Tetralogy of Fallot and transverse aortic coarctation: A rare coexistence and its treatment

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Introduction

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease. Long-term outcomes are favorable after corrective surgery, and 90% of patients reach adulthood. TOF is usually described as isolated obstructions of the right heart and are rarely accompanied by left-sided obstructive lesions such as cor triatriatum, mitral stenosis, subaortic or aortic stenosis, and aortic coarctation (1).

In this report, we describe the presentation and treatment of an extremely rare case of TOF with left aortic arch and coarctation between the left common carotid and left subclavian arteries.

Case Report

A 3-day-old baby boy weighing 2,960 g was born at a gestational age of 39 weeks to a healthy 29-year-old mother by spontaneous vaginal delivery. The infant had a cleft palate and lip and was experiencing feeding difficulty and respiratory distress. On physical examination, a 3/6 systolic murmur was heard in the pulmonary area. The chest radiograph showed coeur en sabot sign. Treatment with prostaglandin E1 (PGE1) infusion was initiated, the patient was referred to our hospital, and a pediatric cardiology consultation was requested. The infant's oxygen saturation on room air was 97%–99%. There was no difference in blood pressure between the upper and lower extremities, and lower extremity pulses were palpable bilaterally. Two-dimensional echocardiogram (ECHO) performed at postnatal 72 hours revealed TOF. A large malalignment ventricle septal defect (VSD), 50% aortic dextroposition, normal tricuspid and aortic valves (Video 1), and infundibular and pulmonary valve stenosis with a maximum gradient of 45 mm Hg were detected (Video 2). Both pulmonary arterial branches were hypoplastic but confluent. An appearance suggesting discrete stenosis proximal to the left subclavian artery was noted, and a maximum 20 mm Hg peak instantaneous gradient without a diastolic pattern was measured at that location. A large patent ductus arteriosus (PDA) with left-to-right shunt was detected. After six hours, the PGE1 was discontinued, and control ECHO showed severe stenosis (maximum gradient 50 mm Hg and diastolic pattern) when PDA started to close. The PGE1 infusion was reinitiated.

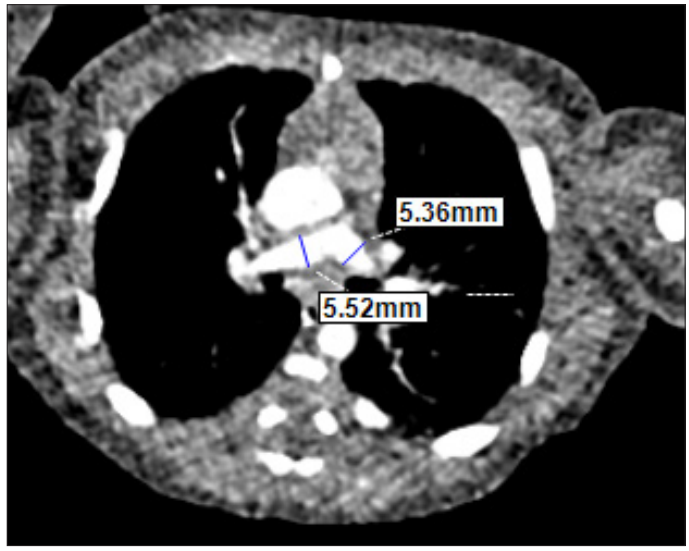

Figure 1. Thoracic computed tomography angiography showing the hypoplastic left and right pulmonary arterial branches. McGoon index 1.69

Thoracic computed tomography (CT) angiography revealed TOF and left aortic arch with severe aortic coarctation distal to the left common carotid artery and proximal to the left subclavian artery (Video 3). McGoon index was 1.69 (Fig. 1). The patient's clinical presentation (hypoplastic pulmonary arteries and both left and right ventricular outflow stenosis) was discussed in the pediatric cardiology and cardiac surgery council. There were concerns that systemic vascular resistance may decrease after the repair of the aortic coarctation and that pulmonary artery stenosis and hypoplasia could result in desaturation postoperatively. The council's decision was to perform aortic coarctation repair surgery with left modified Blalock-Taussig (MBT) shunt. Our patient had severe coarctation of aorta, and we had to perform an emergency operation.

