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CASE REPORT

Bronchial artery-pulmonary artery fistula: case report of a very rare cause of pulmonary hypertension

Ummara Siddique Umer, Aman Nawaz Khan, Shahjehan Alam, Syed Ghulam Ghaus, Seema Gul, Sadia Gul

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Author Information
From: Rehman Medical Institute, Peshawar, KP, Pakistan.
Department of Radiology

Dr. Ummara Siddique Umer
Consultant Radiologist
(Corresponding Author)
Email:
ummara.umer@rmi.edu.pk

Dr. Aman Nawaz Khan
Consultant Radiologist

Dr. Shahjehan Alam
Associate Professor

Dr. Syed Ghulam Ghaus
Consultant Radiologist

Dr. Seema Gul
Assistant Professor

Dr. Sadia Gul
Registrar

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ABSTRACT

Bronchial-pulmonary arterial fistula is a very rare vascular malformation. We report a case of a 20-year-old male with Bronchial-pulmonary arterial fistula, a primary racemose hemangioma, presented with shortness of breath and echocardiographic findings of pulmonary hypertension and was investigated using 128-slice Multidetector Computed tomography (MDCT) scanner. Images of entire chest and upper abdomen were taken in pulmonary artery phase. CTPA showed bronchial artery to pulmonary artery fistula of the right upper lobe near hilar level. Bronchial arteries showed convolution and dilation, were connected with pulmonary arteries, suggesting the presence of primary racemose hemangioma. There was resultant retrograde filling of descending aorta confirming right to left shunt.

Clinical relevance: 128 slice MDCT with its high resolution and multiplanar imaging plays an instrumental role in diagnosing pulmonary vascular anomalies. Establishment of non-invasive treatment strategy for bronchial-pulmonary artery fistula is urgently required.

Keywords: Multidetector Computed Tomography (MDCT), Bronchial-pulmonary artery fistula, CTPA.

Key Points:

- Bronchial artery-pulmonary artery fistula can be diagnosed and assessed in detail using multidetector CT Pulmonary angiography.
- CTPA should be performed to investigate the cause of pulmonary hypertension.

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INTRODUCTION

Bronchial-pulmonary arterial fistula is a very rare vascular malformation complicated with racemose hemangioma.[1] It often causes life-threatened hemoptysis.[2] In normal anatomic state, there are connections between the systemic and pulmonary circulations at capillary and pre-capillary levels. Although these connections exist in all humans, it is very rare for them to be evident radiologically, especially in the absence of an acquired etiology such as infection, trauma, surgery, and neoplasm.[3] Review of literature reveals different nomenclature of such lesions and may be referred to as a bronchial arteriovenous malformation, bronchopulmonary arterial anastomosis, bronchopulmonary shunts, and bronchial artery-pulmonary artery malformation and fistula depending on the vessels involved. Hyperplastic changes of the bronchial arteries may take the form of a racemose arteriovenous angioma or racemose hemangioma.[2] This term of Racemose hemangioma has been reported since the end of 1970s.[1]

Most congenital anomalies are the result of systemic supply to the lung with subsequent drainage through the pulmonary veins such as may be seen with a bronchopulmonary sequestration. Most direct connections between the bronchial and pulmonary arteries that have been described in the literature are acquired, often in the setting of chronic inflammatory pulmonary disease of varied sources, such as tuberculosis or pneumonia.[3] We report a very rare congenital anomaly, a bronchial artery-to-pulmonary artery fistula (BPAF) in a young adult.

CASE REPORT

A 20-year-old male presented to an outside hospital with complaints of shortness of breath and chest pain. Echocardiography revealed pulmonary hypertension. The patient was referred to our hospital for CT angiography of the pulmonary circulation. The study was performed on a 128 slice multidetector CT scanner (Toshiba) from the lung apices through the lung bases. Electrocardiographic gating was not used.

The images were reconstructed at 2-mm-thick intervals along with coronal and sagittal reformations and 3-dimensional volume-rendered images from the raw data. Intravenous contrast (Iopamidol) was injected at a rate of 4 mL/s with a total volume of 100 ml. CTPA findings identified bronchial artery to pulmonary artery fistula of the right upper lobe near hilar level (Figure 1).

