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Ex Utero intrapartum treatment of extralobar pulmonary sequestration: case report and literature review

Sofija Jackevičiūtė[1], Jelena Volochovič[1,2]

[1] *Vilnius University, Faculty of Medicine, M. K. Čiurlionio street 21, Vilnius, Lithuania*

[2] *Vilnius University hospital, Santaros clinic, Obstetrics, and Gynecology Center, Santariškių street 2, Vilnius, Lithuania*

Abstract

Background: Extralobar pulmonary sequestration is an uncommon congenital lung lesion, very rarely causing severe symptoms prenatally or requiring prompt surgical actions taken. The purpose of this report is to present a unique symptomatic case of extralobar pulmonary sequestration which led to an Ex Utero intrapartum treatment being performed for the first time in Lithuania and followed with successful further treatment of pulmonary pathology.

Case presentation: A nulliparous caucasian patient at 22 years of age was sent to a tertiary level clinic after alarming results were found during routine ultrasound examination at 29+4 weeks of gestation. After a detailed examination, extralobar pulmonary sequestration was diagnosed. Due to severely worsening fetal condition on week 32 of gestation the pregnancy was no longer continued and an Ex Utero intrapartum treatment procedure was performed. The pulmonary pathology later was surgically resected at 6 months of age.

Conclusion: This rare case of extralobar pulmonary sequestration allows us to explore key moments in the diagnostic and treatment pathways of this pathology. Magnetic resonance imaging, which allows evaluating parous patients more precisely than ultrasound imaging, with fetal pulmonary vessels being seen more clearly, is inspected as a necessary step in fetal lung sequestration diagnosis establishment. The Ex Utero intrapartum treatment procedure and how anticipated respiratory challenges of the newborn can be tackled with its implementation leading to timely execution of crucial interventions is researched.

Keywords: intrapartum, treatment, procedures.

1. Introduction

Pulmonary sequestration is a rare congenital pathology with an estimated incidence of 0,15% to 6,40% of all congenital abnormalities of the lungs (1). Pulmonary sequestration can be defined as a formation of dysplastic vascularized lung tissue that lacks communication with the tracheobronchial tree thus failing to properly function and impairing the respiratory function of healthy lung tissue. Extralobar sequestrations are predominant in infants and manifest with severe symptoms early on (2, 3, 4).

Clinical significance depends immensely on the size and the location of the sequestration. Prenatal complications can be as follows: fetal growth retardation, preeclampsia, preterm labor, and even miscarriage (1, 3). The most common postnatal complications a newborn may display in the case of severe pulmonary sequestration include respiratory distress, heart failure, pulmonary hemorrhage, fetal hydrops, and difficulty feeding (1, 3). Cases of minor lung sequestrations usually remain asymptomatic throughout life and are often diagnosed accidentally in adulthood.

Pulmonary lesions can be visualized on fetal ultrasound during the second trimester of pregnancy as of the 16th gestational week. The prenatal diagnosis of lung sequestration can be based on Doppler ultrasound imaging as well as magnetic resonance imaging (MRI) scans offering the highest accuracy when diagnosing this pathology *in utero*. Further treatment strategy decisions are made upon evaluating the severity of the sequestration and its potential threat to the newborn's survival. The measures taken for timely treatment can be as follows: conservative surveillance and newborn's lung surgery after labor, intrauterine feeding vessel laser coagulation, an Ex Utero Intrapartum Treatment, or the pregnancy can be terminated according to the countries law. This is a report portraying a case of a successful lung sequestration Ex Utero Intrapartum Treatment.

2. Case Report

A 22-year-old caucasian nullipara at 29+4 weeks of gestation was presented to tertiary level Vilnius university hospital Santaros Klinikos with suspected fetal lung sequestration. Although the previous fetal ultrasound exam at 18+5 weeks was within normal values, a thorough fetal ultrasound at 29+4 weeks described a case of fetal lung pathology: hydrothorax of the left lung, unidentified echo positive left side thoracic structure with set blood circulation (measured size being 2,7x2,4x1,7 cm), seen in Figures 1-3, hence the heart disposition to the right and the compression of the left lung itself along with an excess of amniotic fluid, polyhydramnios. At week 29+4 ultrasound imaging was insufficient for vascular supply origin identification. Because this step is essential in pulmonary sequestration diagnosis differentiation and establishment, we implemented a more accurate means of imaging, a fetal MRI for further case specification. 1,5 T MRI scan was performed using sequences T1, T2, and DWI and as a result, a much clearer conclusion of the pathology was drawn. Left hydrothorax, left supradiaphragmatic formation (2,6x1,9x1,6 cm in size) with vascular support from the aorta branching towards the diaphragm, dextrocardia, and polyhydramnios were detected and led to suspicion of probable lung sequestration.

