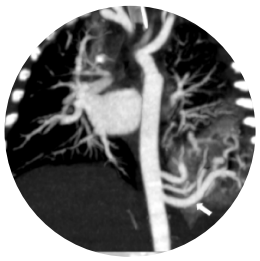

Pulmonary sequestration with three aberrant arteries: a case report

Secuestro pulmonar intralobar con tres arterias aberrantes. Presentación de un caso

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Summary

Pulmonary sequestration (PS) is a congenital anomaly of the respiratory tract in which the lung tissue has no communication with the tracheobronchial tree and receives its supply from aberrant systemic arteries. Despite being a known pathology, it still presents gaps in the therapeutic approach, especially when there are unusual presentations. We describe the case of a newborn with intralobar PS with three aberrant arteries.

Resumen

El secuestro pulmonar (SP) es una anomalía congénita del tracto respiratorio, que consiste en que una zona de tejido pulmonar no tiene comunicación con el árbol traqueobronquial, el cual es irrigado por arterias aberrantes originadas de la circulación sistémica. A pesar de ser una patología conocida, aún hay vacíos respecto al abordaje terapéutico, especialmente cuando se encuentran presentaciones poco usuales. Se describe el caso de un recién nacido con SP intralobar con tres arterias aberrantes.

Case description

We present the case of a male newborn, with suspected pulmonary malformation since 22 weeks of gestation evidenced in prenatal ultrasound extra-institutional, with findings suggestive of bronchopulmonary sequestration versus congenital malformation of the pulmonary airway. Vaginal delivery with term newborn, without complications and transferred to the neonatal intermediate care unit. Chest tomography with angiotomography was performed to characterize the suspected congenital anomaly.

A thoracic angiotomography with arterial and venous acquisition phases was performed (figures 1 and 2), in which an area of increased density secondary to consolidation was visualized in the lung parenchyma of the posterior basal segments of the left lower lobe, following the morphology of the lung parenchyma, without compromising the expansion and apparently involving the posterior and lateral basal segment of this lobe; It is accompanied by adjacent "ground-glass" opacities and it is not possible to identify a clear transition zone between the healthy and diseased parenchyma; it is associated with bronchial dilatations in its interior, which show no connection with the rest of the tracheobronchial tree. In the arterial phase there are three arteries originating in the descending thoracic aorta that irrigate the affected segment and in the venous phase there is drainage to the ipsilateral inferior pulmonary vein.

The patient was taken to thoracoscopy; however, due to his hemodynamic instability when selective intubation was attempted, thoracotomy with resection of the affected segment was performed. Anomalous pulmonary tissue was found adhered to the left lower lobe, where the two main large caliber nutritional arteries are ligated, as well as the venous drainage to the central circulation.

Discussion

Pulmonary sequestration (PS) is a congenital malformation of the respiratory tract, consisting of pulmonary tissue not communicating with the tracheobronchial tree, which receives its irrigation from one or more aberrant systemic arteries (1). It represents the second most common congenital pulmonary malformation, with an incidence between 0.1-6.4%, and it also has an intralobar or extralobar presentation (1). Symptoms vary according to its presentation, the most common are dyspnea, feeding disturbances, *hydrops fetalis*, recurrent infections, congestive heart failure and, less frequently, pneumothorax, hemoptysis and hemothorax; between 10% and 15% are asymptomatic (2).

