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CASE REPORT

Unilateral Absence of Pulmonary Artery with Ipsilateral Interstitial Lung Disease

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ABSTRACT

Unilateral absence of pulmonary artery is a rare congenital malformation that can present as an isolated disease or may be associated with other congenital cardiac defects.Interstitial lung diseaseis usually a bilaterallung disease. Unilateral Interstitial lung disease is a rare condition, can be seen in unilateral absence of pulmonary artery, sarcoma of unilateral pulmonary artery, unilateral pulmonary venous thrombosis, and radiation-induced pneumonitis. Unilateral absence of pulmonary artery with ipsilateral interstitial lung disease is a very rare entity. Here we present a 74-year-old male with a known history of pulmonary tuberculosis presented with covid 19 infection. He underwent High Resolution Computed Tomography, which revealed a congenitally absent right pulmonary artery which was presumed to the cause of his unilateral lung disease.

Keywords: Congenital lung disease, Unilateral absence of pulmonary artery, interstitial lung disease, pulmonary hypertension, congestive heart failure, recurrent respiratory infections

ABBREVIATIONS

PA - pulmonary artery

UAPA - Unilateral absence of pulmonary artery

ILD - Interstitial lungdisease

HRCT - High resolution computed tomography

CT - Computed tomography

MR - Magnetic resonance imaging

INTRODUCTION

An absent pulmonary artery (PA) is caused by the involution of the proximal sixth aortic arch and persistence of the connection of the intrapulmonary PA to the distal sixth aortic arch.Unilateral absence of pulmonary artery (UAPA) is a rare congenital anomaly which occurs during embryogenesis due to interruption of proximal sixth aortic arch[1]. It was first described by Frentzel in 1868; in his initial paper, Frentzel reported that 30% of patients with this finding could remain asymptomatic until adult life[2] . Unilateral interstitial lungdisease (ILD) also a rare entity, sequelae to absence of ipsilateral

pulmonary artery (PA). Other causes of Unilateral interstitial lung disease are sarcoma of unilateral pulmonary artery, unilateral pulmonary venous thrombosis, and radiation-induced ILD[3] .Most commonlyunilateral absence of pulmonary artery accompanied by cardiovascular anomalies such as ventricular septal defects, tetralogy of Fallot, coarctation of the aorta, aortic stenosis, transposition of the great arteries[4] . Unilateral absence of pulmonary artery also may occur as an isolated finding. Patients can present with pulmonary hypertension, congestive heart failure, recurrent respiratory infections or completely asymptomatic. Most patients who have no associated congenital cardiac anomalies have only mild or absent symptoms and survive into adult life.

CASE REPORT

A 74-year-old male patientwith a known history of pulmonary tuberculosis presented with covid 19 infection with complaints of progressive dyspnoea. He had past history of recurrent lower respiratory tract infections. Pulmonary function test showed restrictive pattern. High Resolution Computed Tomography was performed which showed markedly reduced right lung volume with inter and intralobular septal thickening, ground glass opacities, peri-bronchial thickening with traction bronchiectasis and peripheral honey combing. Absence of right pulmonary artery with prominent ipsilateral internal mammary artery noted. Compensatory hyperinflation of left lung herniating across the midline.