The operation was performed by a pediatric cardiac surgeon. The aortic coarctation was repaired using a 1.5 cm-wide bovine pericardial patch between the transverse aortic arch and the distal descending aorta. PDA was closed during the surgery. The left MBT shunt placement planned owing to the patient's McGoon index of 1.69 could not be performed because sufficient flow was not detected in the left subclavian artery. The infant's oxygen saturation fell to 65%–70%. The case was discussed peroperatively, and it was decided to perform shunt surgery between the descending aorta and left pulmonary artery using a 3.5 mm polytetrafluoroethylene (PTFE) tubular graft. On postoperative examination, the shunt was functional, ventricular ejection fraction was normal, and the patient was clinically stable, and has been under follow-up until a full corrective surgery can be performed. Several studies have demonstrated that patients with TOF have the 22q11 deletion, which increases the risk of the presence of aortic arch or vascular anomalies. Our patient may also have the 22q11.2 deletion, and the genetic testing for it is ongoing.

Anatol J Cardiol 2021; 25: 743-4
DOI:10.5152/AnatolJCardiol.2021.39551

Discussion

The original description of TOF includes four abnormalities: a large VSD, right ventricular outflow stenosis, right ventricular hypertrophy, and an "overriding" aorta. The coexistence of aortic coarctation and TOF is very rare. In a large retrospective study, aortic coarctation was reported in only one of 2,235 patients diagnosed with TOF (2). Two types of coarctation have been associated with TOF in the literature. It has been detected distal to the left subclavian artery in patients with left aortic arch and distal to the right common carotid artery in patients with a right aortic arch, as a mirror image of the interrupted aortic arch type B. Perdreau et al. (3) evaluated 12 patients with a combination of TOF and aortic coarctation and observed that coarctation was distal to the left subclavian artery in all patients with a left aortic arch. Our patient is rare in that he had a left aortic arch and coarctation between the left common carotid and left subclavian arteries.

Mild aortic coarctation was detected on ECHO at admission and at postnatal 72 hours while the patient was receiving low-dose PGE1 infusion. However, control ECHO performed within six hours after PGE1 was discontinued revealed a substantial increase in the coarctation gradient. The absence of cases similar to ours in the literature may be because the condition of the patients suddenly deteriorates during PDA closure and returns physiologically to a clinical presentation consistent with type B interruption. The presence of coarctation in its usual location, distal to the left subclavian artery in patients with a left aortic arch, can be explained by the migration of ductal tissues to the aortic wall (4). The worsening of coarctation during PDA closure observed in our patient supports the theory of ductal tissue migration to the aortic wall.

Our patient was treated with aortic coarctation repair with bovine pericardial patch and shunt operation with 3.5 mm PTFE tubular graft between the descending aorta and left pulmonary artery. In previously reported patients, coarctation repair and full Fallot repair, coarctation repair, and MBT shunt procedures were performed depending on clinical severity (5-8).

Close clinical follow-up and repeated ECHO during PDA closure are crucial in the surveillance of patients with TOF and mild aortic coarctation.

Conclusion

In conclusion, the cause of this combination of defects is difficult to explain. The coexistence of aortic coarctation and TOF is very rare. Therefore, early detection of these cardiovascular anomalies is crucial to guide management and consequently improve patient outcomes.

Informed consent: Informed consent was obtained from the patient.

Video 1. Echocardiogram showing tetralogy of Fallot, a large malalignment ventricular septal defect VSD, 50% aortic dextroposition, and normal tricuspid and aortic valves

Video 2. Echocardiogram showing infundibular and pulmonary valve stenosis

Video 3. Thoracic computed tomography angiography showing left aortic arch and severe aortic coarctation

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DOI:10.5152/AnatolJCardiol.2021.39551