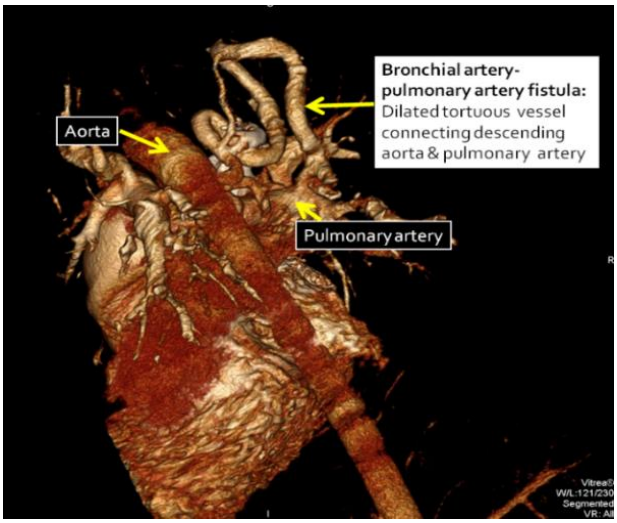
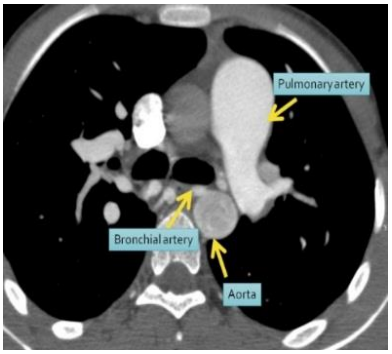
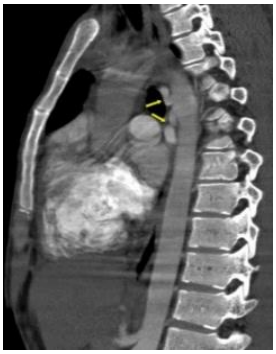


Figure 1: 3D volume image of the case showing bronchial artery-pulmonary artery fistula.

A tortuous dilated bronchial artery was seen arising from the anterior and right lateral descending aorta (Figure 2) having communication with the right pulmonary arterial circulation at hilar level. Bronchial arteries showed convolution and dilation, were connected with branches of pulmonary artery, suggesting the presence of primary racemose hemangioma. Multiplanar reformation in thin slices revealed an apparent tangle of vessels in the right hilum suggestive of a vascular malformation with a dilated feeding artery arising from the bronchial arterial circulation. The angiomatoid convolutions were seen arranged segmentally along the longitudinal axis of a bronchus.[4]



(2A)



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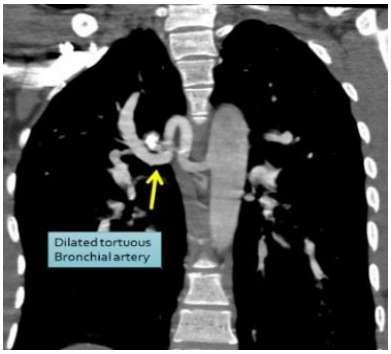
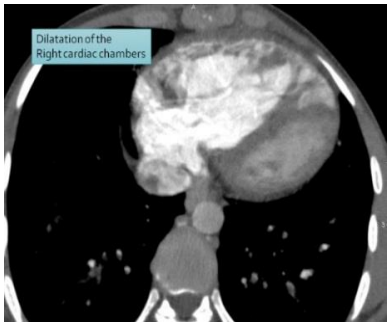


(2C)



(2D)

Figure 2 (A-D): CT pulmonary angiography images of the case with bronchial artery-pulmonary artery fistula. There is dilatation of bronchial arteries seen along anterior aspect of the descending aorta (arrows in 2B and 2C). Right ventricle is dilated. The heart was noted to have right chamber dominance (Figure 2D).

There was resultant retrograde filling of descending aorta confirming right to left shunt (Figure 3). The remainder of study was unremarkable with otherwise normal systemic vasculature, bronchial anatomy, and lung parenchyma.

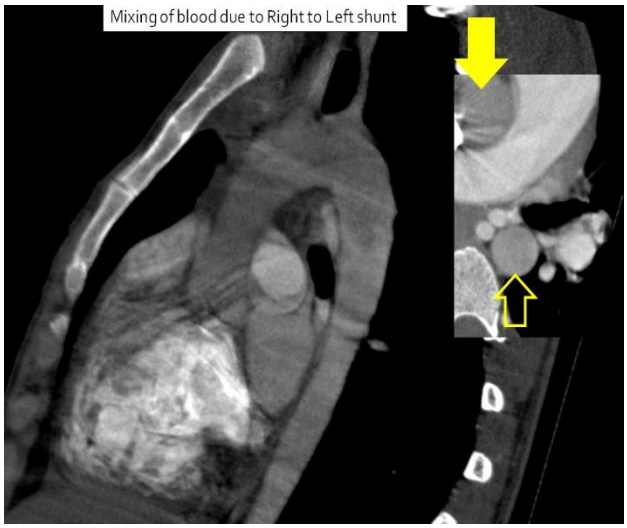


Figure 3: Reconstructed oblique sagittal CT image showing difference in densities of ascending and descending aorta. Small axial image on the right: Open arrow pointing at descending aorta, which is hyperdense due to filling in of contrast from the bronchial artery-pulmonary artery fistula as compared to the ascending aorta (arrow).

