An adult with central cyanosis and differential pulmonary vascularity

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ABSTRACT

A 22-year-old male patient presented with progressive effort intolerance of 2-years duration. Clinical findings and investigations were suggestive of Tetralogy of Fallot (TOF). In addition, there was a conspicuous difference in the pulmonary vascularity with oligemia on the left side and relative hypervascularity on the right side. The right pulmonary artery was arising from the proximal ascending aorta and the main pulmonary artery was continuing as the left pulmonary artery. The anomalous origin of a branch pulmonary artery from the aorta (AOPA) is a rare cardiac anomaly. We report this condition in association with TOF, highlighting the differential pulmonary vascularity.

Keywords: Anomalous origin, congenital heart disease, pulmonary vascularity, pulmonary artery

A 22-year-old male patient presented with progressive effort intolerance of 2-years duration. He was detected to have congenital heart disease during childhood with onset of cyanosis at the age of 5 years. Physical examination revealed central cyanosis (resting saturation: 76%) and clubbing. There was a short systolic murmur at the left upper sternal border and second heart sound was single. A clinical diagnosis of Tetralogy of Fallot (TOF) was considered.

Electrocardiogram showed right axis deviation and right ventricular hypertrophy. Chest X-ray was remarkable for the conspicuous difference in the pulmonary vascularity. There was pulmonary oligemia on the left side, but relative hypervascularity on the right side [Figure 1]. Echocardiography confirmed the clinical suspicion of TOF. The right pulmonary artery could not be delineated on echocardiography. Computed tomography (CT) angiography illustrated the origin of right pulmonary artery from the posterior aspect of proximal ascending aorta [Figure 2]. The main pulmonary artery was continuing as left pulmonary artery. Cardiac catheterization data was consistent with TOF [Table 1 and Videos 1 and 2]. The origin of right pulmonary artery from ascending aorta was illustrated [Figure 3]. The pressure in the right pulmonary artery was similar to aortic pressure with a pulmonary vascular resistance index (PVRI) of 18 wood units. Patient was advised medical management, in view of pulmonary vascular obstructive disease in right lung.

The anomalous origin of a branch pulmonary artery from the aorta (AOPA) is a rare cardiac anomaly, accounting

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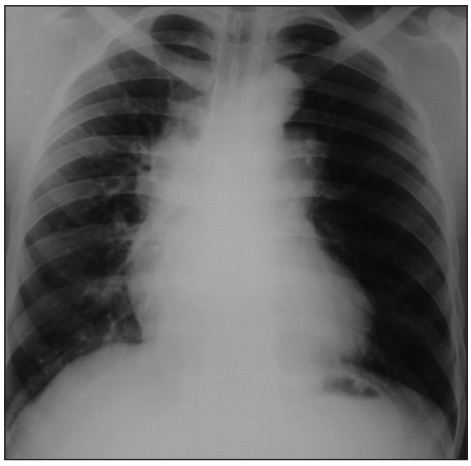

Figure 1: Chest X-ray in postero-anterior view showing a boot-shaped heart. There is pulmonary oligemia on the left side and increased pulmonary flow on right side

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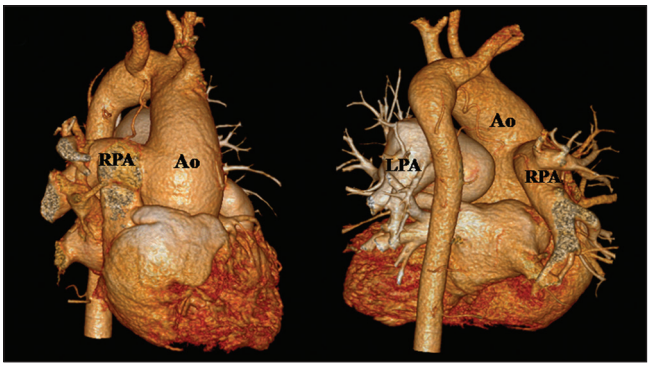

Figure 2: Volume rendered computed tomography depicting the origin of right pulmonary artery from the posterior aspect of ascending aorta; AO: Aorta, RPA: Right pulmonary artery, LPA: Left pulmonary artery

Table 1: Cardiac catheterization data

Chamber	Saturation (%)	Pressure (mmHg)
IVC	65	—
SVC	63	—
RA	64	a4v2 (3)
RV	65	134/6 (9)
MPA	67	20/8 (12)
RPA	81	130/70 (90)
LPA	67	20/8 (12)
LA	98	a8v12 (10)
LV	97	140/7 (10)
AO	83	136/72 (93)

(*Body surface area: 1.64 m^2; † PVRI for right lung: 18 Wood units); AO: Aorta; RPA: Right pulmonary artery; LPA: Left pulmonary artery; IVC: Inferior venacava; SVC: Superior venacava; RA: Right atrium; RV: Right ventricle; MPA: Main pulmonary artery; LA: Left atrium; LV: Left ventricle

for 0.1% of all congenital heart diseases.[1] This developmental defect results from abnormal migration of pluripotent cells. Defective migration of the right sixth aortic arch to the left side results in anomalous origin of right pulmonary artery from ascending aorta.[2] AOPA is commonly associated with DiGeorge syndrome, TOF, aortopulmonary window, patent ductus arteriousus, atrial septal defect, and interrupted aortic arch. Rarely, it may occur in isolation. In a study by Gan HL, cases of AOPA were reported between 60 days–23 years.[3] AOPA should always be suspected in the presence of differential pulmonary vascularity. The important differential diagnosis on chest X-ray is a congenital absence of a pulmonary artery. In AOPA, pulmonary vascular obstructive disease develops rapidly. In the absence of surgical intervention, mortality is as high as 70% before 6 months of age.[4] Hence, early recognition and establishment of vascular continuity between main pulmonary artery and branches on the affected side

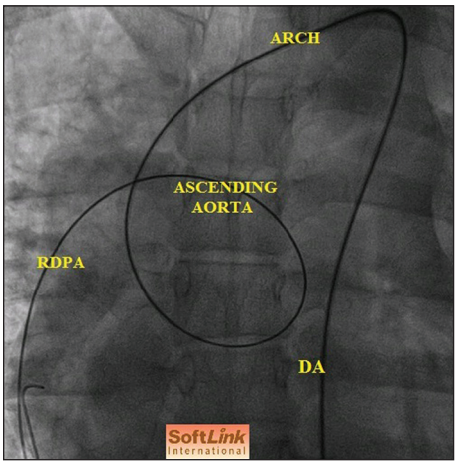

Figure 3: Fluoroscopy highlighting the catheter course and origin of right pulmonary artery from ascending aorta: Descending thoracic aorta > Left sided aortic arch > Ascending aorta > Right pulmonary artery; DA: Descending thoracic aorta; RDPA: Right descending pulmonary artery

is vital, and reduce the likelihood of complications. There are various surgical options for the treatment of AOPA. It consists of surgical division of the anomalously connected pulmonary artery branch and anastomosis directly, or with a graft to the main pulmonary artery.

To conclude, AOPA is a rare congenital cardiac malformation. We report this condition in association with TOF, highlighting the differential pulmonary vascularity.

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