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TETRALOGY OF FALLOT WITH DEXTROCARDIA WITH SINUS SOLITUS PRESENTING IN ADOLESCENCE

Cardiology

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ABSTRACT

Tetralogy of Fallot's (TOF) is the commonest cyanotic congenital heart disease with an incidence of 1 in 2000 newborns. Majority of children present in infancy. Left untreated, only 3% survive to adulthood, Anatomically, it consists of 4 components – Ventricular septal defect; Pulmonic stenosis; Right ventricular hypertrophy; Over-riding aorta. TOF with dextrocardia has rarely been reported.

These sporadic case reports have almost all involved cases of mirror-image dextrocardia with situs inversus and TOF. Only one reported case of TOF and dextrocardia with abdominal situs solitus, detected on ultrasonogram in a fetus of 26 weeks' gestation has been published. The altered spatial orientation of the cardiac structures in case of dextrocardia can present a surgical challenge. A case of TOF with dextrocardia with late presentation during adolescence is presented here. Age at presentation is governed by severity of right ventricular outflow tract obstruction. In the present case, diameter of left pulmonary trunk was 3 mm only. However, right pulmonary trunk had a normal diameter of 10 mm. And this selective obstruction of left pulmonary artery origin is speculated to be the cause of lesser severity of symptoms and, thus, late presentation at the age of 16 years in our case.

KEYWORDS

Tetralogy of Fallot, Dextrocardia, Sinus solitus, Adolescence

1. Introduction

Tetralogy of Fallot's (TOF) is the most frequently diagnosed cyanotic congenital heart defect with an incidence of 1 in 2000 newborn.[1] Majority of children present during infancy. Without surgery, mortality rate gradually increases, ranging from 30% at the age of 2 years to 50% by 6 years of age. The mortality rate is highest in the first year and then remains constant until the second decade. Not more than 20% of patients can be expected to reach the age of 10 years, and fewer than 3% of patients are alive by the end of their second decade.[2]

It is often associated with additional findings such as secundum atrial septal defect or right-sided aortic arch. Its association with dextroversion is rarely reported and can present technical challenges to intracardiac repair. Dextrocardia associated with situs solitus (so called 'dextroversion') is a cardiac positional anomaly in which the heart is located in the right hemithorax with its base-to-apex axis directed to the right and caudally, along with normal anatomical position of other intrathoracic and abdominal viscera. The malposition is intrinsic to the heart and not caused by extracardiac abnormalities. In dextroversion, there is a 90% incidence of additional cardiac malformations, including anomalous pulmonary venous return, TOF, septal defects, pulmonic stenosis, coarctation of aorta and corrected transposition of great arteries (TGA). A rare case of TOF with dextroversion in a 16-year-old female is presented here.

2. Case Report

A 16-year-old female presented with the chief complaint of progressive respiratory distress since the age of 6 years. The distress was mainly on exertion, earlier on walking uphill and now it was present on walking on a flat surface also. It was relieved by squatting and standing still. A history of recurrent cyanotic episodes and lower respiratory tract infections was also present. Perinatal history was insignificant. There was no associated history of seizures, syncope, swelling of limbs, weakness of extremities or paroxysmal nocturnal dyspnea.

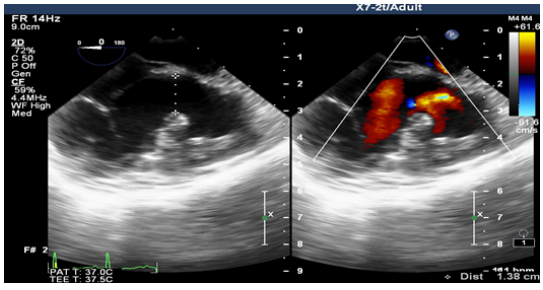
On general physical examination, the patient had tachycardia (pulse rate 118 beats per minute) with regular rhythm and all peripheral pulses were palpable. Systolic blood pressure in the upper and lower limbs was 110 and 100 mm Hg respectively, whereas diastolic pressure was 60 mm Hg in both. Central cyanosis with second degree clubbing in all four limbs was evident. Pedal edema (pitting) was present up to the ankles.

