Arch Bronconeumol. 2020;56(6):390

Sociedad Española de Neumología y Cirugía Torácica SEPAR

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Clinical Image

Right Lung Agenesis Associated with Dextrocardia and Pulmonary Hypertension☆

Agenesia pulmonar derecha asociada a dextrocardia e hipertensión pulmonar

Isabel María Ordóñez Dios,[a,*] Miguel Ángel Rienda Moreno,[b] Javier Lázaro Polo[a]

[a] *Servicio de Neumología, Hospital General Universitario de Ciudad Real, Ciudad Real, Spain*
[b] *Servicio de Radiología, Hospital General Universitario de Ciudad Real, Ciudad Real, Spain*

We report the case of a 37-year-old man diagnosed at birth with right pulmonary agenesis with dextrocardia, operated at the age of 8 years for persistent arteriovenous fistula, and a history of recurrent respiratory infections with bronchial hyperreactivity. Imaging tests (Fig. 1) revealed right pulmonary agenesis with left pulmonary compensatory hyperinflation, anterior transmediastinal herniation, and dextrocardia.

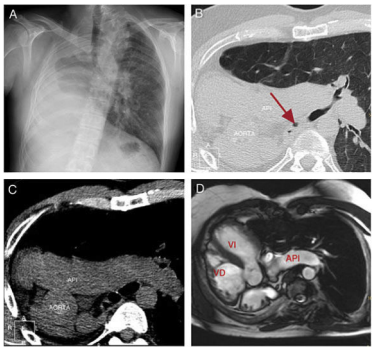

Fig. 1. (A) Chest X-ray, posteroanterior projection, showing mediastinal and left lung shift toward the right hemithorax caused by absence of the right lung (pulmonary agenesis). (B) Tomographic axial slice showing lung parenchyma view. The arrow indicates the rudimentary right main bronchus terminating in the pleural sac. (C) Tomographic axial slice without intravenous contrast showing the shift of vascular structures toward the right hemithorax, and absence of right pulmonary artery. API: Left pulmonary artery. (D) Cine-MR b-FFE oblique axial slice. Dextrocardia, dextro-apex and complete absence of right lung parenchyma are observed. VD: right ventricle; VI: left ventricle.

Pulmonary agenesis is a rare congenital malformation. Congenital anomalies can occur in 3 types:

- Type 1 or agenesis: Complete absence of lung parenchyma and vasculature, and bronchial tree.
- Type 2 or aplasia: Rudimentary bronchi with complete absence of lung parenchyma.
- Type 3 or hypoplasia: Presence of variable amounts of lung parenchyma, bronchial tree, and supporting vasculature.[1]

Its etiology is unknown and appears to be the result of multiple factors, including genetics, vitamin A or folic acid deficiency, and viral infections. Left pulmonary agenesis is the most common (70% of cases), and is associated with a more favorable prognosis, since right agenesis is most often associated with abnormalities, usually cardiovascular and gastrointestinal.[2]

X-ray and chest CT are essential for diagnosis, the latter being the key to confirmation.

References

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☆ Please cite this article as: Ordóñez Dios IM, Rienda Moreno MÁ, Lázaro Polo J. Agenesia pulmonar derecha asociada a dextrocardia e hipertensión pulmonar. Arch Bronconeumol.2020;56:390.

* Corresponding author.
E-mail address: isabelordonezdios@gmail.com (I.M. Ordóñez Dios).

1579-2129/Published by Elsevier España, S.L.U. on behalf of SEPAR.