During week 31+5 of gestation, an ultrasound exam showed the worsening

condition of fetal bilateral hydrothorax and polyhydramnios. Following this alarming change, the standard lung maturation process was started.

Moving forward, at week 32+2 of gestation, genetic consultation clarified that no genetic syndromes or associated abnormalities were present. The aforementioned case was specified as an isolated fetal development anomaly.

A fetal ultrasound on week 32+5 of gestation showed negative progress of fetal lung condition (bilateral hydrothorax, compression of both lungs, and dextrocardia due to the mass effect given by abnormal pulmonary derivative), hydrocele, and the potential threat of fetal hydrops, which led to a decision to no longer continue this pregnancy. The multidisciplinary committee agreed upon an Ex Utero Intrapartum Treatment strategy. During this procedure, a Cesarean section will be performed and before pinching the umbilical cord, while it still supplies the newborn with oxygenated blood, the pediatric surgeon will perform lung decompression, aiding the newborn's respiratory ability.

Beginning with fetal anesthesia and general anesthesia for the nullipara, the C-section was performed ordinarily. After pulling the newborn out of the womb, while the umbilical cord was still attached and hadn't yet been clamped, the neonate alarmingly did not exhibit any activity and was seen to be hypotonic and without elicited reflexes. Therefore, immediately the intubation, resuscitation of the newborn, and drainage of the left pleural cavity took place at the same time. 190 ml of fluid was drained in total. After 3 minutes and 30 seconds, the umbilical cord was clamped. The newborn was evaluated at a 4/5 Apgar score, the umbilical cord pH was at 7,25, within the normal value, which indicated that timely action prevented aided in normal systemic organ function and perfusion. Overall newborn's condition was noted severe thus with CPAP oxygen therapy applied, the baby boy was transported to a neonatal intensive care unit where he was closely monitored onwards.

The placenta was removed, previously attached to the posterior wall, and sent for a histology exam, which later did not reveal any placental pathology. Formerly collected fluid using pleural cavity draining was sent for thorough laboratory testing (microbiology, cytology, biochemistry).

The recovery process of the mother was smooth, as there were no complications after the C-section and the surgical wound was healing properly. Shortly her medical care was continued in outpatient settings.

Contrarily, promptly after being born, the neonate arrived at the neonatal intensive care unit (ICU), where his general condition was severe because of bilateral hydrothorax, respiratory failure, respiratory distress syndrome, heart failure, impaired microcirculation, prematurity, hypoxia at birth, low body mass, non-immune hydrops. Throughout neonates' stay in ICU, the main struggle remained its respiratory capability. A comprehensive examination of the neonate confirmed lung sequestration diagnosis by ultrasound (big 22 mm size sequestration with clearly identified arterial supply), pediatric cardiologist evaluated positive pulmonary hypertension progress. After almost two weeks of care in the neonatal ICU, our patient was stable enough to be transferred to a neonate unit for further care and further lung lesion treatment strategy planning. The patient was discharged

after 22 days of hospitalization, at 36 weeks. The newborn has been carefully observed by a pediatrician, a pediatric surgeon, and a development specialist since. Elective thoracoscopic sequestrectomy was performed at 6 months of age under general anesthesia. The surgery was successful and the postoperative period was smooth. After histological evaluation of the resected tissue, a conclusive diagnosis of extralobar pulmonary sequester was made. As of 15 months of age, the boy is healthy and is further attentively observed by a pediatrician, focusing on his overall respiratory function, infection prevention, comprehensive health, and development.

