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INTERACTIVE CASE

A child with recurrent pneumonia and persistent pulmonary image, whose diagnosis was performed by ultrasonography. What is your diagnostic hypothesis?

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A five-year-old, female student, referred to the pulmonology outpatient clinic for investigation of repeated respiratory conditions and persistent radiological imaging. The child's caregiver reported recurrent pneumonia episodes, with hospitalization for venous antibiotic therapy when the patient was two and three years old. Simple thoracic radiographies showed persistent hypotransparency images in the lower third of the left hemithorax (Figure 1 a and b), and patient had already been submitted to two thoracic computed tomographies for diagnosis elucidation. Patient reported recurrent cough, related to environmental factors, and with clinical improvement after bronchodilator administration. She denied dyspnea, fever, weight loss, observation of episodes compatible with aspiration of foreign body and contact with chronic coughers or tuberculosis. Gestational and perinatal history had no intercurrences.

Upon examination, the child was in good general condition, eutrophic, afebrile, hemodynamically stable, HR = 88 bpm, RR = 21 irpm and with 95% hemoglobin saturation in ambient air. Heart auscultation showed no alterations. Examination of the respiratory system showed decreased expansibility, decreased vesicular murmur and massive percussion in the lower third of the left hemithorax. Other aspects of physical examination revealed no abnormalities.

Laboratory tests, such as hemogram, lipidogram, coagulogram, electrolytes, renal and hepatic functions, were normal.

What is the most likely diagnosis in a patient with recurrent pneumonia history and persistent imaging?

a) immunodeficiency
b) primary ciliary dyskinesia
c) pulmonary malformation
d) neoplasia
e) chronic pulmonary infection, such as tuberculosis

- Patients with recurrent pneumonia in the same location usually present structural alterations such as bronchial extrinsic or intrinsic obstructions, pulmonary sequelae such as bronchiectasis or malformations. Foreign body is always a hypothesis to be considered, even without an observed aspiration episode.
- Humoral or cellular immunodeficiencies and alterations in respiratory tract defense mechanisms, such as those occurring in cystic fibrosis and primary

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Figure 1 a and b. Simple thorax radiographies with persistent hypotransparency image in the lower third of the left hemithorax.

ciliary dyskinesia, are causes of variable localization pneumonia.

- Tuberculosis is a cause of pneumonia with persistent image and chronic symptoms. Rarely causes recurrent pneumonia.

The presence of a persistent image located in the lower third of the left lung, associated to the fact that the patient was submitted to excessive levels of radiation exposure due to numerous radiological examinations, including two chest scans, initially led us to opt for thorax ultrasonography. This examination revealed a predominantly hypoechoic heterogeneous image on the left pulmonary base, posteriorly located, measuring 7cm x 4cm in its largest diameters and well vascularized. Noteworthy, a large branch originating from the aorta was directed to the lesion (Figures 2 a and b). These findings guided the correct request for computed tomography with contrast and thorax reconstruction in order to expose the patient to minimum possible levels of radiation, still reassuring a correct evaluation. Computed angiotomography confirmed the diagnosis of intralobar pulmonary sequestration (Figure 3 a and b).

During follow-up, atopy-mediated bronchial asthma diagnosis was confirmed (spirometry with grade I DVO, increased IgE and positive skin tests) and treatment with inhaled corticosteroid and symptom control were initiated.

After six months of follow-up, patient was submitted to thoracotomy with lesion excision. Macroscopic appearance and histopathological analysis confirmed the intralobar pulmonary sequestration diagnosis. Patient postoperative evolution was excellent.

The definition "Portion of nonfunctioning lung tissue that does not communicate with the adjacent tracheobronchial tree and receive systemic arterial supply" corresponds to:

a) pulmonary sequestration
b) bronchogenic cyst
c) congenital lobar emphysema
d) bronchial atresia
e) pulmonary aplasia

