ISSN: 2581 – 3633
PubMed - National Library of Medicine - ID: 101745081

International Journal of Medical Science and Advanced Clinical Research (IJMACR)

Available Online at:www.ijmacr.com

Volume – 6, Issue – 4, July - 2023, Page No. : 122 - 128

Fascinating case of unilateral absence of the pulmonary artery in a 56-year-old female

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How to citation this article: Jensy Jaison, Udaya Vakamudi, Siddharth Tewari, Rajeshwar Balaji, "Fascinating case of unilateral absence of the pulmonary artery in a 56-year-old female", IJMACR- July - 2023, Volume – 6, Issue - 4, P. No. 122 – 128.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Unilateral absence of the pulmonary artery (UAPA) is an uncommon developmental anomaly resulting in underdevelopment of the proximal portion of the pulmonary artery with preserved intrapulmonary segments. The prevalence of this disorder is estimated to

be 1 in 300,000. Most patients are usually detected incidentally. Common clinical presentation may be breathing difficulty, recurrent respiratory infection, and chest pain. We present an interesting case of an adult woman who was found to have unilateral absence of the pulmonary artery on left side with right sided aortic arch and double superior vena cava, with a brief review of the embryology and various differential diagnoses.

Keywords: UAPA, PIPA, Clinical.

Introduction

Unilateral absence of the pulmonary artery (UAPA) is an uncommon developmental anomaly resulting in underdevelopment of the proximal portion of the pulmonary artery with preservation of the intrapulmonary segments. It is also called proximal interruption of pulmonary artery (PIPA). [1] Clinical manifestation can range from incidental finding to severe haemoptysis. It is usually associated with cardiovascular anomalies, particularly on left side. [2] We present an interesting case of an adult woman who was found to have unilateral absence of the pulmonary artery on left side with right sided aortic arch and double superior vena cava

Case Report

A 56/F presented to the hospital with complaints of breathing difficulty. On examination, she had labored breathing with reduced exercise tolerance and reduced air entry in the left hemithorax . No history of chest pain or palpitations noted. There was no history of recurrent respiratory infections. She underwent a chest X ray (**Figure 1**) which showed reticular opacities in the left lung with gross reduction in the volume of the left hemithorax. Tracheal shift to the left was noted. There was a relatively well-defined opacity in the left hemithorax (subcarinal) with broad base towards the mediastinum. Right sided aortic arch was noted. CT Thorax was advised to rule out the possibility of a mediastinal mass.Lung window showed reticular opacities with interlobular and intra-lobular septal thickening and in the left lung parenchyma, predominantly in sub-pleural and peri-bronchovascular distribution causing architectural distortion and volume loss with ipsilateral mediastinal shift **(Figure 2)**.Peripheral areas of honeycombing was noted in upper and lower lobe of left lung. Right sided aortic arch and double SVC were noted. Left pulmonary artery was not visualized. CT Pulmonary angiogram (**Fig 3A**) was suggested for further evaluation which showed that the left pulmonary artery was completely not visualized from its origin. Right sided aortic arch with aberrant left subclavian artery were also noted . Arterial supply to left lung was seen to arise from the aberrant left subclavian artery beyond the origin of left vertebral artery**(Figure 3B)**. The main and right pulmonary arteries were dilated

(**Figure 4**). Double superior vena cava (SVC) was noted (**Figure 5A and B**). The left sided SVC draining into coronary sinus while the right sided SVC was seen draining into right atrium . Reflux of contrast was noted into the inferior vena cava and hepatic veins. Right atrium and ventricle appeared dilated –suggestive of right heart strain

The patient was advised admission in view of right heart strain and pulmonary hypertension. She refused admission and was lost to follow up

Discussion

Unilateral absence of the pulmonary artery(UAPA) is an uncommon developmental anomaly. It occurs more commonly in association with a cardiovascular anomaly, than as an isolated entity.The prevalence of this disorder is estimated to be 1 in 300,000.[1] The first reported case of UAPA was published in 1868. [2]

Most patients are usually detected incidentally. Common clinical presentation may be breathing difficulty, recurrent respiratory infection, and chest pain. Sometimes they may have haemoptysis and features of pulmonary hypertension. Haemoptysis occurs due to rupture of the pulmonary-systemic collaterals.[3] On examination, there will be reduced breath sounds on the affected side. It has been seen that left UAPA is known to be closely associated with additional congenital cardiovascular abnormalities (septal defects ,patent ductus arteriosus ,Tetralogy of Fallot, Transposition of great arteries,etc), whereas right UAPA occurs mostly as an isolated finding. This is why patients with left UAPA are more likely to present with symptoms at earlier age.[3,4]

