THE SIGNIFICANCE OF ULTRASOUND PRENATAL DIAGNOSIS IN OPTIMAL THERAPY OF THE NEWBORN BASED ON A CASE OF PULMONARY SEQUESTRATION

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ABSTRACT

Pulmonary sequestration is a rare congenital lesion in which nonfunctional lung tissue lacks connection with the tracheobronchial tree and receives systemic blood supply. It can be accompanied by free fluid in the pleura, lung hypoplasia, circulatory insufficiency, polyhydramnios or even hydrops fetalis. For this reason early and accurate diagnosis is required. We present a case of a fetus which developed hydrothorax in the 31st week of gestation. Ultrasonographic examination revealed a hyperechogenic mass in his chest that was classified as pulmonary sequestration. Thoracocentesis was performed twice (in 33th and 35th week of gestation) and preterm labor took place, due to preterm premature rupture of membranes, in 35+5 week of pregnancy. The newborn presented respiratory distress and was admitted to the Neonatal Intensive Care Unit (NICU). The postnatal diagnosis of the lesion was difficult and it took several days to confirm the prenatal finding. We emphasize the importance of cooperation between obstetricians and neonatologists, that consist of complete visualization of the fetal anatomy, proper classification of the anomalies and successful final management not only during pregnancy but also after the delivery.
ulmonary sequestration (known as bronchopulmonary sequestration = BPS) is a congenital malformation, where part of lung tissue lacks connection with the tracheo-bronchial tree. Also, arterial blood supply is incorrect – the lesion receives blood from systemic circulation. The nutrient arteries come from the thoracic aorta in up to 74% of cases, but they can also originate from abdominal aorta or its branches [1]. Other origins of the vessels are extremely rare but even coronary arteries have been mentioned in the literature [2]. The venous blood flow depends on the type of sequestration. We can divide BPS into extralobar (ELS) and intralobar (ILS). ELS, sometimes known as accessory lung, is a mass that develops outside the regular lung and has its own pleura. It drains blood into the systemic venous system (azygos or portal vein). ILS is covered by normal lung tissue or fragmentarily by visceral pleura. The venous blood from that kind of lesion flows back into pulmonary veins. Some authors distinguish a third type, where abnormal lung tissue is linked to the gastrointestinal tract [3]. The incidence of BPS ranges from 0.1 to 6.5% of all congenital pulmonary malformations [1]. Ultrasonographically we observe fetal lung hyperechogenicity with anomalous blood supply, that has to be visualized in Color Doppler as a pathognomonic syndrome for BPS [1, 4]. BPS can be asymptomatic till adulthood but it can also lead to severe complications during pregnancy. Usually, it is fetal hydrothorax with lung hypoplasia, circulatory insufficiency, and polyhydramnions but may even present as hydrops fetalis, which is most severe for further prognosis [5].

Prenatal ultrasonography (USG) allows the visualization and diagnosis of fetal malformation during intrauterine life. Each lesion should be diagnosed and classified accurately. In the literature, we can find numerous papers about prenatal diagnostic tools and further therapy and some of them underline the need for special neonatal care after delivery.

In the presented case, we would like to highlight the significance of cooperation between specialists that allows to carry out proper in- and ex-utero management.

CASE REPORT

The 28 year-old primigravida was admitted to the hospital in the 31st week of gestation due to fetal hydrothorax, that had been observed in routine prenatal USG. On admission, USG examination revealed left-sided hydrothorax but also independent, hyperechogenic piece of the lung within the left supradiaphragmatic area in the fetus. The lesion was relatively large, measuring 34x19mm (Fig. 1 and 2.). Color Doppler revealed abnormal perfusion of the lesion, coming from systemic circulation (Fig. 3 and 4.). Diagnosis of BPS was established. The mediastinum was shifted to the right side of the chest, but the heart was of normal size and no signs of heart failure were observed. The anatomy of remaining organs was normal.

In the 33rd week of gestation, thoracocentesis was performed because of a sudden increase of the fluid within the pleural cavity. Sixty eight ml of yellowish, translucent fluid was drained and relaxation of the left lung was visualized right after the procedure. The thoracocentesis needed to be repeated two weeks later, because of increasing amount of fluid - 90 ml of fluid, similar to the previous one, were evacuated. Five days later (in 35+5 week of gestation) preterm premature rupture of membranes took place and the mother had regular contractions, so she delivered vaginally a female newborn weighing 2680 grams. The newborn got 8, 10 and 8 points in Apgar score at 1st, 5th and 10th minute of life respectively. The pH of umbilical vein blood was within normal values. Despite the good condition at birth, the newborn was admitted to Neonatal Intensive Care Unit (NICU), because of significant respiratory distress. Intermittent mandatory ventilation was used. X-ray performed on the first day of life revealed fluid in the left pleural cavity, that caused mediastinal shift into the right. Pleural drainage was used, but the cause of fluid accumulation in the pleura was not visualized.

