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Tetralogy of Fallot: Cause of Sudden Cardiac Death in Young Individual

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Abstract

Congenital Heart Diseases (CHD) include the abnormalities of the heart and great vessels that are present at birth. Individuals with CHDs may survive till adulthood. We report a case of sudden cardiac death in a young adult who was suffering from Tetralogy of Fallot (TOF) with Atrial septal defect (Pentalogy of Fallot) -a CHD. He collapsed in the hospital premises and was immediately admitted and treatment ensues, but couldn't be saved. Autopsy revealed an oval shaped defect in inter-atrial septa, Right Ventricular hypertrophy, over riding of Aorta and Pulmonary atresia. Coronaries were patent. The authors aim to highlight the need of early diagnosis and timely intervention in such heart diseases. The authors also intend to add to Medical Literature that TOF may be the cause of a sudden death.

Key words: Tetralogy of Fallot, Congenital Heart Disease, Sudden Cardiac Death, Right Ventricular Dysfunction.

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Introduction

Cardiovascular diseases (CVD's) are well established cause of Sudden deaths with increasing frequency in young adults[1,2,3]. Coronary artery disease (CAD) is the main cause of Sudden Cardiac deaths (SCD)[1,4]. In the previous issue of this journal, a case report of SCD of a young individual was reported to indicate increased risk of Sudden Cardiac Death due to underlying CAD in young adults. Besides CADs, other causes of SCD include disease of pericardium & myocardium, valvular disorders or aortic disorders, which in young adults may be due to genetic or hereditary causes1,4. Congenital Heart Diseases (CHD) include the abnormalities of the heart and great vessels that are present at birth. CHDs are one of the most prevalent birth defects with an incidence of 5%. Severe anomalies are incompatible with survivals leading to intrauterine deaths whereas some CHD, like septal defects, unilateral obstructions & outflow tract, produce clinical manifestation only after birth[4]. Individuals with CHDs may survive till adulthood[4]. We report a case of sudden cardiac death in a young adult with history of Tetralogy of Fallot -a CHD. The authors aim to highlight the risk of sudden fatality associated with this condition and also intend to add to Medical Literature that CHDs may be the cause of sudden death.

Case Report

A 25 years old young male collapsed and became unconscious at about 12 pm in Cardiology wing of All India Institute of Medical sciences, New Delhi after which he was admitted and expired during the treatment on same day at about 10:54 pm. In postmortem examination, clothes were intact. Bluish discoloration i.e. cyanosis along with clubbing was present over nail beds of both hands (Image-1). No external injuries were present over the body. Brain was edematous weighing 1050 gms with clotted blood seen in 3rd and 4th Ventricles. Pleura was adherent to overlying chest wall and underlying lungs on both the sides. Lungs were congested and adherent to chest wall. Heart was weighing 275 gms. On dissection an oval shaped

defect of 0.2 x 0.1 cm is present in inter-atrial septa **(Image-2).** Right Ventricular hypertrophy was present **(Image-3).** Over riding of Aorta and Pulmonary atresia is present. Coronaries were patent. Stomach was containing about 100 ml of clotted blood with no peculiar smell and normal mucosa. Other Visceral organs were congested. The hospital record revealed that the deceased was suffering from a Congenital Cyanotic Heart Disease (CCHD) with reduced pulmonary flow - Tetralogy of Fallot with Pulmonary tresia and Hypoplastic Left Pulmonary Artery. The cause of death was concluded as "Myocardial Insufficiency consequent upon Tetralogy of Fallot- a congenital heart disease and the manner was decided as Natural".

Fig. 1 Cyanosis along with clubbing present over nail beds of hands.

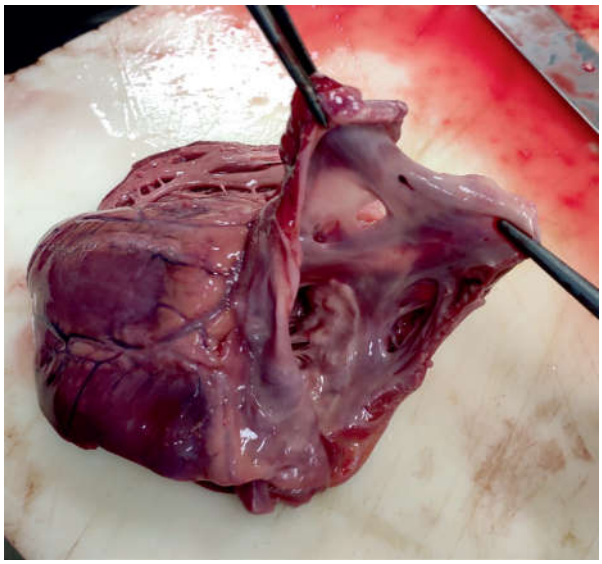

Fig. 2 An oval shaped defect of 0.2 x 0.1 cm is present in inter-atrial septa.