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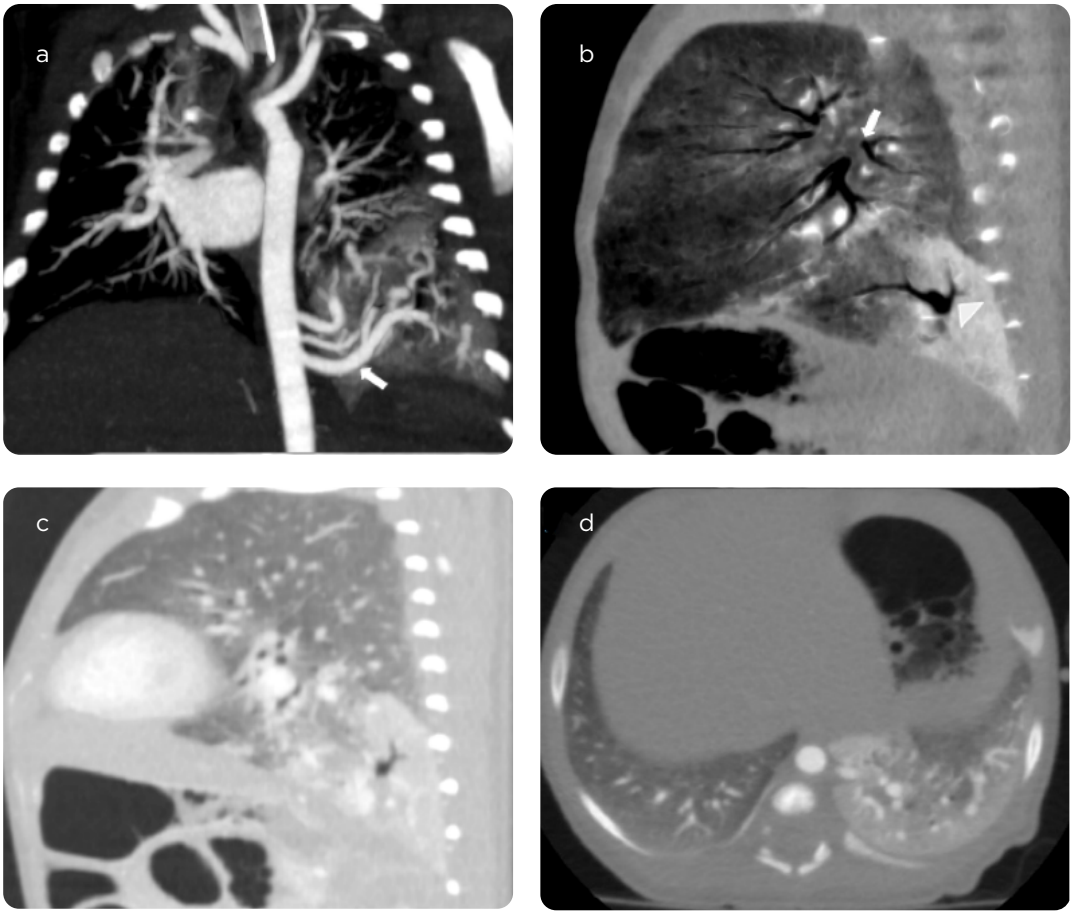

Figure 1. Chest CT with contrast medium, acquisition in arterial phase. a) Coronal reconstruction in maximum intensity projection. Shows the three aberrant arteries originating in the descending aorta (arrow). b) Sagittal reconstruction in minimum intensity projection, showing absence of communication of the sequestration (arrow) with the bronchial structures (arrow head). c) Sagittal reconstruction. d) Axial in lung window: area of consolidation in the basal segments of the left lower lobe, well delimited, with bronchial structures in its interior without connection with the bronchial tree.

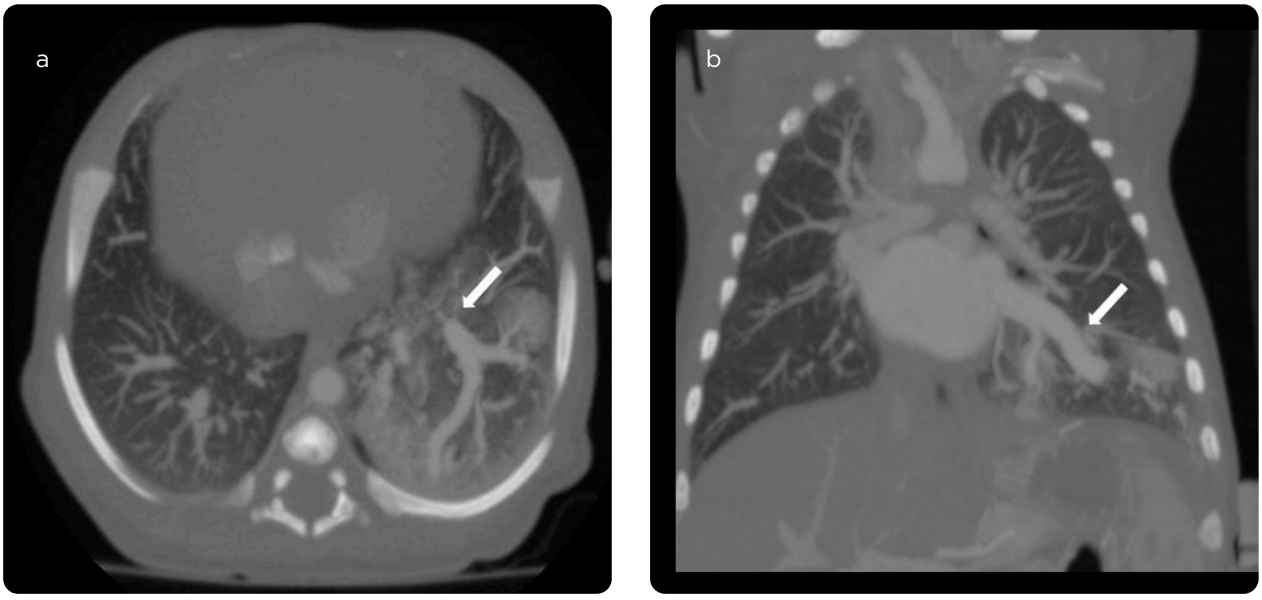

Figure 2. Chest CT with contrast medium, acquisition in venous phase. a) Axial reconstruction. b) Coronal reconstruction. Venous drainage to an inferior lobar vein with connection to the left atrium.

