Type-I right pulmonary artery to left atrial fistula

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Section: Cardiovascular
Area of Interest: Cardiovascular system Paediatric
Imaging Technique: CT-Angiography
Special Focus: Aneurysms Arteriovenous malformations
Case Type: Clinical Cases
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Patient: 12 years, male

Clinical History:
11-year-old male patient, admitted into the coronary care unit (CCU) with recurrent fever, vertigo and exertional dyspnoea. On admission, blood pressure was 80/50 mmHg, heart rate 120 beats per minute and oxygen saturation 87% in room air. Cardiopulmonary auscultation was normal. Other systemic examinations were normal. Haemoglobin level was 20 mg/dL.

Imaging Findings:
Contrast echocardiography(bubble): Ejection fraction 60%, otherwise no abnormal findings.
CT pulmonary angiography: A pear-shaped pouch (5x4x4cm) is noted arising from left atrium and invaginating into medial surface of lower lobe of right lung. No vascular branches / tributaries of the pouch can be identified. All four pulmonary veins drain into left atrium as normal. Right-sided pulmonary veins are significantly narrower (5 mm) than left (10 mm). Pulmonary trunk arises from right ventricle as normal. However, entire pulmonary trunk is engorged (32 mm), as well as right pulmonary artery (23 mm) and its descending branch (15 mm). Superior branch of RPA as well as LPA and its branches are normal in calibre. Descending branch of RPA forms a fistulous communication with left atrial aneurysm described above. However, branching pattern of RPA is normal, with normal vascularisation of entire right lung.

Discussion:
Abnormal communication (fistula) between right pulmonary artery & left atrium (RPA-LA) occurs rarely [1], and is distinct from pulmonary AVM [2]. Mostly congenital in origin, it can also be post-traumatic [1,3]. Due to its rarity and diagnostic complexity, only about a hundred cases exist in the literature [1,4], and its true prevalence is not documented [1]. Classification is into four types, based on pulmonary veins configuration [1]. Surgical correction is curative in most cases and early intervention is advised [1,2]. RPA-LA fistulae can present at any age, with 3:1 male predominance [1]. 70% are diagnosed before twenty and about 20% before ten years of age [1]. Symptoms are centred around cyanosis and exertional dyspnoea. Direct RPA-LA fistula produces persistent central cyanosis [2], whereas aneurysmal fistula (like the present case) produces intermittent cyanosis. Slight systolic or continuous murmur may be present [5,6]. ECG findings are normal or non-specific [2]. Other findings include
clubbing, arterial hypoxia and polycythaemia. Severity of symptoms depends on the magnitude of the right-to-left shunt. Larger shunts present in early neonatal period with severe heart failure. Untreated large shunts progress to pulmonary oedema & hypertension, cardiac failure and death. Moderate shunt fistulae may produce emboli, resulting in infarcts or abscesses elsewhere. In type-I cases, the aneurysm may suffer catastrophic rupture [1,5]. Clinical diagnosis of RPA-LA fistula is very challenging, since it is not the first or commonest cause of the above symptoms. Chest X-ray demonstrates oligaemic lung fields with left-sided cardiomegaly in extreme cases [1]. A very subtle crescentic right paracardiac opacity is present in all cases [1,2,4,7]. Echocardiography is often unable to demonstrate RPA-LA fistula or the associated aneurysm [1]. Pulsed Doppler shows diffuse turbulence spanning over left atrium and the aneurysm upto RPA [1,8]. Contrast echocardiogram shows early filling (within 3-5 beats) of the aneurysm, and is essential for diagnosis [1]. However, echocardiography may sometimes show only turbulence & shunt but fail to detect the anomaly [9]. Selective catheter angiography is confirmatory and gold standard [1,2]. But it is an invasive procedure that may not be available or feasible in every situation, producing delay in diagnosis, decision-making and surgical planning. CT angiography is a more widely available tool that provides accurate non-invasive diagnosis, and early diagnosis means correct treatment before it is too late [10]. CT / catheter angiography clearly demonstrates the fistulous communication, which most often arises from the posterior wall of the descending branch of RPA, and inserts into the LA. Both right & left pulmonary veins are of normal configuration in type-I fistula. PDA is identified in 70% of cases of neonatal presentation. Accurate anatomical identification facilitates choice of proper incision approach and surgical technique. Cardiopulmonary bypass may be required if dissection & ligation of the aneurysm is technically difficult [4,11]. Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Type-I fistula between descending branch of right pulmonary artery and left atrium, Tricuspid atresia, Hypoplastic right ventricle, Atrial septal defect with right-to-left shunt, Unroofed coronary sinus

**Final Diagnosis:** Type-I fistula between descending branch of right pulmonary artery and left atrium

**References:**


Description: Post-contrast axial MPR image showing large aneurysmal sac arising from left atrium, invaginating into mediastinal surface of right lung. Origin: Dept of Radiology & Imaging, Al Haramain Hospital Pvt Ltd, Sylhet, Bangladesh (www.haramainhospital.com). 2019.
Figure 2

Description: Post-contrast coronal MPR images showing all four pulmonary veins entering left atrium at normal position. (right) Origin: Dept of Radiology & Imaging, Al Haramain Hospital Pvt Ltd, Sylhet, Bangladesh (www.haramainhospital.com). 2019.
Description: Post-contrast coronal MPR images showing all four pulmonary veins entering left atrium at normal position. (left) Origin: Dept of Radiology & Imaging, Al Haramain Hospital Pvt Ltd, Sylhet, Bangladesh (www.haramainhospital.com). 2019.
Description: Post-contrast coronal MIP image showing that right-sided pulmonary veins are significantly narrower in calibre than left. Origin: Dept of Radiology & Imaging, Al Haramain Hospital Pvt Ltd, Sylhet, Bangladesh (www.haramainhospital.com). 2019.
Description: 3D reconstruction of CTPA showing all findings in one image: Left atrial aneurysm connecting with descending branch of RPA. All four pulmonary veins insert into LA at normal position. Right-sided pulmonary veins are narrower in diameter than left. Origin: Dept of Radiology & Imaging, Al Haramain Hospital Pvt Ltd, Sylhet, Bangladesh (www.haramainhospital.com). 2019.