CLINIC CASE

AGENESIA PULMONAR DERECHA EN PACIENTE PEDIÁTRICO CON SOSPECHA DE ATELECTASIA TOTAL: REPORTE DE CASO

RIGHT LUNG AGENESIS IN A PEDIATRIC PATIENT WITH SUSPECTED TOTAL ATELECTASIS: CASE REPORT

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RESUMEN:

Se reporta el caso de un paciente masculino de 10 meses y 19 días de vida quien ingresa al servicio de urgencias por cuadro clínico de vías aéreas inferiores, asociado a datos de dificultad respiratoria, al examen físico con evidencia de hipoventilación derecha y crepitantes izquierdos, por lo que se realiza imagen radiográfica con presencia de opacidad generalizada en hemitórax derecho, por lo que se decide realiza tomografía de tórax, corroborando agenesia pulmonar; se valora por el servicio de neumología y cardiología encontrando dextrocardia. Generalmente esta patología es poco frecuente y casi siempre requiere tratamiento de soporte.

Palabras claves: Malformación, agenesia, pulmón, pediátrico, reporte de caso

SUMMARY:

We report the case of a male patient 10 months and 19 days old who was admitted to the emergency department for clinical symptoms of lower airways, associated with respiratory distress, physical examination with evidence of right hypoventilation and left crackles, so a radiographic image was performed with the presence of generalized opacity in the right hemithorax, so it was decided to perform chest CT, corroborating pulmonary agenesis; it was evaluated by the pulmonology and cardiology service finding dextrocardia. Generally this pathology is infrequent and almost always requires supportive treatment.

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Keys words: Malformation, agenesis, lung, pediatric, case report

Introducción

Congenital pulmonary malformations are part of the differential diagnosis of extensive radiopaque lesions on chest X-rays in the pediatric age group. (1).

They may be easy to identify but difficult to diagnose (1). Often, they present with symptoms and chest X-ray findings that are similar (1). More invasive studies may be required to identify the malformation, but in some cases, a definitive diagnosis is not made until surgery (1). An organized approach to the evaluation of these lesions is necessary to avoid unnecessary testing (1).

Clinic case

The patient is a 10-month-and-19-day-old male with a history of hospitalization at 20 days of life due to bronchiolitis requiring supplemental oxygen, as well as a history of urinary tract infection during the third trimester of gestation. He was admitted presenting a 5-day clinical course characterized by clear rhinorrhea, sneezing, episodes of non-emetic dry cough that later became emetic, accompanied by signs of respiratory distress (nasal flaring, intercostal retractions). Upon arrival to the emergency department, the patient exhibited respiratory distress; physical examination revealed decreased ventilation in the right lung and crepitations on the left. Initially, severe community-acquired pneumonia was suspected, necessitating high-flow ventilation. A chest radiograph (Figure 1) showed an 80% opacity of the right hemithorax.

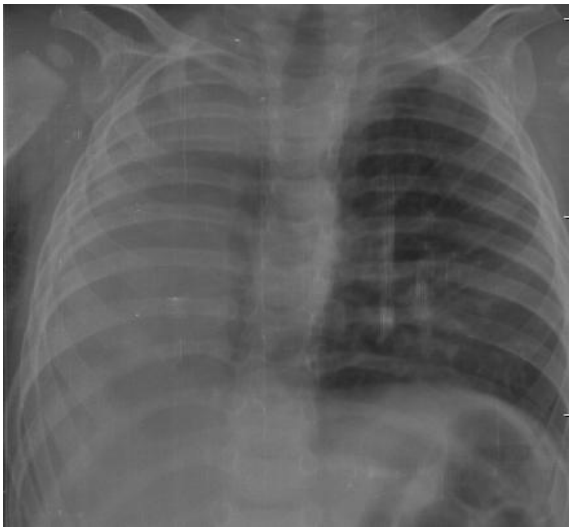

Figure 1. *Chest X-ray: 90% of the right hemithorax is opaque without an aerial bronchogram (Source: ClearCanvas DICOM Viewer – Hospital para el Niño, Toluca)*

Extension laboratory studies (Table I)

	Reports
CBC	Hemoglobin 13.7 Hematocrit 40 Leukocytes 11,950 Neutrophils 55% Monocytes 2% Platelets 477,400 VSG 6
Blood Chemistry	C-reactive protein 0.37 Glycemia 115 Creatinine 0.24 Uric acid 2.7 Urea nitrogen 7.45 Albumin 4.9
Hormones	TSH 0.37 FT4 1.18

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Antibiotic therapy was initiated with cefotaxime (150 mg/kg/day) plus clindamycin (40 mg/kg/day) for 5 days. Additionally, a chest computed tomography (CT) scan was requested, which suggested right atelectasis. The pulmonology service was consulted, and based on the patient's history and imaging studies, they recommended a repeat chest CT with 3D reconstruction (Figure 2).

