Case Report

DOI: 10.7860/IJARS/2022/53180.2774

Radiology Section

Pulmonary Artery Aneurysm with Tetralogy of Fallot: A Rare Entity

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ABSTRACT

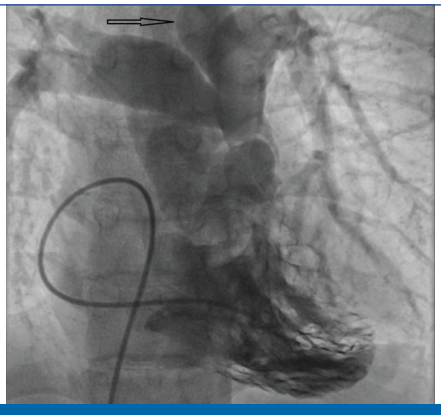
Pulmonary artery aneurysms is a rare entity with varied causes, its association with Tetralogy of Fallot (TOF) having low pressure pulmonary system is further more uncommon. Here, the authors present a case of 15-year-old male patient presenting with symptoms of cyanosis and dyspynoea for which echocardiography, cardiac catheterisation and Computed Tomography (CT) angiography was done and diagnosis of tetralogy of fallot with left pulmonary artery aneurysm was made for which patient was admitted and underwent surgical correction. Pulmonary artery aneurysm with tetralogy of fallot is an extremely rare anatomical combination requiring treatment strategy modification, hence merit proper evaluation.

Keywords: Computed tomography, Cyanotic congenital heart disease, Echocardiography, Increased outflow of aorta, Ventricular septal defect

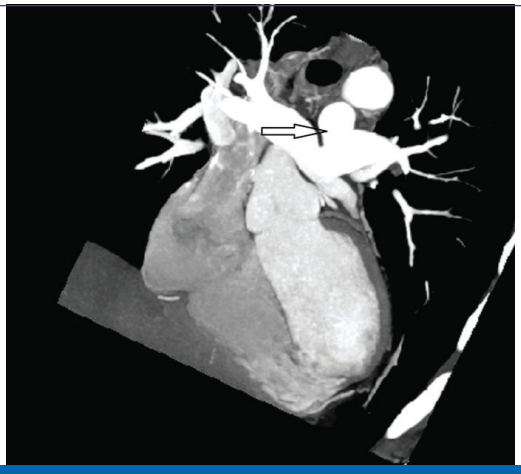
CASE REPORT

A 15-year-old male presented with complaints of dyspynoea and cyanosis for last 4-5 years. There was no evidence of heart failure and was clinically in New York Heart Association (NYHA) class II [1]. There was no history of trauma, infection or any evidence of collagen vascular disease. Chest X-ray was suggestive of mild increased cardiothoracic ratio with coeur-en-sabot appearance with oligemic bilateral lung fields. On electrocardiogram there was right axis deviation with right ventricular hypertrophy.

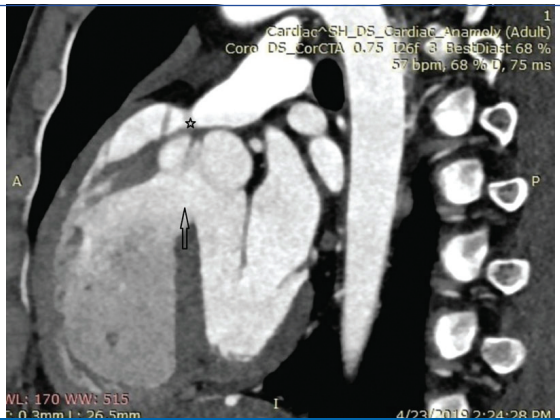
Transthoracic echocardiography was done which suggested tetralogy of fallot physiology including sub aortic ventricular septal defect, overriding of aorta, right ventricular hypertrophy and severe infundibular and pulmonary valvular stenosis (peak gradient-100 mmHg) with good confluent pulmonary arteries. Cardiac catheterisation was performed which revealed similar findings with normal coronaries, no aortic regurgitation and non significant aortopulmonary collaterals but a definite outpouching from left pulmonary artery of size around 1.5×2 cm was observed [Table/Fig-1]. Spiral CT imaging was done for morphological assessment revealed situs solitus, atrioventricular concordance, ventriculo-arterial concordance, left sided aortic arch, subaortic Ventricular Septal Defect (VSD) measuring approx 12.7 mm. Right Ventricular (RV) hypertrophy, 50% overriding of aorta, infundibular stenosis with dilated bronchial circulation and confirmed the diagnosis of associated left pulmonary aneurysm present at the site of patent ductus arteriosus measuring approx 1.5×2 cm [Table/Fig-2,3].



[Table/Fig-1]: Cardiac catheterisation image with catheter in right ventricle showing mild pulmonary stenosis and saccular aneurysm form left pulmonary artery (arrow).



[Table/Fig-2]: Reconstructed coronal multiplanal reformation shows a blind ending saccular outpouching from left pulmonary artery (arrow).