Embryologic basis

The basic arterial pattern of the embryo at the end of 4th week of development is similar to that of fishes . A pair of dorsal aortae is seen on the dorsal body wall of the embryo. The dorsal aortae fuse below the level of the 7th intersegmental arteries to form single dorsal aorta. On the ventral body wall there is an aortic sac, which is the dilated part of the truncus arteriosus. The aortic sac is drawn out cranially as right and left horns . [5] They are connected to dorsal aortae through a series of six aortic arch arteries that pass through the pharyngeal arches (equivalent to gills). The cranial arch arteries degenerate as the caudal ones make their appearance. Therefore, at any given time all of the six arch arteries are never present in human being. The 5th arch artery is rudimentary. The 3rd , 4th and 6th arch arteries along with the aortic sac and dorsal aortae give rise to the definitive arteries of the thorax and neck **(Figure 6).** Blood from the ascending aorta enters the 3rd and 4th arch arteries, while blood from the pulmonary trunk enters the 6th arch arteries. This is because of the peculiar way the spiral septum of truncus arteriosus

extends into the aortic sac. The 6th arch artery opens into the dorsal aspect of the aortic sac, whereas the 3rd and 4th arch arteries open into the ventral aspect.[6]

The main pulmonary artery is arises from the truncoaortic sac . [6] The sixth arch arteries are divided into dorsal and ventral segments due to the appearance of a connection to the lung bud. The right and left pulmonary artery develop from ventral segment of right and left sixth arch artery respectively. The formation of the pulmonary vascular bed is independent of the sixth arch artery and occurs separately. Ductus arteriosus develops from the dorsal segment of left 6th arch artery. Absent pulmonary artery (as in our case) is caused by the involution of the proximal 6th aortic arch artery and persistent connection between the intrapulmonary vascular bed and distal 6th aortic arch artery. [5,6]

The arch of aorta develops from four sources namely aortic sac, left horn of aortic sac, left fourth arch artery and part of left dorsal aorta. Right dorsal aorta below the fourth arch artery disappears. Left subclavian artery develops from the left 7th intersegmental artery. The right subclavian artery develops from three sources, right 4th arch artery, part of right dorsal aorta and right 7th intersegmental artery.[5]

In our case we also saw right sided aortic arch with aberrant left subclavian artery. The arterial supply to left lung noted to arise the aberrant left subclavian artery. Right aortic arch anomaly is a mirror image of the normal pattern. Here, the dorsal aorta distal to the origin of 7th intersegmental artery disappears on left side, while the right dorsal aorta persists. The ductus arteriosus connects the right pulmonary artery with right arch of the aorta .It subsequently forms ligamentum arteriosum. . When the ligamentum arteriosum lies on the left side and runs beneath the oesophagus, it may cause swallowing difficulties. [6]

Double SVC was also seen in our case. During the 4th week of embryonic life there is a symmetrical pattern of venous channels. There are three pairs of cardinal veins. Anterior cardinal veins drain the cephalic half of the embryo. Posterior cardinal veins drain the caudal half of the embryo. Common cardinal veins enter the respective horns of the sinus venosus. During the 5th – 7th weeks a oblique anastomotic channel develops between right and left anterior cardinal veins. This channel diverts the venous blood from the left to the right side, consequent to which the veins on the left side shrink in size. The superior vena cava develops from 2 sources, the extrapericardial part from right anterior cardinal vein caudal to the oblique anastomosis and the intrapericardial part from right common cardinal vein (**Figure 7A and B**). Left superior vena cava occurs due to persistence of left anterior and left common cardinal veins .[6] It is usually asymptomatic. However, sometimes

CVC catheter may be placed in left SVC, and it appears as misplaced catheter on frontal chest X ray. So, knowledge of this anomaly is important and it is easily detected on CT Thorax.