Examination of the cardiovascular system revealed a palpable apex beat localized in the right 5th intercostal space just medial to the mid-clavicular line along with a palpable right parasternal heave. On auscultation, a mid-systolic murmur of grade III/IV intensity was heard in the right 2nd intercostal space, augmented on repeated sitting and standing. First and second heart sounds were normal and cardiac gallop was present. Abdominal examination revealed a firm and tender hepatomegaly 4 cm below costal margin in the mid-clavicular line on right upper quadrant. Spleen was not palpable but Traube's space was tympanic confirming that stomach was on the left side. Respiratory system examination revealed bilateral basal fine end-inspiratory crepts.

Chest radiograph showed oligemic lung fields with cardiac apex on right side and aortic knuckle on the left confirming dextrocardia with features likely suggestive of pulmonic stenosis. Abdominal ultrasonogram showed normally positioned viscera with enlarged liver up to 18 cm with features suggestive of congestive hepatitis, thus giving us a diagnosis of dextrocardia with situs solitus.

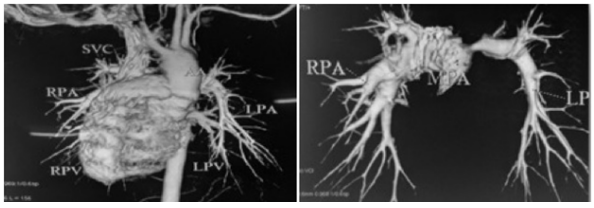
A transthoracic echocardiography followed by trans-esophageal echocardiography was done, which showed dextrocardia, subaortic ventricular septal defect (13mm) with bidirectional shunt, severe infundibular and valvular pulmonic stenosis with a pressure gradient of 120 mm Hg with right ventricular hypertrophy (Figure 1).

Fig 1. Echocardiograph showing subaortic ventricular septal defect (13mm) with bidirectional shunt, severe infundibular and valvular pulmonic stenosis with right ventricular hypertrophy



Cardiac computed tomography scan was diagnostic of dextrocardia with visceral situs solitus. Main pulmonary trunk was 12 mm in diameter whereas diameters at the origins of right and left trunks were 10 and 3 mm respectively (Figure 2).

Figure 2: Cardiac CT showing obstruction at left pulmonary trunk with decreased diameter [SVC- superior vena cava, RPA- right pulmonary artery, LPA- left pulmonary artery, RPV- right pulmonary vein, LPV- left pulmonary vein]



Patient was initially managed on beta blockers to control the tachycardia, and on diuretics in view of right ventricular failure and severe pulmonary hypertension. Patient underwent total correction of the cardiac defect with enlargement of origin of left pulmonary trunk. Post-operative course was uneventful. She was asymptomatic at 6-month follow-up without any persistence or recurrence of symptoms.

3. Discussion

Anatomically, TOF consists of 4 components – Ventricular septal defect, pulmonic stenosis, right ventricular hypertrophy and over-riding aorta. Severity of right ventricular outflow tract obstruction governs clinical presentation. This leads to the systemic circulation of deoxygenated blood, resulting in symptoms of cyanosis, polycythemia and hypoxia. In the present case, diameter of left pulmonary trunk was 3 mm only. However, right pulmonary trunk had a normal diameter of 10 mm. This selective obstruction of left pulmonary artery origin is speculated to be the cause of lesser severity of symptoms and, thus, late presentation at the age of 16 years as in the present case.

TOF with dextrocardia has rarely been reported.[3,4] These sporadic case reports have almost all involved cases of mirror-image dextrocardia with situs inversus and TOF. Only 1 other reported case of TOF and dextrocardia with abdominal situs solitus, detected on ultrasonogram in a fetus of 26 weeks gestation has been published.[5]

Altered spatial orientation of the cardiac structures in case of dextrocardia presents a surgical challenge. Complete surgical correction is now the most important and standard treatment. Palliative surgery includes the Blalock-Taussig shunt or Potts shunt which constructs a communicating channel between the systemic and pulmonary circulation. However, the outcome is poor and it is no longer standard treatment for TOF except when the patient's general condition is not suitable for repair. It may be a bridge between symptomatic relief and total correction.

Thus, children with TOF can present at an older age in case the right ventricular outflow obstruction is partial, as in the present case. Total correction, if feasible, is curative and should be attempted even in late presenters.

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