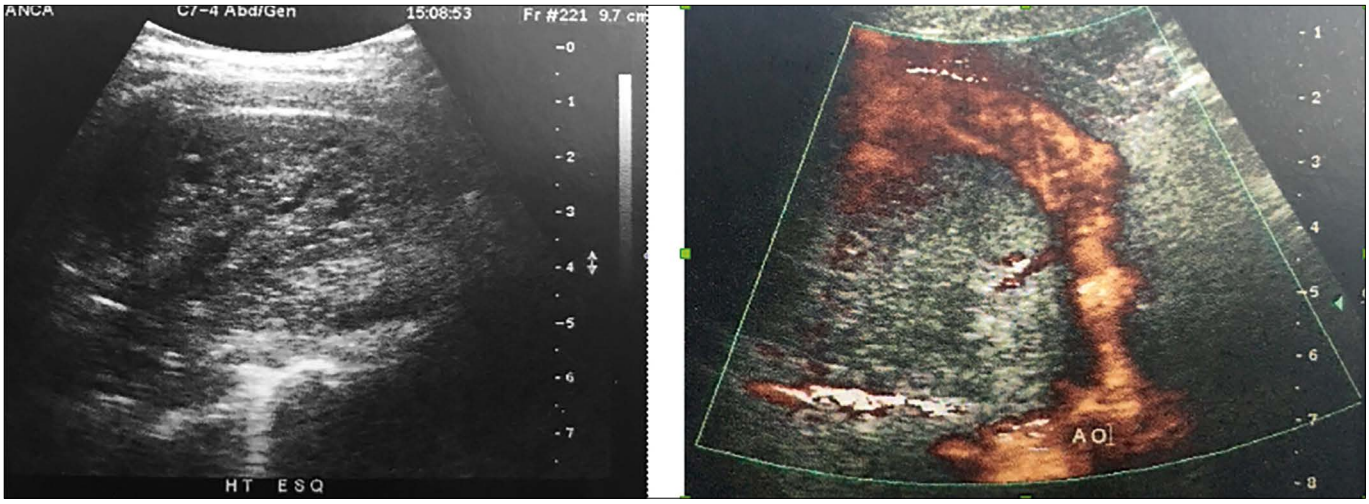
Congenital pulmonary malformations are a heterogeneous group of developmental disorders affecting the pulmonary parenchyma, arterial supply and/or venous drainage. Improvements of imaging methods allowed an increase in the number of diagnoses, better understanding of disease onset, natural history and etiological associations[1].

Current classification is based on the morpho-radiological-pathological findings and proposes the division of these malformations into three groups: pulmonary parenchyma lesions, vascular lesions and mixed lesions[1].

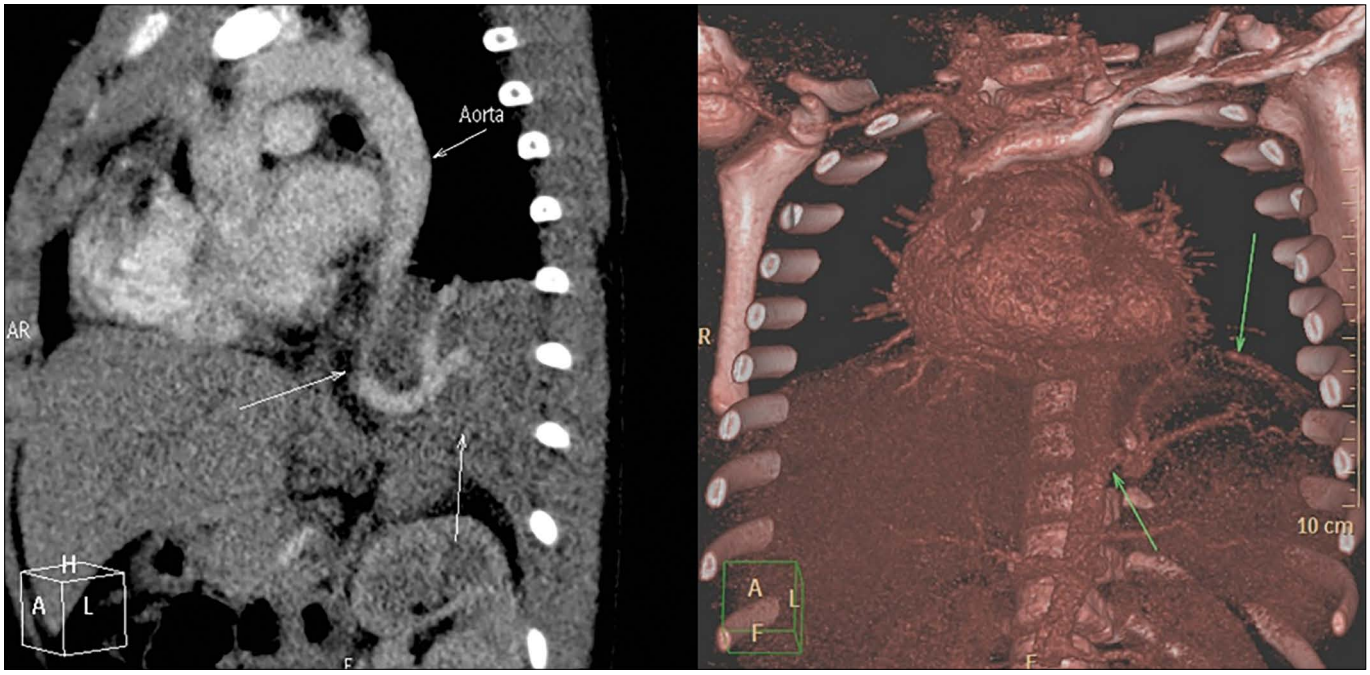
We must also consider that these lesions occur in the context of an embryonic development continuum, at different gestational ages, and may occur in isolation or in association[1].

