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Congenital Aortic Arch Abnormality in Tetralogy of Fallot: A Rare Case Report

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Abstract

A 4-year-old male child was referred to the pediatrics department of our hospital from an Anganwadi with complaints of easy fatigability compared to peers, dyspnea on normal exertion and refusal to feed for 10 days. There was no history of palpitation, bluish discoloration, pedal edema, abdominal distention, cough, or orthopnoea. On auscultation, a grade 2 end systolic murmur was heard. Full blood count revealed an Hb 10 g/dl, MCV 67.2 fL, MCH 20.9 and HCT 32.1%. Liver and kidney function tests were within normal limits. However, the Total T3 (2.85 nmol/L) and TSH were elevated (9.30 mmol/L).

Chest X-ray indicated cardiomegaly with right ventricular enlargement with clear lung fields and pleural spaces. ECG further showed right axis deviation and enlargement of the right ventricle. He was diagnosed with Tetralogy of Fallot on 2D Echo, having confluent good-sized pulmonary arteries (Macgoon's ratio: 1:4), large malaligned VSD with BD shunt, severe valvular and subpulmonary annular stenosis. These findings were further confirmed on MDCT chest with pulmonary angiography. Furthermore, bilateral anomalous origin of both right and left vertebral arteries from the arch of aorta was also observed. Later the patient was scheduled for elective surgical repair. Sauvage patch closure of VSD was done with 5-O continuous prolene sutures followed by reconstruction of the RVOT by augmenting it with a pericardial patch.

The postoperative course was uneventful with minimal drainage. The patient was discharged on Injection Ceftumand Injection Lefoflox with infective endocarditis prophylaxis.

In this case, we aimed to discuss the diagnostic implications of variations in the aortic arch with a special focus on the imaging and approach to the management.

Keywords: Vertebral arteries anomalies; Implications and Diagnosis; Arch of aorta anomalies; Ceftumand injection

Introduction

Congenital anomalies of the aortic arch comprise diverse malformations that may be clinically silent or may present with severe respiratory or esophageal symptoms. They are essential to recognize as they may be associated with vascular rings, congenital heart disease and chromosomal abnormalities that can have important implications for prognosis and management.

Volume-rendered Computed Tomography (CT) and magnetic resonance angiography (MRA) help in preoperative surgical planning by providing information about the complex relationship of the aortic arch and its branches to the trachea and esophagus. Three-dimensional capabilities of both Computed Tomography Angiography (CTA) and MRA are helpful in determining evidence of tracheal or esophageal compression or other high-risk features in patients with a complete vascular ring. Both the endovascular interventionist and the diagnostic radiologist must have a thorough understanding of the major vessels of the aortic arch and their variations. Here we describe one such case of a 4-year-old male

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child having TOF with aortic arch anomaly, in which both right and left vertebral arteries arise from the aortic arch [1].

Case Presentation

A 4-year-old male child was referred to the pediatrics department of our hospital from an Anganwadi with complaints of easy fatigability compared to peers, dyspnea on normal exertion and refusal to feed for 10 days. There was no history of palpitation, bluish discoloration, pedal edema, abdominal distention, cough, or orthopnoea. A grade 2 end-systolic murmur was heard on auscultation in the aortic area. Full blood count revealed an Hb of 10 g/dl, MCV 67.2 fL, MCH 20.9 and HCT 32.1%. Liver and kidney function tests were within normal limits. However, the Total T3 (2.85 nmol/L) and TSH were elevated (9.30mmol/L) [2,3].

Chest X-ray revealed cardiomegaly (CTR-64) with right ventricular enlargement but clear lung fields and pleural spaces. ECG further showed right axis deviation and enlargement of the right ventricle. He was diagnosed with Tetralogy of Fallot on 2D Echo, having confluent good-sized pulmonary arteries (Mcgoon's ratio: 1:4), large malaligned VSD with BD shunt, severe valvular and subpulmonary annular stenosis on 2D Echo. These findings were further confirmed on MDCT Chest with Pulmonary Angiography **(Figure 1)**.

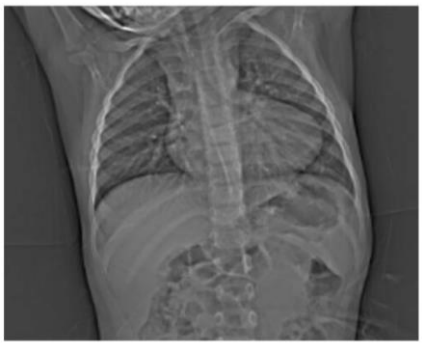

Figure 1: Chest X-ray showing cardiomegaly (CTR-64) with right ventricular enlargement but clear lung fields and pleural spaces.