Discussion

Tetralogy of Fallot (TOF) is the most common in Cyanotic form of Congenital Heart Diseases (CHD) and is symptomatic manifest early in postnatal life due to right to left shunt. Transposition of the great arteries, persistent truncus arteriosus, tricuspid atresia, and total anomalous pulmonary venous connection are some of the other disorders in the group[4,5]. TOF is characterized by[4,5]

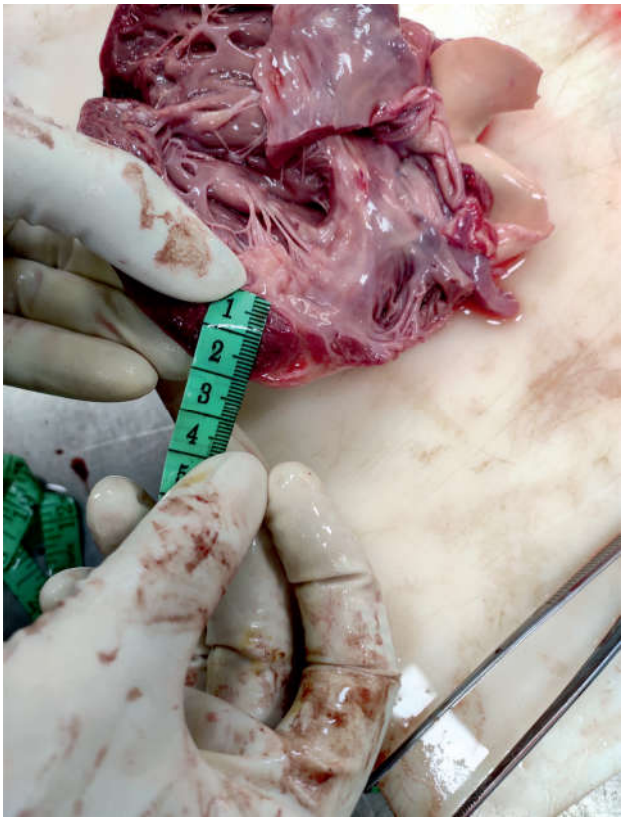

Fig. 3 Right Ventricular hypertrophy

1. Ventricular Septal Defect (VSD).
2. Obstruction of the right ventricular outflow tract: It is usually due to narrowing of the infundibulum (subpulmonic stenosis) but can be accompanied by pulmonary valvular stenosis (sub pulmonary stenosis),
3. Aorta overriding VSD: The VSD is usually large with the aortic valve at the superior border. So Aorta overrides the defect and both ventricular chambers.
4. Right ventricular hypertrophy: The heart is enlarged and "boot-shaped" due to marked right ventricular hypertrophy.

Diagnosis of TOF is confirmed by Echocardiography. The patients with TOF can survive into adult life even if they did not receive treatment. 10% of untreated patients are alive at 20 years and 3% survive for 40 years. The clinical course depends primarily on the severity of the subpulmonary stenosis, which leads to hypoplastic pulmonary arteries and larger overriding of

Aorta4,5,6. The deceased in the present case survived till 25 years of age. The deceased had The irony of the case is that the deceased was collapsed while sitting in the OPD waiting area of cardiology block of AIIMS, Hospital. He was immediately take to emergency and administered treatment but still could not be saved. The terminal event leading to death was mentioned as Cardiogenic Shock in the hospital records.

Surgical repair is the mainstay treatment of TOF. Early repair is advised to minimize the effect of long-standing hypoxia and pressure overload. The optimal timing for surgical correction in children is less than 1 year of age in developed countries. Corrective surgery of TOF is often conducted in newborn period when the symptoms of cyanosis are present. Complete surgical repair is possible but becomes complicated for individuals with pulmonary atresia and dilated bronchial arteries[4,-7]. The s of the surgery are good and most patients with TOF have an uneventful postoperative course. Postoperative Right Ventricle (RV) myocardial dysfunction may lead to surgical complication and even death5,6. Death is caused by ventricular tachycardia and fibrillation and is related to postoperative right ventricular hypertrophy or dilation mainly as a of pulmonary valve stenosis or regurgitation[8-10].

Detailed perusal of Treatment Records revealed that deceased was operated in 2003 with Blalock and Taussig Shunt. Blalock-Taussig shunt (BTS) is the main first step first-surgical management for patients for maintaining pulmonary blood flow11. Yamada11 reported a case of women with TOF with BTS operation at 10 years of age who survived upto 72 years despite no medication. In the present case deceased was operated in 2003 when he was of 9 years old but in 2019 his right side of Shunt was found blocked which decreased his life expectancy. He was currently suffering from occluded right side Blalock and Taussig Shunt, Conical Patent Ductus Arteriosus with distal Left Pulmonary artery (LPA) insertion site stenosis. Major Aorto Pulmonary Collateral Arteries (MAPCA) were also present. Echocardiography report of deceased conducted in June 2019 revealed TOF with pulmonary atresia and overriding of Aorta.

Achour[12] reported a rare case of TOF with pulmonary atresia in a 40-year-old survivor patient, despite a chronic pediatric shunt thrombosis. The patient became symptomatic at 38- years-old with progressive dyspnea on exertion and short cyanosis spells but prolonged survival was due to expansion of several and huge major aorto-pulmonary collateral arteries, a finding similar to the present case.

In the present case, the deceased was also found having Atrial Septal defect (ASD) at Autopsy which was not diagnosed in Hospital investigation reports. This condition is called as Pentalogy of Fallot. Pentalogy of Fallot is a rare form of cyanotic congenital heart disease, characterized by an association of ASD with TOF[13]. Lsaksen et al reported that 4% of major and 18% of minor findings detected on autopsy were not found on ultrasound examination[14]. Shang Gao studied the comparison of Fetal Echocardiographic (FE) diagnoses with cardiac autopsy findings and concluded that 11.7% (20/171) of cases, autopsies disclosed new deformities which were either not diagnosed or misdiagnosed by FE [15].

Conclusion

TOF is treatable with good postoperative recovery. Early diagnosis and timely surgical intervention is of the essence else it may lead to sudden fatality. Verification of an antenatal diagnosis of cardiac anomaly in a fetus or an infant by autopsy plays an important role in the further management and survival. In an undiagnosed case of SCD TOF may be the cause of death and same should also be kept in consideration while treating a collapsed patient in emergency department.

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