DISCUSSION

Absence of pulmonary artery at its origin from the main pulmonary artery is known as unilateral absence of pulmonary artery. In contradiction with pulmonary agenesis where there is complete absence of lung parenchyma with associated blood vessels. An absent PA is caused by the involution of the proximal sixth aortic arch and persistence of the connection of the intrapulmonary PA to the distal sixth aortic arch. Systemic collateral vessels from internal mammary, aortopulmonary, subclavian and intercostal arteries supply the ipsilateral lung parenchyma. Unilateral interstitial lung disease is a very rare lung condition associated with proximal interruption or absence of the pulmonary artery.Diagnosis of UAPA is very difficult and is based on taking a complete medical history, physical examination and imaging examinations[5]. Clinical presentation can vary according to the associated congenital heart abnormalities. Patients with isolated absence of pulmonary artery may be missed in early life and can present late, as happened with our patient. Examination may reveal an asymmetrical chest with abnormal breath sounds in the affected side. There may be a systolic ejection murmur across the pulmonary outflow tract. The clinical presentation varies from dyspnea, recurrent chest infections, pulmonary oedema, pulmonary hypertension, hemoptysis, exercise intolerance and chest pain. Chest radiography shows ipsilateral volume loss with diaphragmatic elevation, linear reticular opacities, tracheo-mediastinal shift to the affected side. The contralateral lung shows hyperinflation and herniation to the affected side. HRCT lung window shows markedly reduced affected lung volume with inter and intralobular septal thickening, ground glass opacities, peri-bronchial thickening with bronchiectasis and peripheral honey combing. Mediastinal window shows absence of affected pulmonary artery at its origin or may terminate within 1cm of its origin[6]. Differential to this condition is Swyer- James syndrome which shows air trapping, hyperlucent with diminished vascularity on expiration. CT pulmonary angiography shows non visualisation of pulmonary artery. MR angiography (MRA) can be used in young patients to avoid radiation exposure. The therapeutic approach for isolated absence of PA should be based on clinical symptoms of the patient.

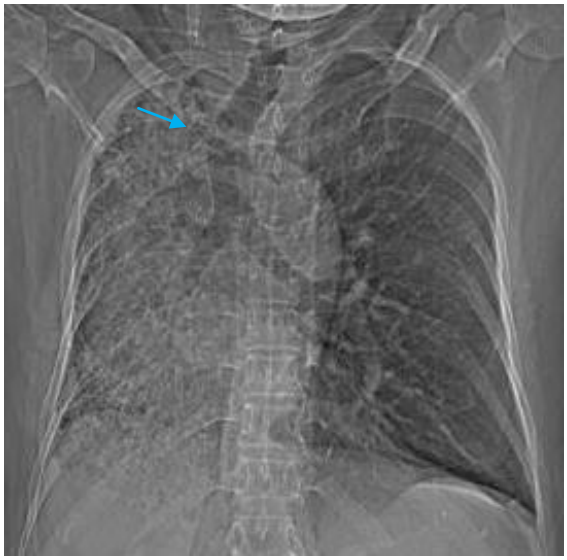

Figure 1a: CT topogram showed reticular opacities in the right lung with volume loss, tracheo-mediastinal shift to the right (blue arrow)

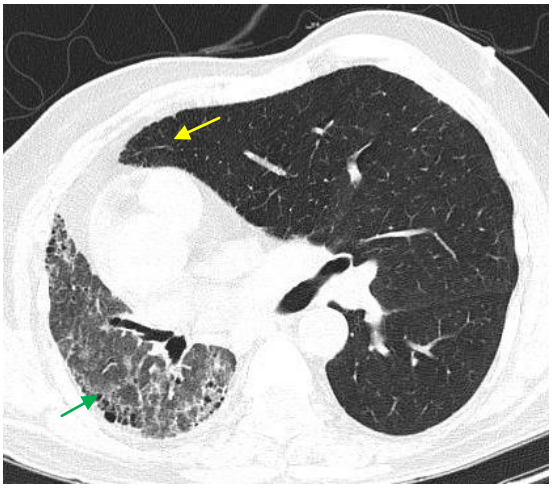

Figure 1b: CT Non contrast lung window showed diffuse inter and intralobular septal thickening, ground-glass opacities, bronchiectasis, and honeycombing (green arrow). Compensatory hyperinflation of left lung herniating across the midline (yellow arrow).