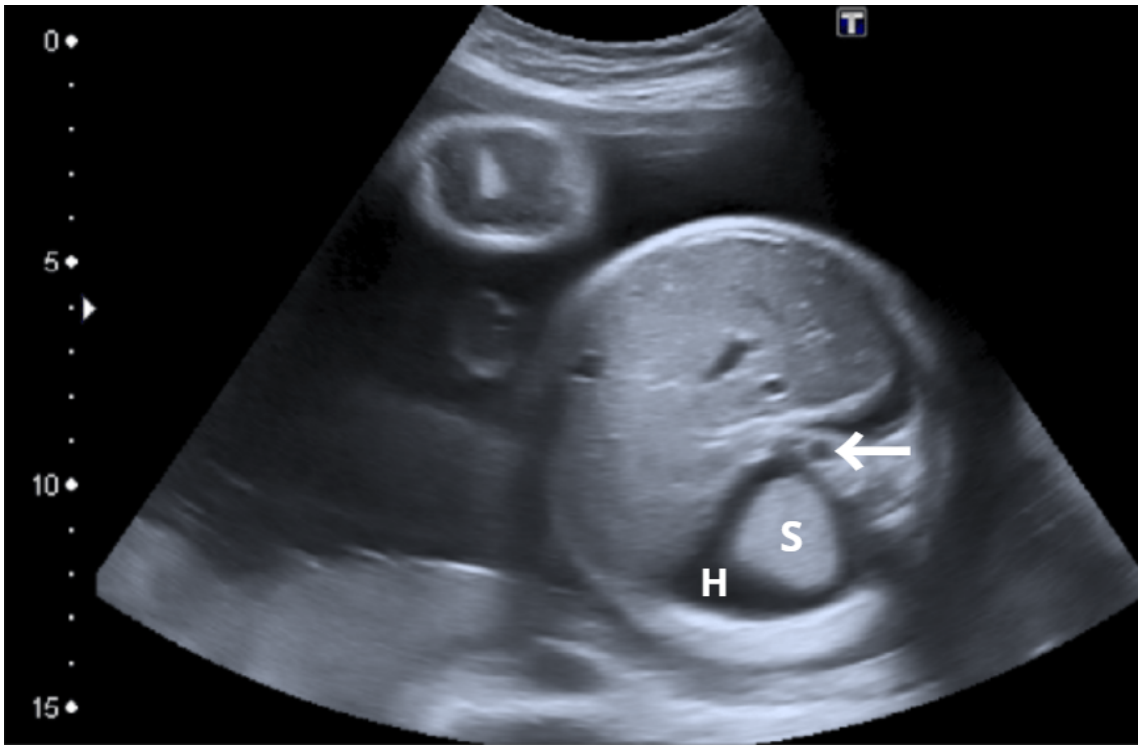

Figure 1. A fetal ultrasound, week 31+6. Abnormal derivative in the thoracic cavity (S) and hydrothorax (H), the white arrow pointing to the aorta.

