Case 14791

Isolated unilateral absent pulmonary artery - A case report
Published on 10.07.2017

DOI: 10.1594/EURORAD/CASE.14791
ISSN: 1563-4086
Section: Chest imaging
Area of Interest: Cardiovascular system Pulmonary vessels Salivary glands Vascular Lung
Procedure: Diagnostic procedure
Procedure: Computer Applications-3D
Imaging Technique: CT
Imaging Technique: CT-Angiography
Special Focus: Pathology Dilatation Case Type: Clinical Cases
Authors: Jamshid Sadiqi, Hidayatullah Hamidi, Abdul Basir Shahin
Patient: 23 years, female

Clinical History:

A 23-year-old female patient with complaints of chest pain and dyspnoea during exercise for one year, which did not respond well to treatment, was suspected for cardiovascular congenital abnormality and referred to radiology department of French Medical Institute for Mothers and Children for contrast-enhanced chest CT scan.

Imaging Findings:

Chest CT scan with IV contrast was performed for the patient. Dilatation of the right atrium was noted (Fig. 1). Dilated pulmonary trunk was seen compared to the ascending aorta in the axial mediastinal window (Fig. 2). MIP and colour LUT images showed absent left pulmonary artery with evidence of abrupt cut-off in proximal part while right pulmonary artery and its branches existed (Fig. 3a-d). Prominent enhanced vessels were noted along the left side of thoracic vertebrae and descending aorta representing collateral vessels (Fig. 4a-c). Volume-rendered images clearly showed absent left pulmonary artery and normal right pulmonary artery and its branches (Fig. 5a-b).

Discussion:

Isolated unilateral absent pulmonary artery (IUAPA) is a rare congenital anomaly. The broader category is UAPA which is accompanied by other congenital heart anomalies like tetralogy of Fallot, cardiac septal defects, coarctation of the aorta and sub-valvular aortic stenosis. The prevalence of UAPA is 1/200000 in population with no sex predilection [1]. The anomaly was diagnosed for the first time in 1868 [2]. Failure of migration and rotation of the sixth aortic arch segment results in absence of the pulmonary artery [1]. UAPA is commonly seen in the site of the chest opposite the aortic arch with only 2% occurring in the same site of the aortic arch [3] as it was in our case. The distal branches of the pulmonary artery remain intact by getting blood from collateral vessels of bronchial, internal mammary, subclavian, sub-diaphragmatic, intercostal and even coronary arteries [2-4]. The clinical features of UAPA are chest pain, pulmonary oedema, dyspnoea during exercise, haemoptysis, pleural effusion, recurrent pulmonary infection, bronchiectasis and even death which is usually caused by severe pulmonary haemorrhage, right side heart failure and respiratory failure. Some of the cases are asymptomatic and detected incidentally [3-5]. One of the consequences of UAPA is pulmonary hypertension. Haemoptysis is another complication which can be serious sometimes, due to unusual high pressure in the collateral vessels [6]. The findings in a chest X-ray are
asymmetric lung fields with absent or diminished hilar vasculature, shifting of mediastinum towards the affected hemithorax and elevation of the hemidiaphragm in the affected side [1-5]. The definitive diagnosis of UAPA can be made by angiography, chest CT, MRI, echocardiography and nuclear imaging. Ventilation perfusion scan can be done to differentiate the pulmonary hypertension of IUAPA from chronic thromboembolic pulmonary hypertension [1-7]. Different treatment strategies exist, such as surgical, pharmacological and behavioural management. In recurrent haemoptysis, pulmonary infection and pulmonary hypertension pneumonectomy and surgical revascularisations are done. Selective embolisation of bronchial or non-bronchial systemic arteries is suitable for patients with massive haemoptysis who are not eligible for surgery. Pharmacological treatment is for patients not tolerating the revascularisation surgery or those who don't improve after surgery [8]. Our patient got medical treatment and advice. In conclusion, UAPA is a rare congenital problem that is usually diagnosed in adolescent and adult population and clinicians should keep in mind the possibility of undiagnosed cases. 

**Differential Diagnosis List:** Isolated unilateral absent pulmonary artery (IUAPA), Primary pulmonary hypertension, Swyer-James-McLeod syndrome, Pulmonary emboli, Chronic thromboembolic pulmonary hypertension

**Final Diagnosis:** Isolated unilateral absent pulmonary artery (IUAPA)

**References:**


**Description:** The axial section of chest CT in mediastinal window demonstrates dilatation of the right chambers of the heart prominently in the right atrium. **Origin:** Radiology Department, French Medical Institute for Mothers and Children, Kabul
Figure 2

Description: Axial section of chest in mediastinal window demonstrates dilated pulmonary trunk compared to the ascending aorta. Origin: Radiology Department, French Medical Institute for Mothers and Children, Kabul
Description: Coronal CT image shows collateral vessels along the left side of thoracic vertebrae and descending aorta. Origin: Radiology Department, French Medical Institute for Mothers and Children, Kabul
Description: Sagittal CT image shows collateral vessels along the left side of thoracic vertebrae and descending aorta. Origin: Radiology Department, French Medical Institute for Mothers and Children, Kabul
Description: Coronal MIP image shows collateral branches with no pulmonary arterial branches in the left side while normal enhancement of right pulmonary vessels are noted in the right hemithorax.

Origin: Radiology Department, French Medical Institute for Mothers and Children, Kabul
Description: The CT enhanced MIP section in coronal plane demonstrates dilated pulmonary trunk with normal enhancement of right pulmonary artery but evidence of abrupt cut-off in proximal part of left pulmonary artery. **Origin:** Radiology Department of French Medical Institute for Mothers and Children
Description: The CT colour LUT image in coronal plane shows dilated pulmonary trunk with normal right pulmonary artery but abrupt cut-off of left pulmonary artery. Origin: Radiology Department of French Medical Institute for Mothers and Children
Description: The CT enhanced MIP section in axial plane demonstrates dilated pulmonary trunk with normal enhancement of right pulmonary artery but evidence of abrupt cut-off in proximal part of left pulmonary artery. Origin: Radiology Department of French Medical Institute for Mothers and Children

Description: Sagittal MIP images demonstrate normal right pulmonary vessels enhancement (right image) and no enhancement of the left pulmonary vessels with abrupt cut-off of left main pulmonary artery (left image). Origin: Radiology Department of French Medical Institute for Mothers and Children
Description: Volume-rendered image in anterior view clearly demonstrates absent left pulmonary artery with normal right pulmonary vasculature. Origin: Radiology Department of French Medical Institute for Mothers and Children
Description: Left lateral volume-rendered image shows absent left pulmonary artery with abrupt cutoff, however, normal right pulmonary artery and its branches are normally seen. Origin: Radiology Department of French Medical Institute for Mothers and Children