Indian Journal of
Applied Radiology

Open Science Publications

Volume 9, Issue 1 - 2023

www.opensciencepublications.com

A Rare Case of Isolated Proximal Interruption of the Right Pulmonary Artery

Case Report

Naukarkar V*, Varma R and Shetty D

Department of Radio diagnosis, Nair Hospital, Mumbai, India

***Corresponding author:** Naukarkar V, Department of Radio diagnosis, Nair Hospital, Mumbai, India, Phone: +91 8767207597; E-mail: naukarkarvikrant@gmail.com

Article Information: **Submission: 02/02/2023; Accepted: 10/03/2023; Published: 15/03/2023**

Abstract

Proximal interruption of the pulmonary artery (PIPA) is a rare congenital anomaly. Presentation varies from mild breathlessness to life-threatening hemoptysis. In isolation, this anomaly has a very good prognosis. But when associated with cardiac abnormalities the prognosis depends on the associated conditions. Therefore, accurate and prompt diagnosis is very essential. Here we describe a case of proximal interruption of the pulmonary artery presenting with mild hemoptysis which was managed conservatively.

Keywords: Pulmonary Artery Anomalies; Interrupted; Radiology; Case Report

Introduction

Proximal interruption of the pulmonary artery is an abnormality where a proximal portion of a pulmonary artery shows abnormal development with preserved, normally developed intrapulmonary vasculature. It creates a condition of chronic hypoxia which leads to a lot of physiological alterations. Here, we describe an interesting case, where PIPA led to collateral vessel formation, the rupture of which caused the patient to present with mild hemoptysis which was managed conservatively.

Case Presentation

A 31-year-old male presented to the casualty department with complaints of acute onset on-and-off fever, mild hemoptysis, vomiting, abdominal pain, and loose stools. On probing further, he also gave a history of mild chronic dyspnea on exertion. The patient was mildly hypotensive with tachypnoea and tachycardia but maintained oxygen saturation. He denied any addictions.There was no history of any surgery in the past. A respiratory examination revealed reduced respiratory sounds on the right side of the chest. Cardiovascular, neurological, and abdominal examinations were normal.

Routine investigations were ordered as per the hospital protocol. Electrocardiographic findings were within normal limits. Hematological cell counts were adequate. Chest X-ray revealed mild elevation of the right dome of the diaphragm and apical pleural thickening. Mild blunting of the right costophrenic recess was noted. Bony thorax and heart size were appropriate. What was surprising to us was the inconspicuous right hilum and indiscernible opacity of the right pulmonary artery (Figure 1). It was decided to go ahead with computed tomography pulmonary angiography with high-resolution thoracic sections. CT revealed the right proximal pulmonary artery interruption after 1 cm of origin. The left pulmonary artery was hypertrophied (Figure 2). There was a paucity of peripheral pulmonary vasculature which appeared narrow in caliber as compared to the left side. Multiple enlarged bronchial, Trans plural collateral vessels were observed supplying right lung parenchyma. The right internal mammary artery was larger in caliber than the left. Mild pleural thickening was noted. The lung window revealed right-sided smooth

ISSN: 2581-3919

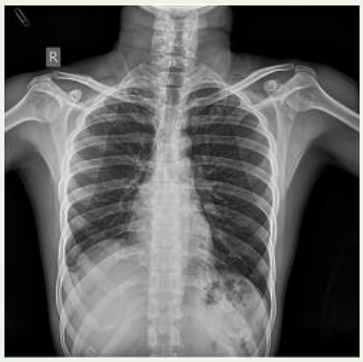

Figure 1: Chest radiograph posterior-anterior projection showing inconspicuous right hilum, indiscernible opacity of the right pulmonary artery, mild right apical pleural thickening, and blunting of right costophrenic recess.