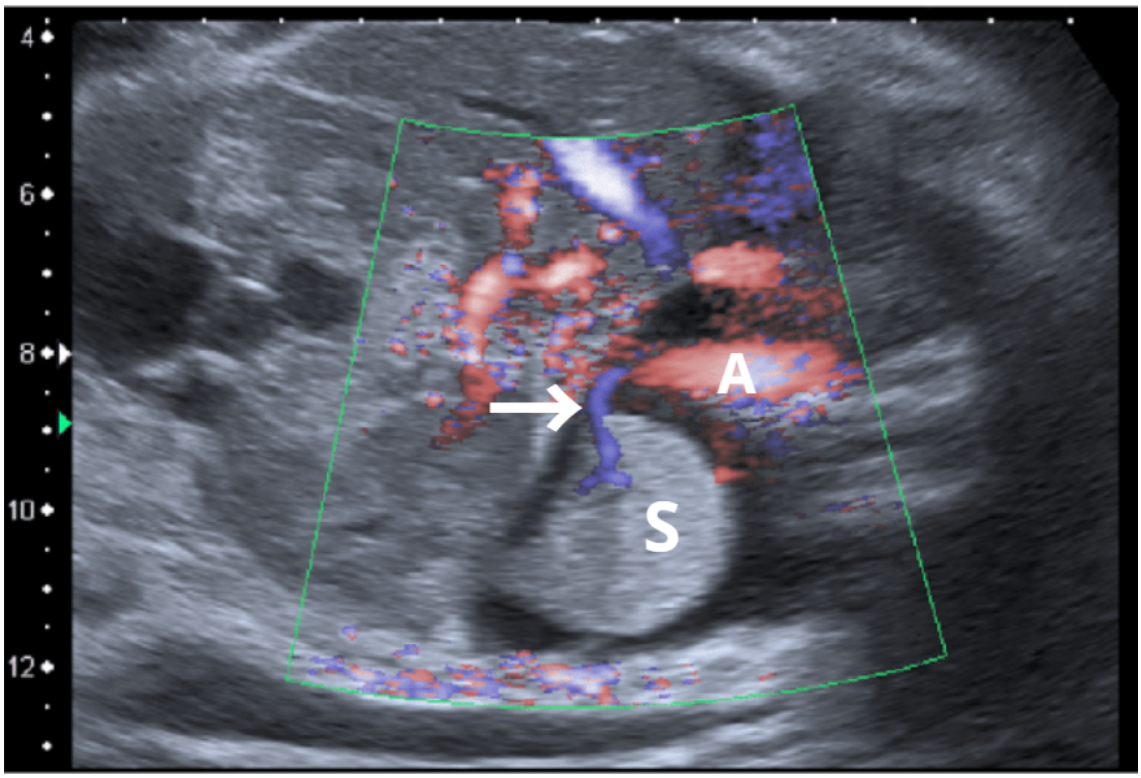

Figure 2. Fetal ultrasound with Doppler, week 31+6. Lung sequestration (S), the white arrow pointing to the feeding vessel branching from the aorta (A).