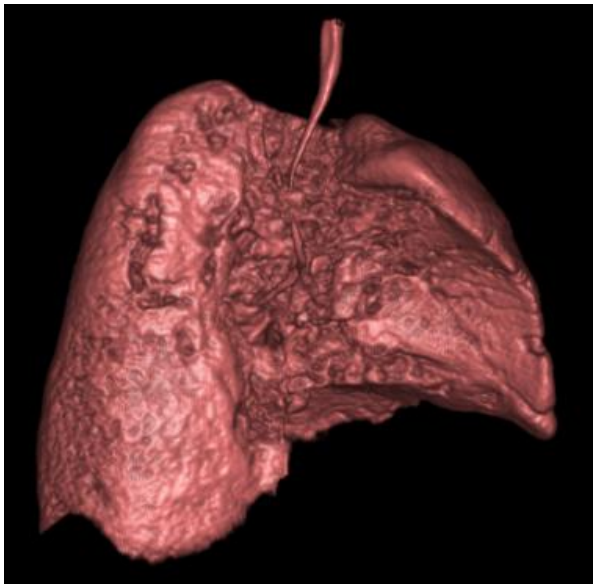

Figure 2. *Chest CT with 3D reconstruction: no right bronchus or right pulmonary branch is evidenced, compensating with hyperinflation of the left lung causing displacement of mediastinal structures to the right (Source: ClearCanvas DICOM Viewer – Hospital para el Niño, Toluca).*

The image revealed absence of the right bronchus, rightward displacement of mediastinal structures, absence of the right pulmonary artery branch, and hyperinflation of the left lung, leading to a diagnosis of right pulmonary agenesis. It was recommended to continue management with inhaled corticosteroids; given the underlying pathology, the condition is associated with poor prognosis and requires a comprehensive evaluation to rule out associated malformations. The cardiology service also evaluated the patient and diagnosed dextrocardia with situs solitus, agenesis of the right pulmonary artery branch and right bronchus, and severe right pulmonary hypoplasia, which did not require treatment. Laboratory findings indicated subclinical hypothyroidism (Table 1). The patient subsequently showed adequate clinical improvement and was discharged home without supplemental oxygen therapy, with planned clinical follow-up in the Pulmonology outpatient clinic.

Discussion

Congenital anomalies of the respiratory system comprise a large number of conditions that can compromise the development of any of its constituent organs (3). Some of these present as characteristic clinical syndromes, while others are considered anatomical variations that do not require treatment (3).

In most cases, it is not possible to determine the triggering cause of the symptoms ; generally, they are caused by two fundamental mechanisms: genetic factors and the influence of surrounding environmental factors (mechanical, physical, chemical, hormonal, and infectious) (3).

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Pulmonary agenesis and hypoplasia are a constellation of pulmonary malformations characterized by lung underdevelopment (1).

In pulmonary agenesis, there is no development of the bronchial tree, lung tissue, or pulmonary vasculature. In pulmonary hypoplasia, there is a decrease in the number and size of bronchial airways, lung alveoli, and pulmonary vasculature (1).

The first case was described by Haberlein in 1887. Most of the subsequently reported cases have shown additional developmental defects involving the skeletal, cardiovascular, gastrointestinal, and urogenital systems (2).

Unilateral pulmonary agenesis has been reported in more than 1,600 cases and is an uncommon alteration, with a described frequency of 0.5 to 1 per 10,000 individuals, resulting from genetic, teratogenic, and mechanical factors (4). There is a female predominance in unilateral agenesis, 1.3:1 (4).

The causes are still unknown, although several hypotheses have been proposed (3):

1. Abnormal development of the first and second branchial arches during embryogenesis.
2. Absence of pulmonary bud development.
3. Teratogenic or vascular injury.
4. Genetic abnormalities involving segment 2p21–p24.
5. Germ plasma defects.
6. Paternal consanguinity.
7. Viral infections.
8. Nutritional factors (vitamin A and folic acid deficiency).

Bilateral pulmonary agenesis is incompatible with life (3). The clinical picture developed by patients with unilateral agenesis can be early respiratory distress ranging from mild to severe, some present with recurrent pneumonias, and in others (3).

The diagnosis is suspected by the absence or decrease of breath sounds in one hemithorax (3).

Chest X-ray shows total opacity in one hemithorax, compensated hyperinflation and herniation of the contralateral lung, retrosternal hyperlucency, and posterior displacement of mediastinal structures (3). The diagnosis is confirmed by performing a computed tomography, ideally with angiotomography or angiography (3).

The differential diagnosis includes total atelectasis of a lung, post-pneumonectomy changes, fibrothorax, foreign body, pulmonary hypoplasia, and hemidiaphragm agenesis (4).

In general, the prognosis of the disease is based on the presence of associated congenital alterations (3). Right pulmonary agenesis has been

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associated with cardiac (14%), gastrointestinal (14%), skeletal (12%), vascular (9%), and genitourinary (9%) malformations (3). Additionally, it carries a worse prognosis due to hemodynamic alterations caused by the displacement of the heart and mediastinum to the right (3).

Conclusión

Recognizing that pulmonary malformations are not frequent, they should be considered in the differential diagnosis in newborns and young children with abnormalities (extensive or total opacities in one or both hemithoraces) on chest X-rays, and that they can be the cause of recurrent pneumonias. Additionally, depending on the clinical history including antecedents, we must make a diagnosis with minimal studies in order not to delay treatment, additionally avoid complications and perform early rehabilitation depending on each specific case.

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