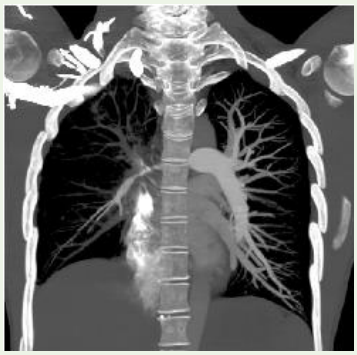

Figure 2: Computed tomography pulmonary angiography coronal reformatted maximum intensity projection mediastinal window image shows interrupted right proximal pulmonary artery with a paucity of distal intrapulmonary arteries. The left pulmonary artery is hypertrophied.

interstitial septal thickening suggestive of hypertrophied collateral vessels. The tracheobronchial tree revealed no abnormalities.

Due to the recent Leptospira outbreak in the rainy season, the patient was evaluated with anti-leptospiral antibodies which came positive. Because of mild symptomatology, it was decided to treat the patient conservatively. The patient was started on ceftriaxone and adequate hydration was maintained to tackle hypotensive status. The patient improved dramatically within a week. For interrupted right-sided pulmonary artery follow-up was suggested as the patient only had associated mild chronic dyspnea on exertion (Figure 3).

Discussion

Proximal interruption of the right pulmonary artery (PIPA) is a rare congenital anomaly with a reported prevalence of 1:2,00,000. Usually, PIPA occurs on the side opposite to that of the aortic arch. It is more common on the right side. When on the left side it is associated with cardiac anomalies and a right-sided aortic arch. The most common cardiac anomalies are tetralogy of Fallot, patent ductus arteriosus, and atrioventricular septal defects. Left-sided anomaly presents earlier and at a young age. Proximal pulmonary arteries develop from the proximal 6th aortic arch in the first trimester of pregnancy. The distal part of the left 6th aortic arch forms ductus

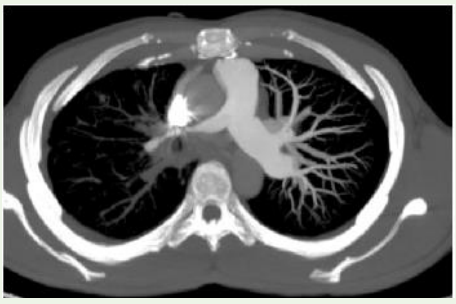

Figure 3: Computed tomography pulmonary angiography axial reformatted maximum intensity projection mediastinal window image shows interrupted right pulmonary artery 1 cm beyond the bifurcation.

arteriosus while the distal part is involuted on the right side. PIPA develops from abnormal development of the proximal part of the 6th aortic arch. Intrapulmonary vasculature has a separate origin and hence is developed normally. Three groups of the anomaly are recognized. In group 1 there is the presence of a left to right shunt most commonly patent ductus arteriosus. In group 2 associated pulmonary hypertension is present. An isolated anomaly without associated pulmonary hypertension is seen in group 3 [1].

Patients with PIPA are mostly asymptomatic or present with mild symptoms. The most common symptoms include chronic exertional dyspnoea, decreased exercise tolerance, recurrent infections, and hemoptysis. Reduced blood flow and hypoplasia of the lung lead to progressive dyspnoea on exertion. Recurrent respiratory infections are caused by the decreased delivery of systemic immunomodulators due to the interruption of blood supply proximally. Rupture of hypertrophied thin-walled collateral vessels can cause hemoptysis [2].

Diagnosis of the abnormality can be suspected on chest radiography based on findings of absent pulmonary artery opacity, and inconspicuous hilum. Other associated findings include ipsilateral lung volume loss with ipsilateral cardio mediastinal shift and elevation of the hemidiaphragm. Compensatory hyperinflation of the contra lateral lung with herniation to the opposite side can be seen. Collateral vessel formation leads to peripheral reticular opacities, pleural thickening, and rib notching. Recurrent infection can cause cystic-bronchiectasis changes.