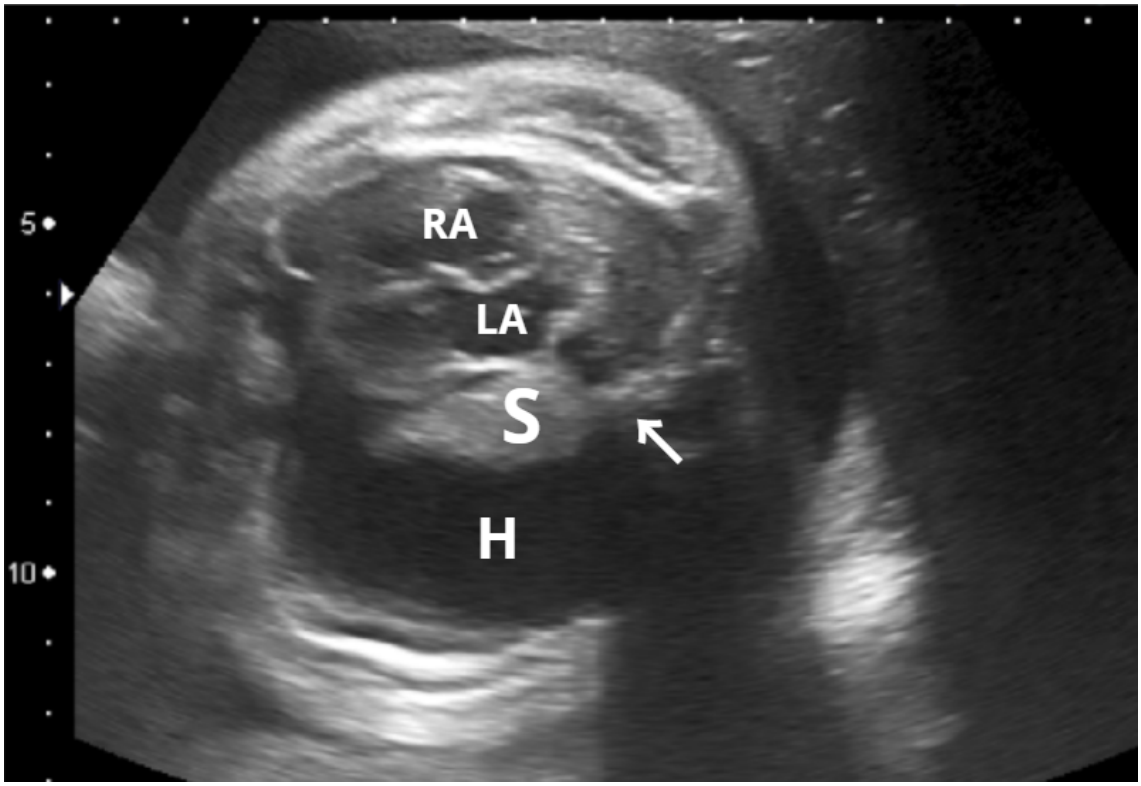

Figure 3. A fetal ultrasound, week 31+6. Lung sequestration (S) with pedicle (white arrow), causing dextrocardia (LA- left atrium, RA- right atrium) and hydrothorax (H).

3. Literature review and discussion

3.1. Prenatal diagnostic approach

To begin with, the first step in fetal pulmonary lesion diagnostics certainly is ultrasound imaging. On routine fetal ultrasound examination, indirect signs of lung sequestration are polyhydramnios, pleural effusion, mediastinal shift, fetal hydrops while the direct signs are the abnormal pulmonary derivative and the pathognomic arterial supply (14). Lung sequestration can be seen on fetal ultrasound as early as the 18th week of gestation, but the definite diagnosis is based on defining the abnormal feeding vessel clearly (1, 11). In our case, on week 29+4 signs of polyhydramnios, left hydrothorax, cardiac dextraposition, and an abnormal pulmonary derivative were seen on ultrasound examination, by week 31+6 we also were able to visualize the blood vessels supporting the derivative (Figure 2). Typical ultrasound imaging paired with Doppler is very helpful in establishing the suspicion of pulmonary sequestration when solid, triangle-shaped echogenic lung mass is detected (11). Even more accurate views can be obtained with 3D ultrasound especially because the origin and the direction of the feeding artery can be distinguished.

The identification of the feeding artery, branching from the aorta is crucial in the differential diagnosis of fetal pulmonary lesions, as the most common congenital lung lesion, congenital pulmonary airway malformation (CPAM), also called congenital cystic adenomatoid malformation, arises from the normal pulmonary vascular tree contrary to lung sequestrations (5, 6, 11). Estimated ultrasound accuracy is 72% (sensitivity 49%, specificity 93%) in pulmonary lesion vascular evaluation, whereas MRI accuracy is 80% (sensitivity 71%, specificity 88%), in some studies reaching up to 98% in comprehensive lung anomaly diagnostics (11, 13). Additionally, compared to MRI, prenatal ultrasound imaging is a much more accessible diagnostic measure, able to fulfill bronchopulmonary sequestration diagnostic criteria. However, due to numerous factors jeopardizing the ultrasound diagnostic capability, such as fetal position, amniotic fluid deficit, etc., vascular supply sometimes can not be visualized reliably (11, 12). Although MRI may not be as widely accessible as ultrasound imaging, MRI is superior as a better view of general fetal anatomy can be seen since more than 60% of extralobar lung sequestrations are associated with other abnormalities, also important is the independence of fetal and placental positions (6, 7, 11). In our case, by the time fetal pulmonary sequestration was suspected, ultrasound imaging did not provide needed vascular identification, thus for case specification an MRI was performed and feeding vessels of the derivative were examined and concluded to be branching from the aorta, hence extralobar pulmonary sequestration was diagnosed.

When neither ultrasound nor MRI is sufficient for feeding vessel recognition, computed tomography, CT, can be implemented postnatally with remarkable 90% accuracy (sensitivity 92%, specificity 88%) surpassing both US and MRI (11).

3.2. Treatment of choice

Treatment options and strategies follow a basic principle: the more severe case of pulmonary sequestration is, the more life-threatening the symptoms become, requiring

increasingly more complicated clinical tactics of choice.

Initially, this uncommon pathology very rarely does call for prenatal or perinatal treatment as a substantial part of fetal lung sequestrations are asymptomatic or are seen to resolve (8). Nevertheless, if respiratory problems are not to be expected, early delivery or a C-section are not indicated (11). Currently, consensus for optimal monitoring of asymptomatic patients does not exist, however regular monitoring by thoracic imaging (X-ray, CT or MRI) is recommended for small and asymptomatic lung lesions (18).