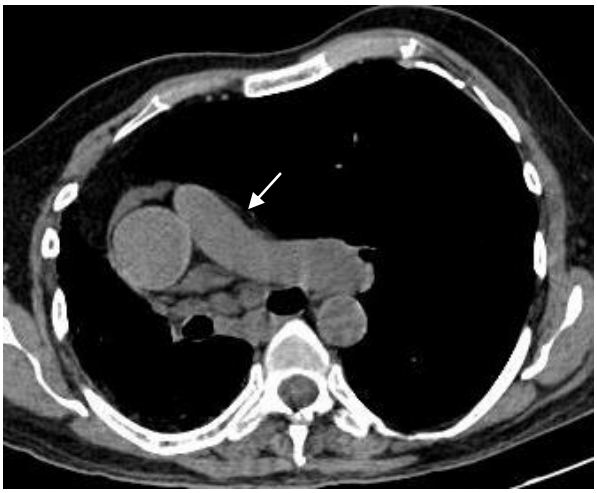

Figure 2: CT non contrast mediastinal window showed absence of right pulmonary artery (white arrow).

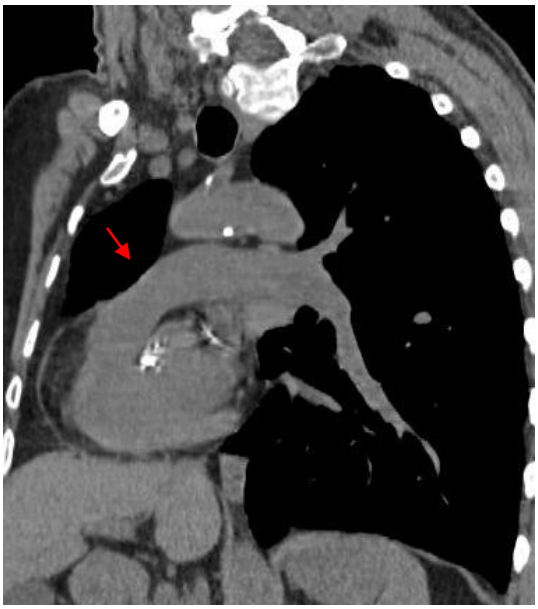

Figure 3: Curved planar reformation of pulmonary trunk showed absence of right pulmonary artery (red arrow).

CONCLUSION

Isolated absence of pulmonary artery with interstitial lung diseaseis a rare entity. Adult patients are often asymptomatic. Pulmonary hypertension is a severe complication of isolated absence of pulmonary artery and significantly increases the morbidity and mortality rate of the disease.Clinical diagnosis of isolated absence of pulmonary artery is difficult, imaging is required. The diagnosis of UAPA can be made by chest radiography and 2D-Echo. Anatomic details and the presence of interstitial lung diseasecan be identified by CT and MRI. The early diagnosis of UAPA can prevent further deterioration and can prevent the potential complications.

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CONFLICT OF INTEREST

No Conflict of interest

REFERENCES

1.Bhamidipati, K. D. P., & Vandana, M. (2021). Unilateral proximal interruption of pulmonary artery with ipsilateral interstitial lung disease–A rare case report. Radiology Case Reports, 16(8), 2021-2024.

2.Fraentzel, O. (1868). Ein Fall von abnormer Communication der Aorta mit der Arteria pulmonalis. Archiv für pathologischeAnatomie und Physiologie und für klinischeMedicin, 43(3), 420-426.

3.Mangla, L., Shamsi, I., Talwar, D., & Talwar, D. (2021). Radiological quandary: Unilateral interstitial lung disease–Truth or dare. Lung India: Official Organ of Indian Chest Society, 38(2), [P202–204.

4.Jan Ten Harkel, A. D., Blom, N. A., &Ottenkamp, J. (2002). Isolated Unilateral Absence of a Pulmonary Artery. Chest, 122(4), 1471–1477.

5.Esquinas-Requena, J. L., Lozoya-Moreno, S., García-Nogueras, I., Atienzar-Núñez, P., Sánchez-Jurado, P. M., &Abizanda, P. (2020). La anemiaaumentaelriesgo de mortalidaddebido a fragilidad y discapacidadenmayores: Estudio FRADEA. Atención Primaria, 52(7), 452-461.

6. Ryu, D. S., Spirn, P. W., Trotman-Dickenson, B., Hunsaker, A., Jung, S. M., Park, M. S., ... & Costello, P. (2004). HRCT findings of proximal interruption of the right pulmonary artery. Journal of Thoracic Imaging, 19(3), 171-175.