DISCUSSION

Blood flow to the lungs is from two different sources: Thin-walled pulmonary arteries circulate low-pressure deoxygenated blood to the lungs for gas exchange, and pulmonary veins drain oxygenated blood back to the heart. Meanwhile, bronchial arteries supply nutrients and oxygen at systemic pressures to the bronchi and lungs.[5] Classically, bronchial arteries arise from the descending aorta, usually around the fifth and sixth thoracic vertebrae. The lung may also receive systemic blood by way of intercostal, internal mammary, esophageal, and coronary arteries.[6,7,8] The venous drainage of systemic arterial supply to the lung is generally via the pulmonary venous circulation.[3]

As bronchial arteries extend out along the bronchial tree, they give off bronchopulmonary arterioles that eventually join with small pulmonary arterioles at the level of the preterminal (respiratory) bronchiole in an anastomotic plexus that supplies the capillaries at the level of the alveolus.[9,10,11] The combined systemic and pulmonary blood flow drains back to the heart through the pulmonary veins.[10] As such, this represents a normal connection between the systemic and pulmonary vascular systems, resulting in a clinically insignificant right to left shunt. Although this arteriolar plexus is functionally closed with little or no flow under normal conditions, channels can open or undergo neovascularization, resulting in a 10-fold increase in flow in disease states such as pulmonary embolus.[10]

Precapillary bronchopulmonary anastomoses are also seen frequently in other pathological states, but the existence of precapillary connections between systemic and pulmonary arteries in normal physiologic states has been the subject of great debate.[12,13] Although it is also conceivable that a bronchial artery-pulmonary artery malformation may form in response to an ischemic event in utero, this seems unlikely in this case given that the tracheobronchial tree and proximal pulmonary arteries were normal anatomically. There are many acquired processes that result in aberrant systemic and pulmonary arterial connections. Infection is a known cause, and bronchopulmonary arterial connections have been the results of bacterial pneumonia, pulmonary actinomycosis and tuberculosis. Although usually presenting as pulmonary arteriovenous fistulas, infections can very infrequently form connections between systemic and pulmonary vessels. Chronic inflammation at these sites can also lead to dilatation of normal anastomotic connections, as well as neovascularization of capillaries at these same sites. Trauma and iatrogenic injury can result in arteriovenous or arterio-arterial fistulas. Finally, neoplasms including lung cancer and lymphoma with systemic-pulmonary arterial anastomoses have been described.[3] None of these predisposing acquired causes were present in this case.

Congenital malformations, especially ones involving the heart and lungs, are the most frequent cause of systemic-pulmonary artery communications. Considering the full gamut of potential systemic and pulmonary artery connections, primary malformation involving a bronchial artery to pulmonary artery or vein is considered to be extremely rare and only 4% of pulmonary arteriovenous malformations have systemic arterial connections.[14]

Bronchial artery malformations are more common in men and are usually unilateral, most often occurring in the right lung.[14] In reviewing the literature, we tried to find a similar case to ours. Most of the cases (if not all) seem to be acquired lesions. The closest example is that by Geyik et al.[9] In this case, there were long internal mammary and phrenic artery collateral vessels that formed a malformation along the periphery and resulted in retrograde flow through the pulmonary artery. The distal connection and multiple feeding vessels led the authors to hypothesize that this truly resulted from an unidentified inflammatory or infectious etiology. In our case, the length of the systemic vessel was shorter and the connection to the pulmonary artery was more proximal near hilum resulting in an anterograde pulmonary arterial flow.

Diagnosis can be made by conventional angiography, but recently CT scan with its multiplanar techniques is being considered the first line imaging in confirming the cause of pulmonary hypertension. Not only the vascular anomalies, but the lung parenchyma, airway and chest wall can be looked at in detail with CT scan. The common congenital causes of pulmonary hypertension seen on CT are pulmonary stenosis, cardiac shunts, patent ductus arteriosus etc.

Bronchial artery-pulmonary artery fistula is a very rare cause of pulmonary hypertension. In our case, the pulmonary-systemic shunting was appreciated by demonstration of contrast in the descending aorta, much before the aortic root. Interventional therapy should be performed in such cases to decrease symptoms as well as prevent future complications from hemoptysis and/or pulmonary hypertension.

CONCLUSION

Bronchial-pulmonary artery fistula is a very rare congenital anomaly. MDCT plays instrumental role in diagnosing pulmonary vascular anomalies.

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