On MDCT Chest with Pulmonary Angiography, bilateral anomalous origin of both, right and left vertebral arteries from the arch of aorta was seen. Later the patient was scheduled for elective surgical repair. As part of the corrective treatment, an intra-cardiac repair was performed with a short trans-annular pericardial patch. Sauvage patch closure of VSD was done with 5-0 continuous Prolene sutures followed by reconstruction of the RVOT by augmenting it with a pericardial patch. The postoperative course was uneventful with minimal drainage. The patient was discharged on Injection Ceftumand and injection Levoflox with infective endocarditis prophylaxis **(Figure 2)**.

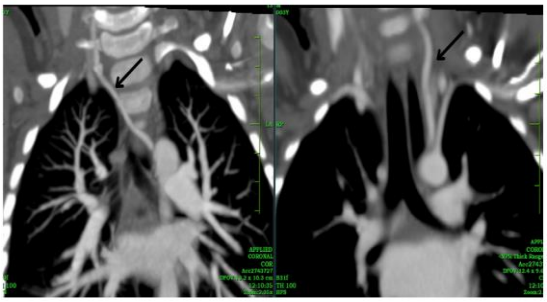

Figure 2: On MDCT chest with pulmonary angiography. (A) Aberrant right vertebral artery is seen arising from the aortic arch as the fifth branch; (B) Left vertebral artery is seen arising directly as the third branch from the aortic arch.

Discussion

Tetralogy of Fallot (TOF) is a complex congenital heart defect that occurs in approximately 3.9-7.7 per 10,000 live births. It is the most common cyanotic heart condition in children who have survived untreated beyond neonatal age, with the need for an intervention in the first year of life. TOF is characterized by a combination of Ventricular Septal Defect (VSD), pulmonary stenosis, overriding aorta and right ventricular hypertrophy. Aortic arch abnormalities are rare in TOF patients, with a reported incidence of less than 1%. However, when present, they can impact surgical management and long-term outcomes.

The development of the human heart starts around the 20th day of gestation, with the fusion of the outer endocardial tubes into a single tubular structure, the cardiac tube. Subsequently, the cardiac tube folds and loops, with the development of an atrium that is cranial and dorsal and a primitive ventricle moving downward, ventrally and to the right. The right ventricle is the dominant chamber in the embryo and fetus, receiving 65% of the venous return and is the main contributor to the lower part of the body, the placenta and the lungs. The right ventricle can be described by three components: the inlet, which consists of the tricuspid valve chordae tendineae and papillary muscles; the trabeculated apical myocardium; and the infundibulum or conus. The exact embryologic process that contributes to the development of TOF still is unknown, but an association that had been observed is an anterior and cephalad deviation of the infundibular septum that results in a misaligned ventricular septal defect, with an overriding aortic root causing a subsequent right ventricular outflow obstruction.

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The ventricular septal defects seen in patients with TOF are usually perimembranous that can extend into the muscular septum. Different factors can contribute to the right ventricular outflow obstruction, including the pulmonary valve that is usually bicuspid and stenotic, the hypo-plastic pulmonary valve annulus and the deviation of the infundibular septum that causes a subvalvular obstruction and the hypertrophy of the muscular bands in this region. The degree of the overriding aorta usually varies and receives blood flow from both ventricles [4-6]

While TOF is a relatively common congenital heart defect, aortic arch abnormalities are a rare finding in these patients. TOF is a complex congenital heart defect that occurs in approximately 3.9-7.7 per 10,000 live births. Aortic arch abnormalities are rare in TOF patients, with a reported incidence of less than 1%. However, when present, they can impact surgical management and long-term outcomes.

Aortic arch abnormalities involve malformations of the aortic arch and its branches that can lead to various cardiovascular complications. Normally, the aorta originates from the left ventricle and gives rise to the aortic arch, which curves anteriorly and to the left before descending as the thoracic and abdominal aorta. The aortic arch branches into three major vessels, the brachiocephalic artery, the left common carotid artery and the left subclavian artery [7].

