shunt is currently the most common aortopulmonaryshunt procedure and consists of a Gare-Tex conduct anastomised side to side from the subclavian artery to the homolateral branch of pulmonary artery with post operative complications including chylothorax, diaphragmatic paralysis, Horner's syndrome.

Corrective surgical therapy consists of relief of right ventricular outflow obstruction by removing obstructive muscle and by patch closure of VSD by a transatrial-transpulmonary approach.

A small but not insignificant group of patients with TOF, die suddenly after years of repair via right ventriculotomy due to sustain recurrent monomorphic ventricular tachycardia as there is an increase in scar tissue with age. Revision of the ventricular scar and radiofrequency ablation of residual obviate the risk of monomorphic ventricular tachycardia and substantially increase the prospects of long term survival.

Before corrections, patient with TOF are susceptible to several serious complications.

Cerebral thrombosis is commonly seen along with extreme polycythemia and dehydration usually before the age of 9 years. Brain abscess is extremely rare. Bacterial endocarditis may also occur. Heart failure is not a usual feature, but is usually seen in young infant with Pink TOF. A brain abscess should be suspected when a cyanotic patient experiences protracted headache, focal neurological signs, seizures and fever[6]. The diagnosis of a fresh brain abscess is readily made by computed tomography by the distinctive ring enhancement. Brain abscess may be due to a septic embolus, contagious extension of infection or direct mechanical introduction of infectious organisms by a surgical procedure or trauma[7]. Two preconditions appear necessary; Firstly, Bactermia and second, a focal zone of cerebral vulnerability[8].

If diagnosed in early phase of cerebritis treatment with antibiotic therapy alone can be adequate[9]. Most frequently, however, treatment requires aspiration employing computed tomographic or magnetic resonance directed stereotactic control.Culture and sensitivity tests of the aspirate set the stage of specific antibiotic therapy.

Our case was unique in many ways: Firstly, survival of a Pink tetrology of fallot with absent pulmonary valve till age of 24years has not been reported anywhere. Second, inspite of being having severe aneurysmal dilatation of pulmonary artery patient has not presented with respiratory tract obstruction. Third the absence of Pulmonary valve which is seen in only 2-6% TOF cases.

In conclusion, we report a case of Pink TOF with Absent Pulmonary Valve who has survived till 24years which is more than ever reported. Even though the expected presentation was to have repeated respiratory tract infection, here the patient has presented with Cerebral Abscess and recovered very well after acute surgical intervention with an uneventful follow up.

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