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CASE REPORT

Rare Combination of Right Aortic Arch, Aberrant Left Subclavian Artery and Absent Left Pulmonary Artery in a Boy with Tetralogy of Fallot

Mohammed Nasir[1], Muluken Ahmed[2]

[1]Pediatrics Department, Hawassa University, Hawassa, Ethiopia; [2]Pediatrics Department, Arbaminch University, Arbaminch, Ethiopia

Correspondence: Mohammed Nasir, Pediatrics Department, Hawassa University, PO Box: 05, Hawassa, Ethiopia, Tel +251911739081, Email mn2572338@gmail.com

Introduction: Right aortic arch is associated with an aberrant subclavian artery and absence of left pulmonary artery in patients with Tetralogy of Fallot. However, the occurrence of a combination of the right aortic arch, an aberrant left subclavian artery, and an absent left pulmonary artery in a single patient is rare. Therefore, the purpose of this case report was to discuss the rare association between a right aortic arch, the absence of a left pulmonary artery, and an aberrant left subclavian artery in an 8-year-old Ethiopian boy with Tetralogy of Fallot.

Case Presentation: An 8-year-old male child who presented with easy fatigability, dizziness, and intermittent difficulty in swallowing solid food over the past two years. He had grade clubbing, cyanosis of the lips, and an ejection systolic murmur at the left lower sternal border. Tetralogy of Fallot with absent left pulmonary and right aortic lesions was diagnosed using echocardiography. CT tomography revealed a detailed vascular anatomy and confirmed the diagnosis of an aberrant left subclavian artery.

Conclusion: In the evaluation of patients with Tetralogy of Fallot, it is important to pay close attention to vascular abnormalities such as right aortic arch, aberrant left subclavian artery, and absent left pulmonary artery for not missing them.

Keywords: Tetralogy of Fallot, right aortic arch, aberrant left subclavian artery, absent left pulmonary artery

Introduction

A right aortic arch with an aberrant left subclavian artery, a right aortic arch with mirror image branching and varied branching patterns, or a right aortic arch with an isolated left subclavian artery occurs in 13–34% of patients with Tetralogy of Fallot (TOF).[1] In half of the cases, the right aortic arch was associated with an aberrant left subclavian artery.[2] The right aortic arch is also associated with the absent of left pulmonary artery (LPA).[3]

However, to our knowledge, there have been no reports of a combination of the right aortic arch, absent left pulmonary artery, and aberrant left subclavian artery in the same patient. The purpose of this case report is to discuss the rare association between a right aortic arch, the absence of a left pulmonary artery, and an aberrant left subclavian artery in an 8-year-old Ethiopian boy.

Case Presentation

An 8-year-old child presented with easy fatigability and intermittent difficulty swallowing solid food over the past two years. The patient experienced dizziness. During a visit to the cardiac clinic of Ethiopia, a physical examination revealed the following vital signs: body temperature, 370°C; blood pressure, 90/40 mmHg; and pulse rate, 104 beats per minute. He had blue lips and fingers. He also had grade 1 clubbing, with a systolic ejection murmur at the left sternal junction. Echocardiography revealed severe pulmonary valvular stenosis with right ventricular dilatation, subaortic ventricular septal defect (VSD), and the absence of the left pulmonary artery (Figures 1 and 2). A chest CT scan revealed that the main pulmonary artery (MPA) was 103 mm in size and the right pulmonary artery (RPA) was 101 mm. The RPA size divided by the body surface area (BSA) was used to calculate the