From an embryological perspective, the vertebral artery is an important vessel that arises as a secondary development, on each side of the midplane, from a series of dorsal rami of dorsal intersegmental arteries belonging to the neck. These rami undergo longitudinal linkage just dorsal to the ribs (post costal anastomosis). All of the original stalks then atrophy except the most caudal one in the series. The resulting longitudinal vessel is the vertebral artery; it takes origin, along with the subclavian from the seventh intersegmental artery. The seventh cervical intersegmental continues as the left subclavian and hence as the distal part of the right [1].

A left vertebral artery of aortic origin may be because of the persistence of the dorsal division of the left sixth intersegmental as the first part of the vertebral artery instead of that of the left seventh intersegmental artery [2].

The origin of the left vertebral artery from the arch of aorta has been documented by different authors with a range of 3.1%–8.3% [3].The right vertebral artery may arise from the first part of the subclavian, nearer than normal to the brachiocephalic (1% of cases) or to the anterior scalene muscle,directly from the arch of aorta (3% of cases), from the right common carotid, when the right subclavian arises from the aorta beyond the left subclavian, or from the brachiocephalic trunk [3].

Aortic arch abnormalities can result from various embryological abnormalities, including abnormal migration of neural crest cells, which contribute to the development of the aortic arch and its branches. Other factors that may contribute to aortic arch abnormalities include genetic mutations and environmental factors. However, the exact cause of aortic arch abnormalities in TOF patients is not well understood [8].

Patients with TOF have varying degrees of cyanosis depending on the severity of right ventricular outflow tract stenosis and pulmonary artery anatomy. In the majority of cases of TOF, the heart disease is clinically well tolerated. If the pulmonary arteries are of adequate size and the pulmonary obstruction is mild, oxygen saturation may be over 90%. In our case, the child complained of easy fatigability compared to peers, dyspnea on normal exertion and refusal to feed for 10 days. However, there was no evidence of cyanosis.

The physiological process surrounding the hypercyanotic episodes or “Tet spells” in tetralogy of Fallot consists of either a decrease in systemic vascular resistance or an increase in pulmonary resistance contributing to a right-to-left shunt across the ventricular septal defect, causing marked desaturation [9,11].

Echocardiography is the first line investigation to diagnose and monitor TOF, However, it plays a limited role in assessing extra-cardiac vessels. Complementary imaging is justified in a patient in whom some clinical or echocardiography findings are missing in planning for the decision on treatment. Cardiac MRI fulfils this role without irradiation and provides both anatomical and functional information. Despite all of its advantages and technological advances, MRI has certain limitations [12]. Apnea images cannot be obtained in infants and sedation (general anesthesia) is often required. In addition the images available do not have the necessary spatial resolution to assess small anatomical structures. The tracheobronchial tree is also better assessed by CT than MRI [13]. Furthermore, as the investigation time is longer than for cardio-thoracic CT, this may represent a limiting factor in patients who are hemodynamically unstable and/or who require a doctor at the bedside.

Previously, invasive angiographic techniques were considered the gold standard for the assessment of large arterial abnormalities. However, the complexities and complications inherent to invasive imaging have meant that more recently non-invasive techniques such

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as echocardiography, Magnetic Resonance Imaging (MRI) and Multi Detector CT (MDCT) have been increasingly used in congenital cardiovascular disorders. MDCT is now a fundamental tool for the diagnosis and pre-surgical work-up of aortic abnormalities due to its high spatial resolution, large area of coverage and short scan time.

Surgical correction is the mainstay of management of TOF and is performed during the first year of life, ideally before eleven months of age. It involves closing the VSD and broadening the pulmonary outflow tract using a trans-annular or infundibular patch with pulmonary valvuloplasty. This reduces the hypoxia time, risk of death and risk of "Tet spells". The prognosis of patients who have undergone surgical correction is excellent, with an early mortality rate of <2%. In our case, an intra cardiac repair was performed with a short trans-annular pericardial patch. Sauvage patch closure of VSD was done with 5-O continuous prolene sutures followed by reconstruction of the RVOT by augmenting it with a pericardial patch.

Conclusion

In conclusion, aortic arch abnormalities are an incidental finding that are likely to go unnoticed during angiographic or interventional cardiology examination. Therefore, it is essential for radiologists to be vigilant during interpretation of imaging.

Ultimately, a thorough knowledge of the vertebra basilar changes associated with individual vertebral arteries will aid in the interpretation of images and the results of skull base and other head and neck surgeries. Advances in technology has increased our knowledge of the many variations in the human body and recognizing them can help prevent unwanted complications from various procedures.

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