Definitions of congenital malformations are:

- Pulmonary sequestration: A portion of nonfunctioning lung tissue that does not communicate with the adjacent tracheobronchial tree and receives systemic arterial supply;



Figures 2 a and b. Thoracic ultrasonography showing a predominantly hypoechoic heterogeneous image in the left pulmonary base, posteriorly located, measuring 7cm x 4cm in its largest diameters, well vascularized. Also, images show a large branch originating from the aorta and directed to the lesion.



Figures 3 a and b. Computerized thorax angiotomography confirming the pulmonary sequestration diagnosis.

- Bronchogenic cyst: defect in the ventral tree bud or branch between 26-40 gestational days;
- Congenital lobar emphysema: hyperinflation and distension of one or more lung lobes;
- Bronchial atresia: Obliteration or stenosis of segmental, subsegmental or lobar bronchi at or near its origin;
- Pulmonary aplasia: total absence of lung and pulmonary artery with rudimentary source bronchus[1].

It is characteristic of intralobar pulmonary sequestration:

a) Mandatory intrathoracic arterial supply
b) Venous drainage performed by the pulmonary veins
c) Location in infradiaphragmatic region
d) Isolated pleural lining
e) Presence of intact lung tissue in its interior

- Pulmonary sequestration can be classified as intralobar and extralobar.
- Intralobar pulmonary sequestration, which is the more frequent type of pulmonary sequestration (75% of cases), is covered with the same visceral pleura as the rest of the lung[2].
- Intralobar pulmonary sequestration may receive arterial supply from thoracic or extrathoracic arteries (branches of the descending aorta), but venous drainage occurs through the pulmonary veins[3].
- Extralobar pulmonary sequestration is covered by its own visceral pleura, separated from the rest of the lung. It presents systemic arterial supply (descending aorta or its branches, such as celiac trunk) and usually systemic venous drainage (azygos, portal vein, subclavian, internal mammary)[2].
- In intralobar sequestration cases, pulmonary tissue is in a rudimentary, fibrotic form, and may have cysts in its interior[3].

Intralobar pulmonary sequestration in childhood is more often associated with:

a) Recurrent infection in the same pulmonary location
b) Localized pulmonary bleeding
c) Congenital anomalies of the digestive system
d) Pneumothorax
e) Severe respiratory distress at birth

- Children with intralobar pulmonary sequestration are usually asymptomatic or present recurrent infections in the same pulmonary site, with rare symptoms at birth. In a review of congenital pulmonary malformations cases, Hylas et al. observed history of prior pneumonia in 48.57%[4] of them.
- Hemoptysis episodes are rare in childhood. In some series of adult cases, these episodes occurred as the second most common manifestation (51% of cases), followed by chest pain and recurrent infections[5].
- The association with other congenital malformations is described in both types of sequestration, and may include bronchogenic cyst, bronchial atresia, congenital pulmonary airway malformation (CPAM), Scimitar Syndrome, and communication with the gastrointestinal system. In cases of intralobar sequestration, association may vary between 6 and 12%, with bronchial atresia and CPAM being more frequent[6].
- About 50% of the extralobar sequestration cases are associated with type 2 CPAM or small cysts. This type of sequestration is also more commonly associated with congenital heart diseases, diaphragmatic hernia, and pulmonary hypoplasia[1,7].

The most common location of pulmonary sequestration is:

a) Superior lobes
b) Inferior lobes
c) Medium lobe
d) Lingula
e) Abdomen

- Intrapulmonary sequestration is the most common form and 60% of these cases occur in the posterior basal segment of the left inferior lobe. 98% of cases occur in inferior lobes.
- Patients with extralobar sequestration have focal lung masses in the neonatal period, typically in the lower left hemithorax (95% of cases). Other less common locations are the mediastinum, the pericardium, and the infradiaphragmatic region[8].
- Bilateral involvement is uncommon.
- Regarding sexes, extralobar form affects 4 times more males, while the intralobar form incidence is the same for both sexes[9].
- Extralobular sequestrations may be located outside the rib cage. In the abdomen, attention should be drawn to the region above the left adrenal, and the differential neuroblastoma diagnosis should be performed during gestational ultrasonography[7,10].