Due to prenatal USG finding contrast-enhanced computed tomography (CT) was performed. It showed atelectasis in left lower lung lobe (segments IX and X) and presence of fluid in the pleural cavity. Mediastinum was again dislocated to the right. Continuous drainage was used and differential diagnosis between lung sequestration and compressed lung tissue was performed.

The X-ray in the 5th day of life revealed complete segment X atelectasis and only a trace of fluid in the left pleural cavity, so the drain was removed. On 8th day of life the condition of the newborn was stable, so she was extubated. In the following days, the patient did not need oxygen and no dyspnea was observed, but baby stayed at NICU, due to further diagnostics of atelectatic lung tissue.

After 2 weeks the fluid was visualized again. X-ray indicated a thymoma or mesothelioma, but lung USG performed on the same day revealed a hyperechogenic tumor measuring 28x38mm. It was located in the same area as the lesion observed prenatally – at the base and posterior part of the left lung. The image corresponded to lung sequestration or teratoma, that was considered due to significantly high alpha-fetoprotein (AFP) level in baby’s blood (1210ng/ml). Oncological and thoracoscopic consultations were obtained. As a consequence thoracic magnetic resonance imagining (MRI) was performed and a tumor of the left lung was visualized, with the largest diameter of 4 cm.

On the 29th day of life, the patient was transferred to Oncology Department. Based on prenatal examinations no neoplastic tumor was excluded and contrast-enhanced CT was performed again to visualize lung sequestration. The girl was referred for surgery which took place in the 5th week of life. The lesion was removed entirely and resected tissue.

The patient was discharged one week post surgery in good general condition. She develops normally at 5-year follow-up and is under the care of the oncologist, surgeon and pulmonologist. Last chest examination did not reveal any pathology.
DISCUSSION

Reported case concerned ELS located in the left part of the chest. It is usually recognized prenatally in contrast to ILS, which often is a later finding [6]. The estimated size of ELS lesions varies from 5 to 15 mm and according to other authors, may be larger after infections [7]. In our case, left – sided ELS was larger than usual at the very beginning of diagnosis and was growing with the advancing gestational age.

Huber was the first one, who reported lung sequestration-like lesion in 1777, then Rokitansky described similar anomaly in 1861, but it was Price in 1946 who first used the term „sequestration” [1, 12, 13]. Since that time the final explanation of the origin of BPS has not been found and it is still the subject of debate. The origin of BPS seems to be different depending on the type. ELS is considered to be always a congenital lesion, while ILS can be congenital or acquired [13]. Congenital lesions form from lung bud from foregut and are known as malformations. The acquired one can be an effect of sequential events, that start with bronchial obstruction and are usually accompanied by inflammation [3, 13, 14]. It is known that inflammation can occur even in ELS, which is an independent lesion, because of the existence of pores of Kochs and canals of Lambert, as a bronchoalveolar connections [15].

ELS occurs less often (25%), comparing with ILS (75%). Very infrequently ILS and ELS occur simultaneously [8]. ILS has no prevalence by gender, but ELS is definitely more frequently observed in males (up to 80% of cases) [7]. ELS is located in the left part of the chest up to 80% of cases, especially between the diaphragm and lower lung lobe, but about 10% of ELS can be located subdiaphragmatically [7], which is important for differential diagnosis. ILS is observed on both sides of the thorax, with similar incidence [1].

In the literature, many cases of coincidence of ELS and congenital cystic adenomatous malformation type II (CCAM type II) have been described, suggesting a common embryological origin [1]. CCAM should be taken into consideration in the differential diagnosis, along with congenital diaphragmatic hernia. The ultrasonographic image can also suggest the anomaly of the spleen, kidney, adrenal gland or pancreas [9]. Hyperechogenicity of the lung, as seen on US scans, can be caused by mucus retention in the obstructed bronchial tree [4].

BPS sometimes coexist with other anomalies (more often it is ELS than ILS), such as a diaphragmatic hernia, skeletal deformations, heart, renal and cerebral anomalies but it is considered as an isolated lesion, with no proof for being hereditary. If we do not observe other abnormalities in the fetus, there is no need for prenatal chromosomal investigation [1, 9].

BPS sometimes regress spontaneously, more often when it does not cause any complications during pregnancy (especially hydrothorax or hydrops fetalis) [9]. It is one of the most important issues when planning the prenatal intervention. It is also the point in parental counseling [4]. If BPS is not recognized during pregnancy, nor during the neonatal period, it usually causes recurrent pneumonias, hemoptysis, cough, chest pains, shortness of breath or dyspnea [11]. Surgical treatment needs to be considered even in asymptomatic patients because the lesion can be infected and cause permanent inflammation.

Prenatally, it can be difficult to diagnose BPS, especially ELS, when the image can be identical with normal lung, sometimes only with slightly higher hyperdensity [3]. Here, color Doppler may help visualize pathognomonic syndrome, which is the systemic blood supply. Color Doppler has to be performed very carefully because the vessel can be small and located close to the heart or aorta. All lesions must be confirmed postnatally by contrast-enhanced CT, which is the test of choice [7].