Computed tomography pulmonary angiography (CTPA) is widely done to diagnose PIPA since it is rapid, readily available, and hasan excellent spatial resolution (Figure 4). Collateral vessels are better appreciated. Thin 0.625 mm sections are obtained from lung apices to the top of the diaphragm after giving 1.5 ml/kg contrast at the rate of 3.5-4 ml/sec bolus dose. Maximum intensity projection reformats are made. A high-resolution lung window better assesses the bronchial tree and lung parenchyma. Normally developed bronchial tree is seen. CT findings include proximal interruption of the pulmonary artery with preserved or paucity of the distal pulmonary vasculature. Lung parenchyma is perfused by systemic hypertrophied collateral vessels from bronchial, trans-pleural intercostal, internal mammary, phrenic, subclavian, and brachiocephalic arteries. These changes can lead toperipheral reticulations, pleural thickening, and rib notching. Sometimes an aberrant artery arising from the aorta

Citation: Naukarkar V, Varma R, Shetty D. A Rare Case of Isolated Proximal Interruption of the Right Pulmonary Artery. Indian J Appl Radiol. 2023;9(1): 179.

can be seen supplying the lung parenchyma. Ipsilateral volume loss of lung parenchyma with mediastinal shift, crowding of ribs, and elevation of the hemidiaphragm is seen. Compensatory contra lateral lung volume expansion and hypertrophy of the pulmonary artery are noted. Intraparenchymal and subpleural cysts and bronchiectasis changes can be seen in cases of recurrent infections.

Magnetic resonance angiography (MRA) of the thorax is an alternative to a CT scan. Respiratory and electrocardiographic gating is used to acquire better images. T1 and T2 weighted TSE images in axial and coronal planes are obtained in minimum free breathing or breath holding as per the patient's condition. Pre- and post-gadolinium contrast images with subtraction are also acquired. However, due to limited availability, long duration, and cost consideration CT is preferred over MRI. The only indication of MRI is in young patients to avoid radiation exposure (Figure 5).

Digital subtraction angiography best depicts proximal interruption of the pulmonary artery and offers simultaneous intervention in cases of hemoptysis.

Other investigations include a ventilation-perfusion scan (V/Q scan). It shows decreased perfusion on the side of interruption with maintained ventilation. Echocardiography can be done to look for associated cardiac anomalies [3].

PIPA is managed conservatively depending on the symptomatology. Rarely in case of severe symptoms intervention is needed. Recurrent infections are treated with appropriate antibiotics. Chronic breathlessness due to pulmonary hypertension seen in 19-27% of patients requires treatment with phosphodiesterase inhibitors and endothelin receptor antagonists. Hemoptysis is seen in 10-20 % of cases. It is self-limiting most of the time. When significant, percutaneous trans-arterial embolization of bleeding collateral vessels is done. Various embolization materials include gel foam, polyvinyl alcohol, glue, and coils. It carries a success rate of 73-99 % and a recurrence rate of 10-55 %. In cases of recurrent hemoptysis even after embolization, pneumonectomy is performed. Surgical reconstruction of interrupted pulmonary artery can be done but the procedure carries high morbidity and mortality [4] (Figure 6).

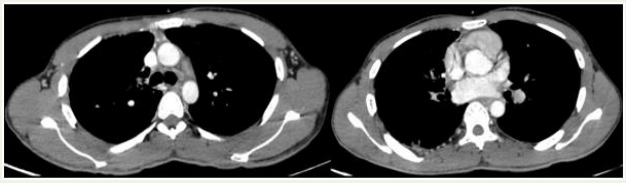

Figure 4:(High-resolution contrast-enhanced computed tomography thorax axial reformatted mediastinal window images show dilated bronchial and trans-pleural intercostal collateral vessels. The internal mammary artery on the right side is hypertrophied as compared to the left.