The recommended complementary exam for diagnosis confirmation in these cases is:

a) Thorax ultrasonography
b) Thorax angiotomography
c) Arteriography
d) Transthoracic echocardiogram
e) Thoracic radiography with contrasted esophagus

- During the investigation of suspected pulmonary sequestration case, imaging examinations have two main objectives: to discard the presence of other pathologies and to characterize the presence of anomalous arterial supply[11].
- Ultrasonography is excellent for initial lesion evaluation, especially in neonates and infants. It is a non-invasive technique, available even at the bedside, without exposure to radiation and that can reveal from homogeneous lesions to heterogeneous mass with cysts. Arterial supply and, possibly, venous drainage can be demonstrated with color Doppler[3].

- In recent years, multidetector tomography associated with angiotomography has been the exam of choice for evaluation of pediatrics congenital malformations. The reconstruction allows an overall sequestration evaluation and is the recommended exam for diagnostic confirmation and detailed anatomical analysis[1,3].
- The tomographic evaluation with accuracy of pulmonary malformations depends on basic parameters, such as milliamperage, kilovolt peak, table speed, collimation detector and reconstruction thickness, adjusted for diagnostic suspicion in order to obtain the best possible definition of the image, still preserving the patient from exposure to high radiation doses[1,12].
- Magnetic resonance imaging (MRI) is a useful method for the evaluation of solid and vascular lesions, providing images without exposure to radiation. Nevertheless, it has limitations in the study of pulmonary parenchyma and in the time required for examination, which for pediatric population means sedation[2].
- Transthoracic echocardiogram does not allow the pulmonary sequestration diagnosis, however, it should be requested in order to discard structural cardiomyopathies hypothesis, as well as the evaluation of possible hemodynamic repercussions[3].
- Thoracic radiography with contrast-enhanced esophagus is indicated in the suspicion of concomitant malformations of the digestive tract[3].
- In this scenario, the use of color Doppler ultrasonography as an initial examination in diagnostic investigation of persistent pulmonary imaging in the left inferior lobe was noteworthy, since it is a widely available, noninvasive and free of ionizing radiation technique, that can successfully reveal pulmonary sequestration and associated anomalous arteries[2].

Regarding intralobar pulmonary sequestration treatment, the correct alternative is:

a) surgical resection should be performed regardless of the symptoms.
b) pneumectomy is the surgical procedure of choice and aims to prevent the evolution to future neoplasia.
c) Embolization of anomalous vessels through catheterization followed by coil implantation is possible.
d) conservative treatment can be chosen, even in hybrid lesions.
e) extra-numeral anomalous arterial supply is a contraindication for surgery.

- Surgical resection is invariably the treatment of choice for these intrathoracic lesions. In the case of intralobar sequestration, a lobectomy or segmentectomy is usually performed, while in extralobar cases it may be possible to remove only the lesion[13,14].
- Sequestration cases can have complications with recurrent infections, massive bleeding, and pneumothorax. Some studies report the possibility of association or even progression to neoplasms, especially in cases of radiologically hybrids lesions[14].
- In cases of small and anatomically well-studied lesions, embolization of anomalous vessels through catheterization followed by coil implantation has been described since 1983 as an alternative to the surgical procedure. The first descriptions of these procedures were for cases of extralobar sequestration, but recent studies have been successful in adult and children intralobar cases[14].
- In cases in which embolization is chosen, lesion involution should be monitored[13,15].

CONCLUSIONS

Respiratory infections are the major cause of illness in children. Pneumonia risk factors include children with less than five years of age, prematurity and malnutrition. According to the literature, the occurrence of two or more episodes within 12 months indicates need for evaluation.

In the case of a patient in diagnostic investigation, with no clinical evidence of respiratory infection, no history of foreign body aspiration or tuberculosis and presenting condensation in the posterior segment of the left inferior lobe on thorax radiography, pulmonary malformation is one of the diagnostic hypotheses that should be considered.

Request of appropriate imaging examinations should be emphasized, to allow the correct etiologic diagnosis without exposing the pediatric patient to excessive radiation. The location of the persistent image in inferior lobes, especially the left, indicates pulmonary sequestration as an important diagnostic hypothesis.

The case reported shows that ultrasonography examination performed by experienced professionals can be used as an initial strategy in the diagnostic investigation of pulmonary malformations, not only during the prenatal but also in the postnatal period.

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