Imaging features and differential diagnosis

Chest X ray will show reduced volume in the affected hemithorax with ipsilateral mediastinal shift absent hilar shadow/pulmonary artery, elevation of the ipsilateral diaphragm and reduced pulmonary vessels in the affected lung. Hyperinflation of the contralateral lung may be seen .Rib notching may be seen on affected side. [7]

CT Angiogram is used to confirm proximal absence of the pulmonary artery. The intrapulmonary arterial branches are preserved. Interrupted pulmonary artery is usually seen opposite to the side of the aortic arch, as in our case. Collaterals arising from the enlarged bronchial, intercostal, internal thoracic artery, phrenic artery or thoracic aorta may be seen. On MRI, diagnosis can be confirmed on steady state free precession (SSFP) or spin echo sequences[7,8]

HRCT Thorax usually shows reduced volume of the affected lung with fibrotic changes that may include mosaic attenuation, bronchiectasis, intraparenchymal and subpleural cysts with honeycombing. Fine peripheral reticulations with pleural thickening may be noted. Rib notching can be seen due to collateral circulation. The unaffected lung shows normal appearance or may show hyperinflation. [7]

V/Q scan will show preserved ventilation with absent perfusion in affected lung. Transfemoral catheter directed pulmonary angiography is not done now due to its invasive nature unless endovascular intervention is required.[8]

In case of hemithoracic volume loss with small hilum, differentials should include hyperlucency, Scimitar syndrome, Swyer James syndrome ,pulmonary artery stenosis or proximal interruption of the pulmonary artery(PIPA).

In pulmonary agenesis, there is complete absence of lung tissue, bronchi and vasculature distal to the carina. In pulmonary aplasia, there is a rudimentary bronchus ending in a blind pouch. There is no lung tissue or pulmonary vasculature. In pulmonary hypoplasia, lung is small in volume with a small pulmonary artery with transpleural collaterals on affected side. Hyperaerated opposite lung is seen .[4,7]

Scimitar syndrome is commonly seen almost exclusively on right side. It is characterised by anomalous pulmonary vein connecting the pulmonary veins of right lung to the infra diaphragmatic inferior vena cava. The right pulmonary artery is absent or reduced in caliber. Small hyperlucent right lung with disorders of lobulation/segmentation is seen. Presence of classic

"Scimitar sign" with demonstration of the draining vein on CT helps to differentiate from pulmonary hypoplasia or PIPA [1,7]

Swyer James syndrome usually appears on chest X ray as unilateral small hyperlucent lung with air trapping on expiration due to post-infective obliterative bronchiolitis. Bilateral involvement may be seen. Concomitant bronchiectasis is more in favour of small airway disease. Expiratory air trapping is not a feature of PIPA. On V/Q scan, PIPA shows preserved ventilation with absent perfusion, whereas in Swyer James syndrome there is a decrease in ventilation and perfusion of the affected lung [7,8]

Fibrosing mediastinitis is a rare non-malignant acellular collagen and fibrous tissue proliferative condition occurring in young adults causing compression of mediastinal structures. On imaging, there will be infiltrative soft tissue mass in the mediastinum causing compression of the pulmonary vessels, superior vena cava, , trachea-bronchial tree and oesophagus. Affected lung may show interlobular septal thickening due to veno-lymphatic obstruction. Often ,there is a calcified hilar mass (~85% cases) or hilar fullness .This is in contrast to small hilum seen in PIPA [2,4]

Pulmonary artery branch stenosis can occur anywhere along the course of pulmonary arteries, till the level of segmental artery .It is commonly associated with cardiovascular anomalies (TOF,VSD,ASD) and syndromes (Alagille syndrome, Williams syndrome).Unilateral stenosis will usually show evidence of post-stenotic dilation ,which is not seen in PIPA.[8]

Treatment of PIPA is usually conservative. Patient typically comes with dyspnoea and recurrent respiratory tract infection due to reduced lung volume and associated parenchymal changes. This is treated symptomatically. Pulmonary hypertension should be managed, as it increases risk for right heart failure and worsens prognosis.

In case there is haemoptysis, transcatheter selective embolization of the bleeding vessel has shown good results. In case of recurrent haemoptysis, pneumonectomy has been suggested.[8,]

Source of funding: This research did not receive any specific grant from funding agencies in the public, commercial, or not for- profit sectors

Institutional Ethics Committee approval was not required for this case report.

Author's Contribution

JJ: involved in initial diagnosis, jointly prepared manuscript and critically reviewed the manuscript.

UV: involved in initial diagnosis and critically reviewed the manuscript.

S T: reviewed images and wrote the manuscript

R B: involved in initial diagnosis

The manuscript has been read and approved by all the authors, the requirements for authorship as stated earlier in this document have been met, and each author believes that the manuscript represents honest work.

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