Differential diagnoses include other congenital pulmonary malformations, such as congenital malformation of the pulmonary airway, bronchogenic cyst, focal bronchiectasis and congenital lobar emphysema (2).

Intralobar sequestration is that which shares pleura with the healthy lung and has venous drainage to the pulmonary veins (1). Until now, the theory suggests a congenital origin of the primitive intestine, before the formation of the pleura; however, more recent literature has raised the possibility of an acquired cause, given its greater presentation in children and adolescents (1). On the other hand, extralobar sequestration has its own pleura and both its irrigation and venous drainage depend on the systemic circulation; the most accepted theory for this subtype has been the congenital origin, with an accessory pulmonary yolk of caudal location (1).

Diagnostic imaging plays a fundamental role in the diagnosis. Ultrasonography is recommended as the first modality for the approach of thoracic masses in infancy and prenatal life (1). Since the 1980s, ultrasound findings for PS have been described that include a well-defined echogenic mass and may occasionally include cystic images, features that allow differentiation of PS from pleural effusion or diaphragmatic hernia (3). However, its performance in identifying nutritional vessels is not yet defined, so it should not be the only test performed for decision making (4).

Computed axial tomography (CT) or magnetic resonance imaging (MRI) are the images of choice for the characterization of sequestrations (5). Intralobar sequestrations will be visualized as masses or consolidations with homogeneous or heterogeneous density, with or without cysts, which may be filled with air and/or fluid. Extralobar sequestra tend to be rounded or pyramidal masses in the pleural space near the posteromedial aspect of the ipsilateral hemidiaphragm, and since they have no communication with the tracheobronchial tree they are collapsed, which increases the density of the parenchyma in "ground glass" or consolidation (5). On the other hand, MRI also allows differentiating the relationship of the PS with the parenchyma and pleura, with a good resolution of soft tissues as well as a better distinction between solid and cystic components, with a good capacity to identify the nutritional artery and venous drainage (4).

Intralobar pulmonary sequestration represents 75% of the forms of appearance of this pathology, and its arterial supply is provided by the descending aorta (72%), the abdominal aorta (21%) and the intercostal arteries (3%), respectively, in order of frequency (6). They usually have a single nutritional artery. Less frequently, cases with two nutritional arteries have been documented in 16% of them (1). Another investigation that sought to describe the characteristics of 2625 cases of pulmonary sequestration found that patients with more than two arteries, such as the one presented in this report, accounted for only 4.92%. This same study found that the most frequent venous drainage was to the pulmonary veins in more than 90% of the cases, followed by the azygos vein with 4% (7).

Treatment is controversial due to the fact that in most cases it is unknown how this disorder will develop over time; however, given the high risk of complications -especially recurrent infection- surgical resection of the affected segment is recommended, which is the preferred therapeutic management (2). However, bleeding from these aberrant vessels can be lethal if it is not controlled, so it is necessary to find other alternatives of care to avoid these possible risks (8).

In 1998 the first endovascular therapy for the management of PS was performed, which subsequently reported a series of complications with this exclusive management such as incomplete embolization, recanalization or even recurrent infections or hemoptysis by not completely excluding the non-functional parenchyma (8). However, a shorter hospital stay has been demonstrated in endovascular management (9.6 versus 4.7 days), with 7% of patients presenting complete regression of the PS and 60% incomplete regression, in addition to greater blood loss in patients managed surgically mainly due to lack of identification of the aberrant circulation before the procedure (9). However, there is little literature regarding treatment methods and long-term evolution of patients (9).

So far, it is suggested to perform hybrid management, surgical plus endovascular, mainly in patients with high risk of hemorrhage and recurrence, either by multiple nutritional vessels or of a large caliber (without defining a standardized measure yet) and in the case of large pulmonary lesions (> 3 cm) (8). The minimally invasive approach via video-assisted thoracoscopy is recommended because it has better results than traditional open management; however, it has been relegated by surgeons for fear of injuring the vessels that irrigate it, so previous endovascular management could be a solution to reduce this type of complications and increase the number of more conservative interventions (10). The literature on hybrid treatment in these patients is scarce, so additional studies are required to standardize indications and management recommendations, as well as to evaluate the long-term evolution of patients with PS who undergo this procedure.

Conclusion

We report a patient with an intralobar pulmonary sequestration with three aberrant arteries with prenatal suspicion and postnatal confirmation of the diagnosis by chest angiotomography. Her treatment was surgical. Hybrid management of pulmonary sequestration with multiple nourishing arteries seems to have fewer complications, such as bleeding or incomplete resection; nevertheless, in clinical practice it is not usual probably due to the little information that exists on the subject, especially in long-term evolution. It is important to point out the gaps that remain in the literature regarding hybrid management and to highlight the importance of standardizing this procedure.

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