CASE REPORT
Pre-natal diagnosis of pulmonary sequestration
Diagnóstico pré-natal de sequestração pulmonar

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Received, 2023, August 1st; accepted 2023 sept 10
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How to cite
Saad TA, Couto JCF, Baylon e Silva MH, Alves FMT, Villela OCC. Pre-natal diagnosis of pulmonary sequestration. JODI 2023;1:e0230006

Abstract
Pulmonary sequestration consists of a rare lung malformation, characterized by a mass of non-functional lung tissue, which does not maintain normal communication with the tracheobronchial tree and whose blood supply is from an anomalous systemic artery. Its prenatal diagnosis is still rare, usually performed in centers of reference by trained professionals. This is a case report of a prenatal diagnosis of pulmonary sequestration, correlating the pre and postnatal findings in different methods of diagnostic imaging.

Keywords: lung malformation; prenatal diagnosis; ultrasonography; magnetic resonance imaging

Resumo
O sequestro pulmonar consiste em uma malformação pulmonar rara, caracterizada por uma massa de tecido pulmonar não funcional, que não mantém comunicação normal com a árvore traqueobrônquica e cujo suprimento sanguíneo é proveniente de uma
artéria sistêmica anômala. Seu diagnóstico pré-natal ainda é raro, geralmente realizado em centros de referência por profissionais capacitados. Apresentamos um relato de caso de diagnóstico pré-natal de sequestro pulmonar, correlacionando os achados pré e pós-natais em diferentes métodos de diagnóstico por imagem.

**Palavras-chave:** malformação pulmonar; diagnóstico pré-natal; ultrassonografia; imagem de ressonância magnética

**Case report**

33 years old patient, first pregnancy, had an ultrasonography (USG) routine performed in the 22\textsuperscript{nd} week of gestation that showed single fetus, male, presenting a triangular hyperechogenic image at the left lung base, measuring 1.6 x 3.3 cm (Figure 1A), with individual vascularization from the aorta (Figure 1B), consistent with pulmonary sequestration. After two weeks, the control USG showed slight image increase, measuring about 2.8 x 3.4 cm. No other associated changes were observed.

At 29 weeks of gestation, a magnetic resonance imaging (MRI) was performed and showed a triangular formation on the left lung base measuring 3.6 x 2.4 x 2.6 cm (Figure 2). She underwent cesarean section at 39 weeks without complications. At the postpartum a computed tomography (CT) and subsequently a computed tomography angiography (CTA) were performed, which confirmed the prenatal diagnosis (Figure 3). The child is currently being followed up and remains asymptomatic and the image is stable.

*Figura 1* - Ultrasonography showing triangular hyperechogenic image on the left lung base (1A) with individualized vascularization from the aorta (1B)
Discussion

Pulmonary sequestration is the second most common pulmonary malformation, yet it is a rare disease [1], representing about 0.15 to 6.4% of all congenital lung malformations [2]. It is characterized by a mass of non-functional lung tissue, which does not maintain normal communication with the tracheobronchial tree [1-4], with anomalous systemic arterial blood supply [1,2,5]. This irrigation can be derived from the abdominal aorta, subclavian artery, and the renal artery, however, over 80% of cases are irrigated by descending thoracic aorta [6].
From the presence or absence of its own envelope, it was possible to distinguish two subgroups of pulmonary sequestration: the intralobar (IPS) and the extralobar (EPS) [7,8]. The EPS has independent pleural lining of the normal lung [7,9]. The arterial supply in most cases is from the abdominal aorta or from one of its branches [10]. Its most common location is the posterior thoracic region [9], and in 80% of cases are on the left side [6]. The association with other anomalies such as pectus excavatum [1], diaphragmatic hernia, hydropsy, cystic adenomatoid malformation of the lung, gastric or colonic duplication, tracheoesophageal fistulas, spinal and cardiac anomalies are reported in about 60% of the cases [9].

The IPS is defined as parenchymal lung segment that shares the same visceral pleural lining of a normal lung [11], but cannot connect to the tracheobronchial tree [10]. The most frequent location is the posterior lateral segment of the left lower lobe [2,10]. The blood supply is given by anomalous systemic arterial circulation, represented in 75% of cases by the aorta, and in the remaining 25% by the thoracic or abdominal vessels [2]. Unlike the EPS, 90% of cases of IPS are presented as an isolated finding, without any correlation with other sonographic changes [2].

Due to the lack of specific symptoms, the diagnosis is usually late, after birth, and the prenatal diagnosis is even more rare and challenging, with few cases reported.

The prenatal diagnosis of PS can be made through routine obstetric ultrasound, especially from the second trimester of pregnancy [2]. The association with Doppler can increase diagnostic accuracy, as it enables the identification of an artery from a systemic origin promoting vascularization of the sequestration [1]. This finding on the dopplerfluxometry is considered the main criteria for the diagnosis of PS [2].

The sonographic diagnosis of PS in the prenatal period should be expressed by the presence of an echogenic intrathoracic mass, usually with a pyramidal shape [2], which is likely to trigger mediastinal shift, with non-pulsatile characteristic, irrigated by systemic artery [9]. Other possible findings associated with this lung abnormality include pleural effusion, polyhydrarnnios and hydropsy [9].

The fetal MRI is important in the evaluation and differentiation of fetal thoracic masses (1), and its findings associated with ultrasound in this case helped composing the diagnosis of fetal pulmonary sequestration.

The main differential diagnosis should be made with cystic adenomatoid malformation of the lung (CAML), which has higher incidence than PS [2]. Besides the CAML, diagnostic hypotheses as teratoma, neuroblastoma, diaphragmatic hernia, laryngeal-bronchial atresia, congenital lobar emphysema, pulmonary arteriovenous malformation and bronchial mucus plug should be considered [2,9].
The evolution of PS depends on its type (IPS or EPS), location, presence of other associated anomalies and hydropsy [2,9]. Hydropsy is the main criteria for a poor prognosis, followed by pulmonary hypoplasia, ascites and polyhydramnios [2]. Otherwise, in the absence of pulmonary hypoplasia and hydropsy, the prognosis is usually good [2].

Conclusion

The prenatal diagnosis of PS is rare, being discovered especially after the second trimester of pregnancy. Ultrasonography is the method of choice for being harmless, quick and at a low cost, but the examiner experience is fundamental for the accuracy of the diagnosis. Fetal magnetic resonance imaging may be another option for prenatal diagnosis.

The early diagnosis can allow the early initiation of a targeted monitoring and treatment, contributing to the quality of life of the patient.

References


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