Most BPS cases are diagnosed because they may affect fetal condition and pregnancy outcome. Pleural effusion can cause fetal hydrothorax which may compress the diaphragm or cause a mediastinal shift. Differential diagnosis of primary or secondary hydrothorax has to be performed in such cases and it is worth considering that 25% of hydrothorax cases are associated with congenital lung malformations [16]. Pleural effusion is also a risk factor for polyhydramnios than can be caused by difficulties in swallowing the amniotic fluid, due to mediastinal shift. The mediastinal shift can next cause hydrops fetalis. It is connected with constricted venous return to the heart and is a cause of fetal or newborn death [10].

After the delivery, the most pressing problem that can exist in the patient with congenital lung malformations is lung hypoplasia. It is caused by lack of space for normal lung tissue to develop in the chest. Lung hypoplasia can lead not only to permanent hypoxia but also to pulmonary hypertension and it is very difficult to manage after delivery [17]. That is why the most important goal for prenatal intervention is to allow the lungs to develop normally. Space in the chest can be limited by pleural effusion fluid; in such cases thoracocentesis is recommended. It was proposed in 1982 and since that time is the most common procedure in fetal hydrothorax management [16, 18]. It is usually performed several times before the delivery because we do not solve the cause for increasing amount of fluid, only evacuate the overflow. Right after the procedure relaxation of the lungs must be checked, due to prior difficulties in imaging. The pleuro-amniotic shunt can be considered as another option. It is beneficial that it can be applied during one intervention, but in the literature we can find reports of its closure or dislocation [5].

Due to increased interest in fetal medicine new techniques come into consideration. Most of them use occlusion of the nutrient vessel of BPS. It can be done by injection of sclerosing substance, alcohol, laser or radiofrequency ablation [16]. The best choice remains unknown, awaiting further investigations. Open fetal surgery is not often considered, because of possible regression of the lesion and in severe cases, such as in hydropic fetuses, it is a very challenging procedure [10]. According to our and others’ experience adequate fetal monitoring and therapeutic management are crucial in pulmonary sequestration [16]. Monitoring the pregnancy with the diagnosis of BPS should include USG once a week and fetal echocardiography with the
same frequency to exclude circulatory insufficiency and heart failure. The goal is to avoid hydrotic changes and complications due to the location of the sequester and to prevent fetal death or preterm labor. Delivery should always take place in third reference level hospital and NICU should be prepared for admission. If fetal hydrops develops after 32 weeks of gestation caesarean section after fetal maturation with glucocorticosteroids should be performed. In mild cases, there are no indications for preterm delivery, but fetal medicine solutions should be considered, as well as EXIT procedure (ex utero intrapartum therapy) [5].

After the delivery, the newborn with BPS diagnosed prenatally should have extended diagnostic imaging performed. X-ray and contrast-enhanced CT are the investigations of choice [9].

Final management of the lesion remains surgical. After the first month of life neonatal pulmonary surgery is known as safe, especially in ELS [3]. Also, the anesthetic risk decreases after first 4 weeks of life [19]. During the procedure, the most challenging part is finding the nutrient vessel and avoiding bleeding but this should not be a reason for postponing it.

Histopathological findings may vary. The most common are cystic forms, that produce a lot of pleural effusion. Another finding can be a pseudotumorous form, that usually is asymptomatic but occurs very rarely [1]. Other types of histopathological changes can be found as the lung tissues malformations can coexist. In our case histopathology revealed bronchopulmonary dysplasia, that can be a part of the cystic form.

CONCLUSIONS

Fetus and newborn are in fact the same patient. In the emerging field of feto-maternal medicine, obstetricians and neonatologists, as well as pediatric surgeons, should cooperate closely to treat the patient from the very beginning in a coordinated way, following a conclusive differential diagnosis, that will not be disrupted by new ideas, that are in conflict with the previous findings.

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REFERENCES


LIST OF FIGURES

Fig. 1. Hyperechogenic lesion in the left fetal lung. Transverse scan of the chest.

Fig. 2. Bronchopulmonary sequestration in the left supradiaphragmatic area. Sagittal section of the chest.

Fig. 3. Visualization of separate circulation – abnormal artery of the lesion.

Fig. 4. Visualization of circulation originating from the aorta.

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FIG. 1. HYPERECHOGENIC LESION IN THE LEFT FETAL LUNG. TRANSVERSE SCAN OF THE CHEST.

FIG. 2. BRONCHOPULMONARY SEQUESTRATION IN THE LEFT SUPRADIAPHRAGMATIC AREA. SAGITTAL SECTION OF THE CHEST.

FIG. 3. VISUALIZATION OF SEPARATE CIRCULATION – ABNORMAL ARTERY OF THE LESION.

FIG. 4. VISUALIZATION OF CIRCULATION ORIGINATING FROM THE AORTA.