Case 12255

Congenital atresia of the left pulmonary veins and single right meandering vein
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Section: Chest imaging
Area of Interest: Lung Pulmonary vessels
Procedure: Diagnostic procedure
Imaging Technique: CT
Special Focus: Congenital Case Type: Clinical Cases
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Patient: 19 years, female

Clinical History:

A 19-year-old woman was diagnosed with bronchial asthma. A chest radiography was undertaken and showed volume loss in left hemithorax with small ipsilateral pulmonary artery and a tubular opacity in the right perihilar area. Chest CT angiography was performed to complete the study.

Imaging Findings:

Chest radiography shows volume loss in left hemithorax with reduced size of the left main pulmonary artery and a serpentine tubular structure in the right medial lobe in a juxta-hilar location suggesting an abnormality in the course of the right pulmonary veins (Fig 1a, b).

Angio CT examination reveals congenital atresia of the left pulmonary veins (Fig. 2), with left lung hypoplasia and a small left pulmonary artery (Fig. 3). There is also development of systemic collateral arterial circulation with prominent bronchial and intercostal arteries. Smooth thickened interlobular septa are observed, reflecting pulmonary venous hypertension secondary to the atresia of the left pulmonary veins (Fig. 4). In the right lung, a single right inferior pulmonary vein entering in the left atrium is observed. This single vein is markedly dilated describing a tortuous way ("meandering vein"), collecting all of the pulmonary vein drainage of the right lung (Fig. 5).

Discussion:

Imaging studies confirm a congenital atresia of the left pulmonary veins and the presence of a single meandering vein in the right lung.

A) The congenital atresia of the pulmonary veins is a rare congenital anomaly and probably results from a deficiency in the incorporation of the common pulmonary vein into the left atrium. It can occur in either of the two lungs, without predominance of right or left side. This condition is usually diagnosed in childhood and most frequent symptoms include recurrent pulmonary infection, mild dyspnoea, or haemoptysis [1] due to hypertrophy of bronchial arteries. In approximately 50% of patients it is associated with other congenital heart defects or anomalous pulmonary venous return [2]. Multidetector CT is the preferred imaging technique and confirms the suspected diagnosis made in the chest X-ray. The CT findings include: small ipsilateral hemithorax with a small ipsilateral pulmonary artery and absence of any draining ipsilateral pulmonary vein into the left atrium, the left atrial wall being completely smooth, with no evidence of even a rudimentary pulmonary vein [2, 3]. Mediastinal abnormal soft tissue can also be observed representing the pulmonary-to-systemic venous collateral circulation [4]. Pulmonary artery hypoplasia
explains the systemic-to-pulmonary collateral arteries observed across the bronchial and intercostal arteries. Interlobular septal thickening, peribronchovascular thickening and ground-glass opacities reflect pulmonary venous hypertension and swollen lymphatic vessels. This case shows the main components of this entity: pulmonary hypoplasia with a small hemithorax associated with ipsilateral mediastinal shift, small ipsilateral pulmonary artery, development of arterial supply through systemic branches, absence of pulmonary venous connection to the left atrium [4] and parenchymal abnormalities such as interlobular septal thickening and peribronchovascular thickening.

B) On the other hand, the right lung shows a single pulmonary vein with a meandering course. An anomalous unilateral single pulmonary vein is a single vein that drains only to the left atrium after collecting all pulmonary veins [5]. This failure can also be associated with ipsilateral lung hypoplasia and it should be distinguished from the hypogenetic lung (scimitar syndrome). However, the anomalous vein has a normal drainage in the left atrium [6], so that no bypass occurs. The other entity that should be differentiated is anomalous unilateral single pulmonary arteriovenous malformation. A vein with meandering course does not require treatment, because it is not a vascular shunt. However, the scimitar syndrome and arteriovenous malformation syndrome may require surgery or embolization to correct the shunt.

Differential Diagnosis List: Congenital atresia of the left pulmonary veins and single right meandering vein., Left lung findings: a) Congenital atresia of the left pulmonary veins b) Pulmonary hypoplasia secondary to unilateral absence of the pulmonary artery c) Sywer-James-MacLeod syndrome, Right lung findings: a) Scimitar syndrome. b) Single right meandering vein. c) Unilateral single pulmonary arteriovenous malformation

Final Diagnosis: Congenital atresia of the left pulmonary veins and single right meandering vein.

References:

Figure 1

Description: A serpentine tubular structure is noted in the medial lobe. In the left lung, loss of lung volume and hypoplasia of the main pulmonary artery are seen. Origin: Department of Radiology, University Hospital Dr Peset, Valencia, Spain.
Description: A serpentine tubular structure is noted in the medial lobe. In the left lung, loss of lung volume and hypoplasia of the main pulmonary artery are seen. Origin: Department of Radiology, University Hospital Dr Peset, Valencia, Spain.
Description: CECT (axial view) shows congenital atresia of the left pulmonary veins. Origin: Department of Radiology, University Hospital Dr Peset, Valencia, Spain.
Description: CECT (axial view) Left lung hypoplasia and a small left pulmonary artery are noted.
Origin: Department of Radiology, University Hospital Dr Peset, Valencia, Spain.
Figure 4

Description: Smooth thickened interlobular septa are observed. Origin: Department of Radiology, University Hospital Dr Peset, Valencia, Spain.
**Figure 5**

**Description:** CECT (MIP reconstruction, oblique view) shows unilateral single pulmonary vein with a meandering course. **Origin:** Department of Radiology, University Hospital Dr Peset, Valencia, Spain.