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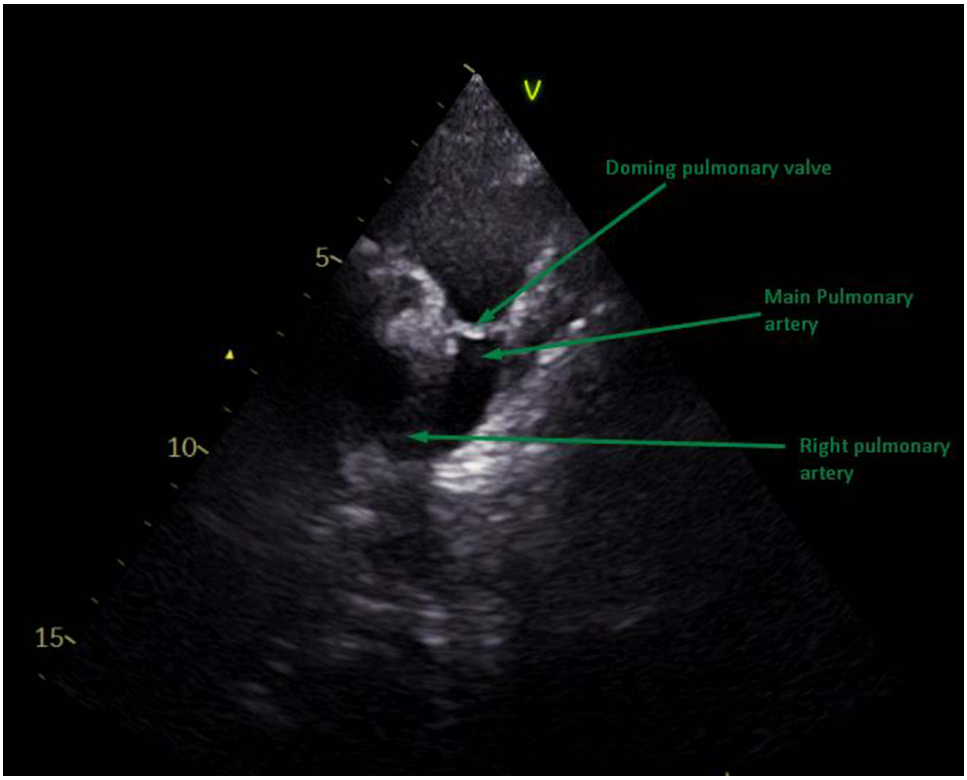

Figure 1 Echocardiography showing continuation of main pulmonary artery as right pulmonary artery and absence of left pulmonary artery.

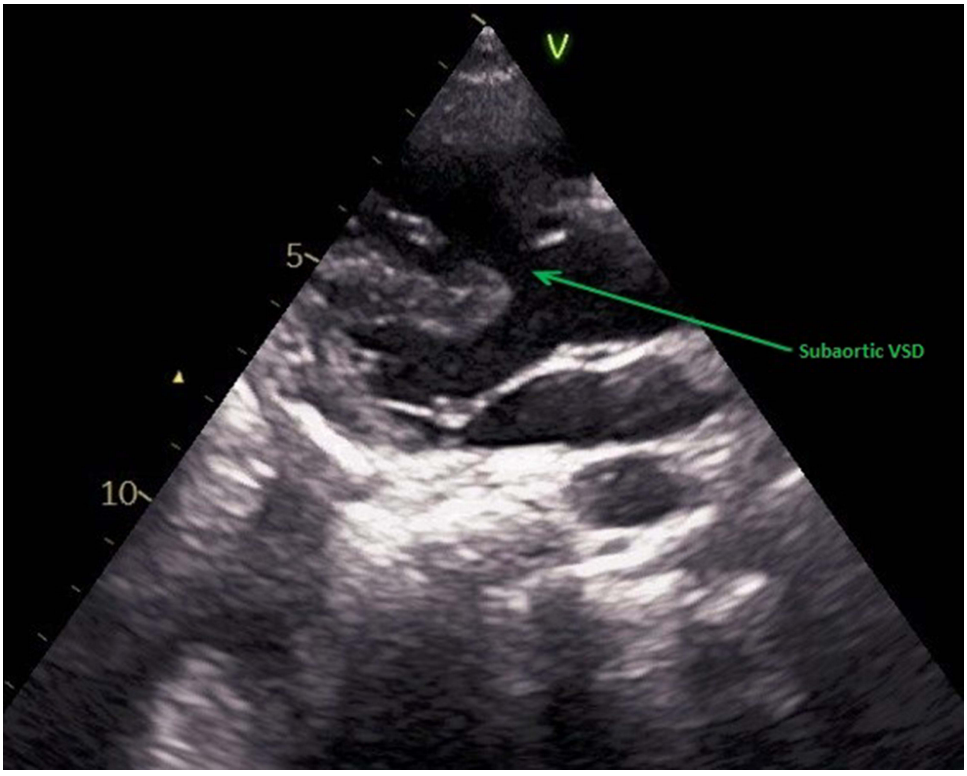

Figure 2 Echocardiography showing large subaortic VSD.

