

Prenatal Diagnosis of an Extralobar Infradiaphragmatic Pulmonary Sequestration: A Case Report and Review of the Literature

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Abstract

Pulmonary sequestrations (PSs) are masses of non-functioning pulmonary tissue. Most PSs are situated in thoracic cavity; very rarely PSs are located below the diaphragm, particularly in the left suprarenal area. A 33-year-old primigravida underwent ultrasound scan at 24+2 weeks of gestation. Ultrasonography showed a hyperechogenic mass situated below the diaphragm. Color Doppler Flow Imaging (CDFI) showed the supplying vessels within the mass originated in the abdominal aorta. The fetus was preliminarily diagnosed with infradiaphragmatic PS. The patient opted to terminate the pregnancy. She underwent odinopoeia to deliver a male fetus who underwent post-mortern examination confirmed prenatally diagnosed abnormalities.

Keywords: Pulmonary sequestrations; Infradiaphragmatic; Ultrasonography; Color Doppler flow imaging

Introduction

Pulmonary sequestrations (PSs) are some rare congenital anomalies consisting of a non-functioning bronchopulmonary mass separated from the normal tracheobronchial tree, which receives arterial blood supply directly from the systemic arteries [1]. The incidence of PSs has been estimated at 0.15% to 1.7% [2]. PS often presents as an incidental mass on radiographic examination. The question of malignancy develops, and evaluation often leads to surgery [3]. Among this group of anomalies, more than 90% are found in the thorax, and less than 10% of them arise under the diaphragm, mainly in the left upper abdomen [4]. The availability of accurate fetal images, basically ultrasound and Color Doppler Flow Imaging (CDFI), has allowed the detection of these anomalies during prenatal life with increasing frequency. The analysis of the images helps in the differential diagnosis, avoiding the need for invasive procedures. We present a rare case of subphrenic fetal PS in a 33-year-old primigravida.

Case Report

A 33-year-old primigravida underwent routine ultrasound scan at 24+2 weeks of gestation estimated by last menstrual period. Ultrasonography showed a $2.10 \times 1.70 \times 1.50$ cm hyperechogenic mass situated at fetal adrenal area upper right left kidney. The hyperechogenic mass was quasi-circular, had homogeneous internal echo and clear boundary with adrenal gland (Figure 1A). CDFI showed the supplying vessels within the mass originated in the abdominal aorta (Figure 1B). Thus, the results of ultrasonography and CDFI showed that there was a

hyperechogenic mass, its supplying vessels originated in the abdominal aorta, at left adrenal area of the fetus, which suggests the fetus had infradiaphragmatic pulmonary sequestration.

Then, the patient underwent further observation. At 26 weeks of gestation, she performed a detailed ultrasound examination again revealed that the hyperechogenic mass did not have the tendency of shrink or spontaneous regression, which suggested that the mass did not be excluded adrenal tumors. After the patient was informed of the prognosis of the syndrome, she opted to terminate the pregnancy. She underwent odinopoeia via craniotomy to deliver a male fetus who underwent postmortern examination that revealed a mass situated behind the pancreas and upper right of left adrenal area, which confirmed prenatally diagnosed abnormalities (Figure 2A). The weight of the mass was 7 g. The diolame of the mass was intact and the surface was smooth. The histologic study (HE dye) of the specimen revealed the presence of disorganized lung tissue (Figure 2B).

Discussion

During the fourth week of gestation, the laryngotracheal bud arises from the ventral wall of the primitive foregut. It bifurcates dozens of times, developing the tracheobronchial tree and the pulmonary parenchyma, coupled with a similar branching process of the primitive lung artery. For unknown reasons, abnormal accessory buds may emerge from the primitive foregut, developing disorganized lung-resembling structures. Usually these structures are not connected to the main airway and, since they emerge far from the pulmonary artery and acquire systemic blood supply [5]. These congenital abnormal non-functioning structures are known as PSs. Some of these abnormalities are embedded within a normal pulmonary lobe, sharing its visceral pleura; they are classified as intralobar PSs. On the other hand, those



Figure 1: The results of ultrasonography and CDFI images. (A): Ultrasound image showing a $2.10 \times 1.70 \times 1.50$ cm hyperechogenic mass situated at fetal adrenal area upper right left kidney. The white arrow points to the mass. (B): CDFI showed the supplying vessels within the mass originated in the abdominal aorta.

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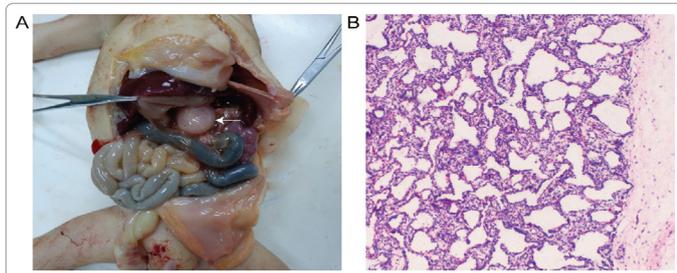


Figure 2: The results of the post-mortem examination and the histologic study of the specimen. (A): The post-mortem examination revealed a mass situated behind the pancreas and upper right of left adrenal area. The white arrow points to the mass. (B): The histologic study (HE dye) of the specimen revealed the presence of disorganized lung tissue.

that are completely isolated from the pulmonary lobes are regarded as extralobar PSs.

Depending on the level of the foregut at which they emerge, extralobar PSs may be supradiaphragmatic or, rarely, infradiaphragmatic (i.e., intraabdominal), with an approximate ratio of 9 to 1, respectively. According to several reports, intraabdominal pulmonary sequestrations (IPSs) are typically situated in the left suprarenal area [4,6-8]. Extralobar PSs are associated with other congenital malformations in more than 50% of cases, such as congenital diaphragmatic hernias, congenital adenomatoid malformation type II, and congenital heart disease [9].

The current available accurate fetal images have allowed the detection of these abnormalities during prenatal life with increasing frequency. Gestational age at diagnosis ranges from the third to eighth month. Gestational age at diagnosis in the case reported in this paper was sixth month (24+2 weeks). IPS is only one of the several possible diagnoses of a suprarenal mass and there is a difficulty distinguishing the mass from an adrenal tumor [10,11]. Ultrasonography is usually method to diagnose PSs that are described as hyperechogenic lesions, with or without cystic components. Although the presence of systemic arterial flow on CDFI scan is the most distinctive feature, its sensitivity is low. Further evaluations are suggested a magnetic resonance imaging (MRI) or computer tomographic (CT) scan [4]. The determination of the levels of tumor markers in urine is a key step in the diagnostic process of a suprarenal mass of unknown origin.

Conclusion

Although the fetus in this case report had no other congenital malformations, it is well documented in the literature that PSs are highly associated with them. Diaphragmatic hernia and cardiovascular anomalies are the most frequently reported [9]. Because IPSs arise from the primitive foregut, a communication with the digestive tract may exist and must be excluded [6].

Although the natural history of IPS is not completely known, surgical excision is the treatment of choice. Complete excision also eliminates the potential risk of high-output congestive heart failure and allows a thorough microscopic examination of the specimen. Several reports have showed that PSs can occur complete spontaneous regression [12,13]. Once proved that the lesion is unique and that the images have the described features, a confident diagnosis of IPS can be made, the fetus can be spontaneously delivered and surgery can be carried out after birth and odinopoeia can be avoided.

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