When neonatal respiratory complications are probable, delivery should be planned carefully together with neonatology experts and pediatric surgeons in a tertiary level center where required help and experience are available in need of urgent resuscitation or intervention as was done in our case (11).

Prenatal treatment options include thoracoamniotic shunting and intrafetal laser coagulation. Prenatal hydrops management is essential for healthy lung tissue development, that is why thoracoamniotic shunt placement or laser coagulation helps reduce the adverse consequences pulmonary sequestrations have on residual healthy lung maturation.

Treatment using a thoracoamniotic shunt hasn't shown promising results in cases of severe fetal lung sequestrations used for lung decompression, however, it is encouraged when signs of hydrops are seen before week 30 of gestation (1, 4). Since our patient had severe lung sequestration and only since week 32 hydrops was suspected, a thoracoamniotic shunt was not selected as a treatment strategy. According to one relatively large cohort study, only 59% of fetuses survived thoracoamniotic shunting and the main prognostic markers associated with poor survival were polyhydramnios, hydrops, mediastinal shift, and shunt-birth interval lower than 4 weeks, all of which were seen in our case study (21).

Intrafetal laser coagulation aids fetal lung condition by reducing sequester size, increasing total normal lung tissue volume, and reversing hydrops and pleural effusion in more than 75% of reported cases (18). In comparison with thoracoamniotic shunt, laser ablation has shown better results in bronchopulmonary sequestration treatment as total lesion regression was seen more often (18). Although sometimes a second ablation procedure is needed, higher gestational age is seen with laser ablation rather than shunt placement (19). Reasons, why laser coagulation was not selected in our case, are as follows: it was too late time-wise in the pregnancy to expect an effect of sequestration reduction after the procedure; fetal complications were already severe; the vascular location was particularly risky for coagulation, it being so close to the aorta.

An option for perinatal treatment exists as well. Perinatally, Ex Utero intrapartum therapy (EXIT) procedure can be applied in cases of large pulmonary sequestrations, hybrid lesions, and CPAM (17). When the fetal respiratory function is compromised, EXIT provides very valuable time for life-saving interventions (pleural drainage, ECMO cannulation, chest decompression, mass excision, etc.) during C-section and helps to delay surgery (17). This procedure requires a high level of experience, however, the general survival rate of fetuses is 90% (17, 20). In our case, the choice to perform EXIT was made upon

evaluating the rapidly worsening fetal condition, prematurity, and the high probability of respiratory decompensation after birth.

Furthermore, prenatal and perinatal interventions are focused on symptom management, targeting the complications of lung sequestration. Though sometimes sequestrations are seen to resolve completely after shunting or vascular coagulation, the sequestrations themselves may still cause symptoms postnatally and remain in need of resection later on (21). Postnatal extralobar pulmonary sequestration resection shows excellent results as well as low complication rates (11, 18). The notion is that when applicable and indicated, resection is an excellent strategy with lobectomy being the superior choice to segmentectomy (4, 8, 14, 17). Surgery should always be done on symptomatic patients 6 to 12 months old when no coinciding pathology interferes with the surgery (18). Resection is recommended for the prevention of infection, hemorrhage, malignancy, and curative purposes even for asymptomatic or low-risk patients (11, 14). Although the complication rate isn't yet clearly established, infection is the most common unresected pulmonary sequestration complication, usually requiring urgent surgery (18). For preventative and curative reasons elective postnatal sequestrectomy was performed in our case when the patient was 6 months old.

4. Conclusions

In this case study, prenatal ultrasound presented a suspicion of pulmonary dysplasia with the possibility of it being lung sequestration and an MRI helped to identify more accurate size, evaluate the anatomy of blood vessels, and conclude it as an extralobar pulmonary sequestration. In this case, the decision to perform an EXIT procedure and immediately drain the pleural cavity was made upon evaluating the worsening clinical condition of the fetus. With this procedure performed we were able to manage the anticipated life-threatening respiratory failure and postpone surgical resection to the recommended age for resection and later perform the sequestrectomy successfully.

This case report and literature review highlight the importance of prenatal diagnostic precision and clinical action planning. With this novel procedure implementation, we were able to expand our clinic‘s experience and, most importantly, save a child‘s life.

Informed Consent

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