Sir,

Unilateral pulmonary vein atresia is a rare congenital anomaly where long segments of pulmonary veins of one lung are atretic. Patients often present with recurrent episodes of respiratory tract infections and hemoptysis. It is often associated with congenital heart disease or anomalous pulmonary venous return. Due to the severity of the condition, patients are usually identified within the first 3 years of life. Our case was unique because the patient had isolated unilateral pulmonary venous atresia with no associated congenital heart disease and was diagnosed in adulthood. To our knowledge, there are less than 50 cases of this condition, reported in adults.

A 20-year-old female presented with complaints of dyspnea on exertion since childhood, which was progressively worsening since 3 months. Dyspnea occurs on running or climbing stairs. The patient had no history of cough or fever or wheezing or palpitations, but had a history of recurrent episodes of chest infections since childhood for which she had taken treatment and recovered completely. On clinical examination, the trachea was deviated to right with decreased chest movement on right side. On auscultation, breath sounds were decreased on right side in all areas. The patient was advised a chest radiograph. Posterior–anterior chest radiograph [Figure 1] shows a small right hemithorax, with prominent reticular opacities in mid and lower zones. There is associated mediastinal shift to right with elevation of the right hemidiaphragm. Contrast enhanced computed tomography (CECT) thorax was performed for further evaluation. Mediastinal window [Figure 2] shows absent right superior and inferior pulmonary veins with a smooth right border of left atrium. A low-attenuation soft tissue is seen at the mediastinum adjacent to the left atrium in the subcarinal location, not causing narrowing of right main bronchus with enhanced vascular channels within, which represent prominent bronchial collaterals. The right main pulmonary artery is smaller in caliber as compared to the left [Figure 3]. There is associated ipsilateral mediastinal shift. Lung window [Figure 4] shows a hypoplastic right lung with diffuse interlobular septal thickening.

Based on these imaging findings, the final diagnosis of isolated unilateral pulmonary venous atresia was made.

Congenital unilateral pulmonary vein atresia results from the failure to incorporate common pulmonary vein into the left atrium during the embryological development. This results in complete or partial atresia of pulmonary veins on one side.\[1\] There is no right- or left-sided predominance of this condition, and in up to 50% of cases it is associated with cardiac defects or anomalous pulmonary venous drainage.\[1,2\] The condition usually manifests in infancy or within the first 3 years of life.\[1\]

Patients often present with a varying degree of clinical severity, ranging from recurrent pulmonary infections, dyspnea on exertion, and hemoptysis to being completely asymptomatic.\[1,3\]
Chest radiographs show small hemithorax, ipsilateral mediastinal shift, reticular opacities, and Kerley B lines.[1]

CECT shows characteristic findings of a hypoplastic lung, smooth margins of left atrium without evidence of rudimentary pulmonary veins,[1,2] and smooth thickening of interlobular septa likely due to the dilation of pulmonary lymphatics and bronchial veins.[4] There is associated small ipsilateral pulmonary artery, attributed to preferential perfusion to contralateral side and confluent low attenuation soft tissue in the mediastinum adjacent to the left atrium that contains pulmonary to systemic collaterals.[1] In adults, this mediastinal soft tissue can pose a diagnostic dilemma with fibrosing mediastinitis and mass lesion being considered as the differentials.[1] However, a small hemithorax in the absence of bronchial obstruction suggests a congenital anomaly.

Treatment of unilateral congenital pulmonary venous atresia may be conservative in relatively asymptomatic patients, and pneumonectomy is done in patients with progressive dyspnea, significant pneumonia, or recurrent pneumonia.[3]

Patient was given antibiotics and symptomatic treatment. Following recovery, she was referred to a higher center for further management.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

Ramakrishna Narayanan,
Balasubramanyam Shankar, Samir Paruthikunnan

Department of Radiodiagnosis, Kasturba Medical College,
Manipal, Karnataka, India
E-mail: drrkris@gmail.com

REFERENCES
1. Porres DV, Morenza OP, Pallisa E, Roque A, Andreu J, Martínez M. Learning from the pulmonary veins. Radiographics 2013;33:999-1022.
2. Heyneman LE, Nolan RL, Harrison JK, McAdams HP. Congenital unilateral pulmonary vein atresia: Radiologic findings in three adult patients. AJR Am J Roentgenol 2001;177:681-5.
3. Kim Y, Yoo IR, Ahn MJ, Han DH. Asymptomatic adults with isolated, unilateral right pulmonary vein atresia: Multidetector CT findings. Br J Radiol 2011;84:e109-13.
4. Dixit R, Kumar J, Chowdhury V, Rajeshwari K, Sethi GR. Case report: Isolated unilateral pulmonary vein atresia diagnosed on 128-slice multidetector CT. Indian J Radiol Imaging 2011;21:233-6.

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

Access this article online

Quick Response Code:  
Website: www.lungindia.com
DOI: 10.4103/0970-2113.189990

How to cite this article: Narayanan R, Shankar B, Paruthikunnan S. Isolated unilateral pulmonary vein atresia. Lung India 2016;33:571-2.