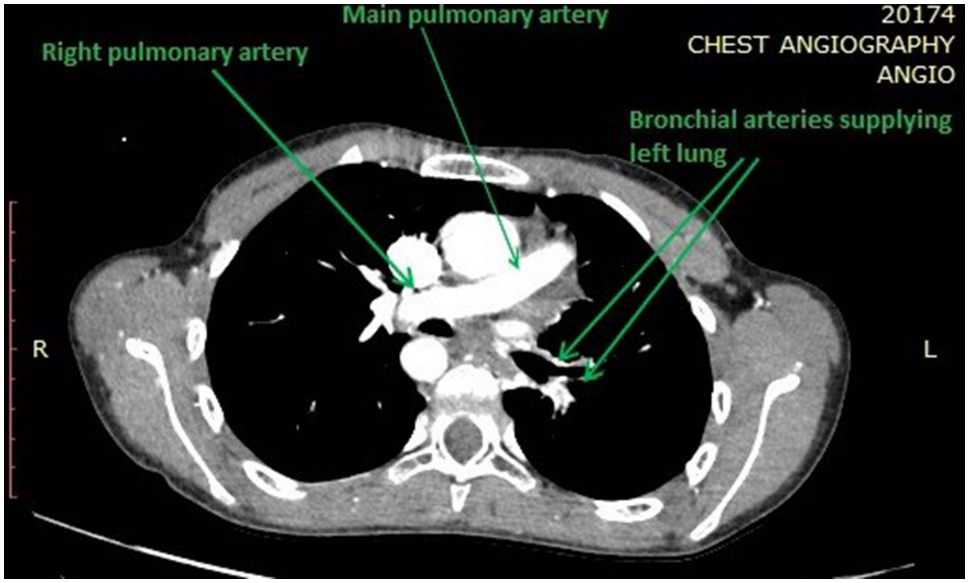

Figure 3 Chest CT scan showing continuation of main pulmonary artery as right pulmonary artery and absence of left pulmonary artery and left lung supplied by collaterals.

Nakata index, which was 134 mm^2/m^2 (Figure 3). An additional diagnosis of an aberrant left subclavian artery with the right arch was made via CT scan (Figure 4A and B).

Discussion

The right aortic arch is a rare congenital anomaly that occurs in approximately 0.1% of both fetuses and is defined by the position of the arch on the right side of the trachea.[4] This developmental anomaly occurs between weeks 4 and 5 of embryogenesis, when the six pairs of aortic arch vessels that connect the ventral aorta to both dorsal aortae begin to migrate and form their respective structures.[5] The right arch is formed when the right fourth arch transforms into the right aortic arch, and the right dorsal aorta remains as the thoracic aorta with retraction of the left dorsal aorta.[5]

It is associated with TOF, and 13–34% of patients with TOF have a right arch.[1] Additionally, 0.04–0.1% of patients with a right aortic arch have an abnormal left subclavian artery in autopsy specimens, and radiological examination revealed almost the same proportion, 0.05%.[6] Furthermore, there are also a few reports of its association with a rare abnormality: the absence of the left pulmonary artery.[7] Both the right arch and the absent left pulmonary artery are associated with Tetralogy of Fallot.[1,8]

Patients with a right aortic arch and abnormal left subclavian artery are often asymptomatic. However, symptoms of compression of adjacent structures such as the esophagus and trachea rarely occur.[2,9] Furthermore, if the left pulmonary artery is absent, the patient may experience respiratory issues due to derangement of pulmonary function and a high prevalence of pulmonary hypertension in individuals with a unilaterally absent pulmonary artery.[10]

Chest radiography is helpful for assessing the right aortic arch, whereas barium enema may be used to detect esophageal compression.[2] The diagnosis of an anomalous left subclavian artery and an absent LPA is established by a CT scan, MRI, or 2D echocardiography with color Doppler.[9,11] When looking for patent ductus arteriosus (PDA) or collaterals associated with an absent LPA, the aforementioned investigative modalities are also crucial.[12]

Surgical intervention is required in patients with a right aortic arch and an aberrant subclavian artery.[1] There is still no consensus on the treatment of unilateral absent LPA. Some advice observing asymptomatic patients, giving them a vasodilator if they have pulmonary hypertension, and using aggressive management techniques such as embolization, lobectomy, and pneumonectomy if hemoptysis occurs.[13]