[Table/Fig-3]: Reconstructed saggital image shows four components of TOF sub-aortic VSD (arrow), overriding of aorta, pulmonary stenosis (star) and RV hypertrophy.

So, after establishing the diagnosis of tetralogy of fallot with left pulmonary artery aneurysm plan for surgical correction was made.

A standard median sternotomy was performed and pericardium was reflected towards the left side and cardiopulmonary bypass was established after aortobicaval cannulation. Patient was cooled down to 28°C and delnido's cardioplegia was delivered after aortic cross clamp. Right atrium was opened, anatomy of ventricular septal defect was observed and infundibular resection was done from right

atrial side. Main pulmonary artery was opened and incision was extended distally till the confluence and proximally till 2 cm below annulus. Transpulmonary artery release of supravalvular teethering, pulmonary commisurotomy and infundibular resection was performed. Left pulmonary artery aneurysm opening was seen which was present at the site of patent ductus arteriosus insertion of size of around 2×1.5 cm which was closed with polytetrafluoroethylene patch using interrupted prolene 5-0 sutures. Ventricular septal defect was closed with dacron patch using interrupted pledgeted prolene 5-0 sutures and trans annulur patch was done with autologous unfixed pericardial patch. Patient were slowly weaned off and peak trans annular gradient was measured by direct needle prick method which came out to be 36 mmHg. Haemostasis achieved and routine closure was done and patient was shifted in Intensive Care Unit (ICU) in stable condition with minimal inotropic support of 5 mcg/kg/min of dopamine and dobutamine. Patient was discharged from hospital on 6th postoperative day in stable condition and was followed-up after a week with fresh echocardiography and chest X-ray which were uneventful.

DISCUSSION

Pulmonary Artery Aneurysms (PAAs) with tetralogy of fallot is an extremely rare entity with only four cases have been reported so far in the literature. All the cases were of middle age with mostly the aneurysm involving the main pulmonary trunk till its bifurcation [2-5]. Afrikh K et al., also reported a similar case but there the whole of the left pulmonary trunk was dilated, whereas in the present case there is a saccular outpouching from proximal left pulmonary artery at the site of patent ductus arteriosus origin [2].

In 66% of cases PAAs are seen with pulmonary arterial hypertension [6], thus, it is mostly associated with congenital heart diseases such as patent ductus arteriosus, ventricular septal defects and atrial septal defects that cause volume and pressure overloading right cardiac chambers and pulmonary arteries [7]. This itself signifies its rare association with tetralogy of fallot. PAAs are also associated with pathologies that cause inflammatory changes in vessel wall like mycosis, syphilis, tuberculosis, Behcet's disease and trauma [8]. Isolated PAAs, although very rare but also known to exist.

These aneurysms are usually divided into proximal (central) PAAs and peripheral PAAs. Proximal PAAs involve main pulmonary artery and right and left pulmonary artery, beyond that comes under peripheral PAAs. Central PAAs are usually asymptomatic with symptoms are related to complications such as bronchial or tracheal compression, dissection, rupture or thrombus causing unexplained dyspynoea [9]. Peripheral PAAs can be life threatening as they can lead to massive haemoptysis in cases of rupture with even unruptured cases had a high mortality rate [10].

In the past pulmonary artery angiography used to be the gold standard in diagnosis of PAAs but now spiral CT angiography is considered as the most useful tool in its diagnosis, as it has good temporal and spatial resolution like in the present case [11]. Images are acquired from thoracic inlet to L1 vertebrae following which base images and various image reformatting techniques were used to systematically evaluate the situs, aorta, pulmonary artery, pulmonary veins, cardiac chambers, atrioventricular concordance, ventriculo-arterial concordance, coronaries, tracheo-bronchial anatomy and associated anamolies [12]. Treatment of these PAAs can be conservative or surgical. Surgical repairs are recommended when aneurysms are of large size >6 cm or if they are symptomatic, regardless of the size, as the risk of rupture or dissection are high in such cases [13]. Surgical techniques include aneurysmorrhaphy or arterioplasty, pericardial patch reconstruction, interposition grafting with allograft or synthetic textile grafts and closing the opening of aneurysm with pericardial or synthetic textile patch, as was done in this case [14]. Peripheral PAAs in the past were treated with aneurysmectomy or lobectomy but presently endovascular coil embolisation is the preferred method of choice, as it is less invasive and has fewer complications [15]. The guidelines for intervention in PAAs are not clarified as the natural history of these aneurysm remains largely unknown.

CONCLUSION(S)

Association of PAAs with tetralogy of fallot with decreased pulmonary blood flow is extremely rare as in this case. It further requires a composite diagnostic approach and a planned surgical technique for a proper management.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Nov 10, 2021
- Manual Googling: Jan 15, 2022
- iThenticate Software: Mar 09, 2022 (11%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Nov 07, 2021**
Date of Peer Review: **Jan 06, 2022**
Date of Acceptance: **Jan 15, 2022**
Date of Publishing: **Apr 01, 2022**