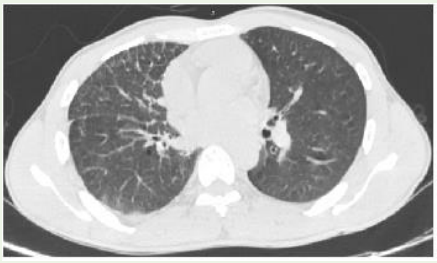

Figure 5: High-resolution computed tomography thorax axial reformatted lung window image shows smooth interstitial septal thickening on the right side suggestive of hypertrophied collateral vessels.

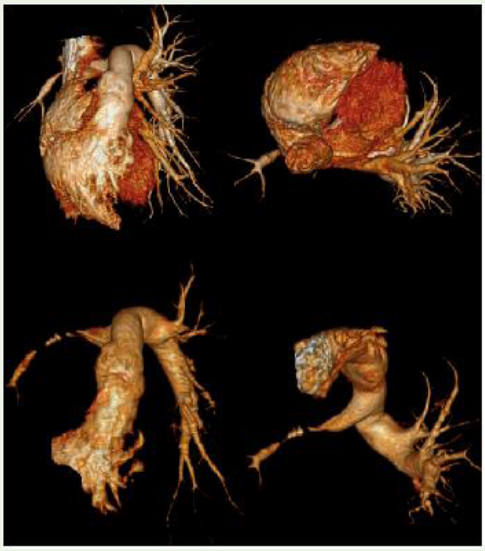

Figure 6: Computed tomography pulmonary angiography coronal and axial reformatted 3D volume rendered images show interrupted right proximal pulmonary artery with a paucity of distal intrapulmonary arteries. The left pulmonary artery is hypertrophied.

Hypogenetic lung syndrome is one of the differentials where hypoplasia of lung parenchyma with anomalous venous drainage is seen. A scimitar-shaped abnormal draining vein along the right heart border differentiates this condition from PIPA. Swayer James syndrome can mimic PIPA where unilateral lung hypoplasia with increased lucency will be seen due to post-infectious obliterative bronchiolitis. Unlike PIPA air trapping will be seen on expiratory images. Primary pulmonary hypoplasia is characterized by the hypoplasia of normal lung parenchyma, bronchial tree, and pulmonary vasculature. On the contrary in PIPA bronchial tree is normal. Pulmonary artery branch stenosis is one of the differentials but can be ruled out by the absence of post-stenotic dilatation [5].

Conclusion

Proximal interruption of the pulmonary artery is an uncommon congenital anomaly with varied presentation. Due to the benign nature of the disease, it needs to be differentiated from other more ominous conditions by appropriate investigations. PIPA is mostly managed conservatively unless complicated by significant hemoptysis, where in percutaneous transarterial embolization or pneumonectomy is performed.

References

1. Davis SD (2000) Case 28: Proximal interruption of the right pulmonary artery. Radiology 217: 437-440.

Citation: Naukarkar V, Varma R, Shetty D. A Rare Case of Isolated Proximal Interruption of the Right Pulmonary Artery. Indian J Appl Radiol. 2023;9(1): 179.

2. Anand SH, Jasper A, Mani SE, Joseph E, Mathai J (2015) Proximal Interruption of the Pulmonary Artery: A Case Series. J Clin Diagn Res 9: TD04-TD06.

3. Williams EA, Cox C, Chung JH, Grage RA, Rojas CA (2019) Proximal Interruption of the Pulmonary Artery. J Thorac Imaging 34: 56-64.

4. Liu B, Monroe EJ, Kogut MJ (2015) Proximal interruption of the pulmonary artery: Transcatheter embolization for emergent management of massive hemoptysis. Radiol Case Rep 8: 865.

5. Zhang M, Cao Y, Zhou N, Wang R, Wu H et al. (2022) Proximal interruption of the pulmonary artery: A review of radiological findings. Front Pediatr 10: 968652.

Citation: Naukarkar V, Varma R, Shetty D. A Rare Case of Isolated Proximal Interruption of the Right Pulmonary Artery. Indian J Appl Radiol. 2023;9(1): 179.