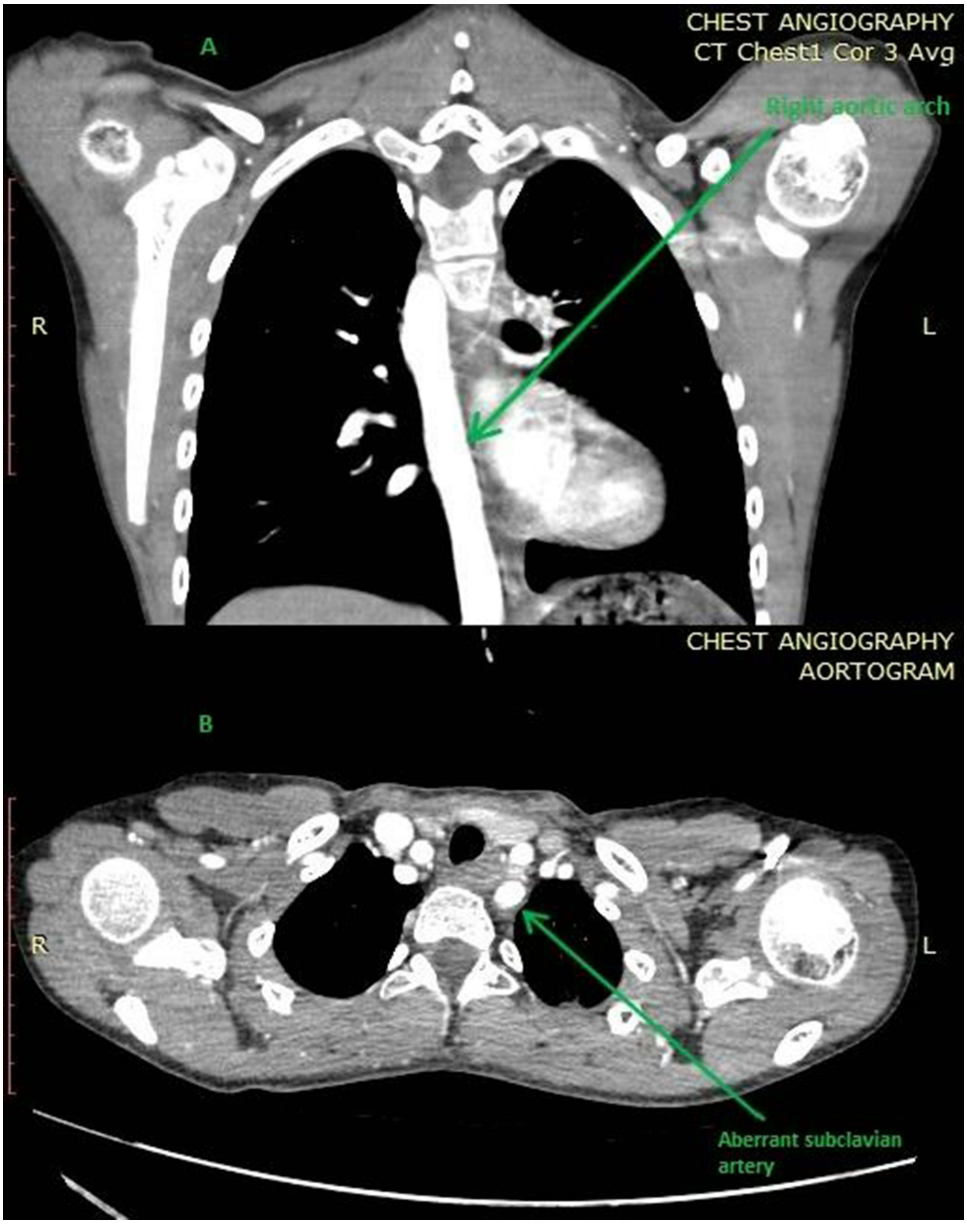

Figure 4 Chest CT showing right aortic arch (**A**) and aberrant origin left subclavian artery (**B**).

Conclusion

In the evaluation of patients with Tetralogy of Fallot, it is important to pay close attention to vascular abnormalities such as right aortic arch, aberrant left subclavian artery, and absent left pulmonary artery for not missing them. A detailed assessment of the aortic arch is crucial for the complete diagnostic of congenital heart disease because the complex anatomy may influence the surgical decision.

Data Sharing Statement

The datasets used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Ethical Clearance

Ethical clearance was obtained from Saint Paul's Hospital Millennium Medical College Research Ethics Review Board, Addis Ababa, Ethiopia.

Consent

Verbal informed consent was obtained from both parents and the child to use their patients' medical information and images for publication. The consent was verified by a research ethics review board.

Acknowledgment

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Disclosure

The authors declare no conflicts of interest in this work.

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