Rare localization of an extralobar pulmonary sequestration in a child as a diagnostic challenge: a case report and review of the literature

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We present a child with a rare finding of an extralobar pulmonary sequestration localized in the upper mediastinum. Findings of the prenatal screening enabled early postnatal diagnostic measures that revealed a heterogeneous mass situated next to the thymus. Based on the localization and inconclusive computed tomography images, the preoperative prediagnoses included enterogenic cyst, thymus cyst and teratoma. Intraoperative features of the vascular supply and structure of the mass drew the surgeons' suspicion to extralobar pulmonary sequestration, which was confirmed in the histopathological examination. The scans were reevaluated after the surgery. Surgical management was implemented prior to the occurrence of any symptoms, which led to a positive general outcome.

The presented case should raise the awareness of radiologists, pediatric surgeons and other consultants involved in the diagnostic process of mediastinal lesions in children. The rare localization and lack of visualization of a systemic feeding artery can divert suspicion away from extralobar pulmonary sequestration in the preoperative differential diagnosis.

Key words: extralobar pulmonary sequestration, mediastinal malformation, prenatal diagnosis, postnatal management.

Pulmonary sequestration (PS) is a rare congenital lung anomaly presenting itself as a mass of non-functioning pulmonary tissue without communication with the tracheobronchial tree. Seventy-five percent of PS cases are intralobar pulmonary sequestration (ILPS) located within the pleural investment of the normal lung parenchyma. It mainly affects the lower lobe of the lung. Approximately 25% of PS are extralobar (ELPS), usually situated at the base of the left chest and diaphragm. They are surrounded by their own pleura[1,2]. Arterial supply of the ELPS in 80% of cases originates from a systemic artery, arising directly from the thoracic or abdominal aorta[3].

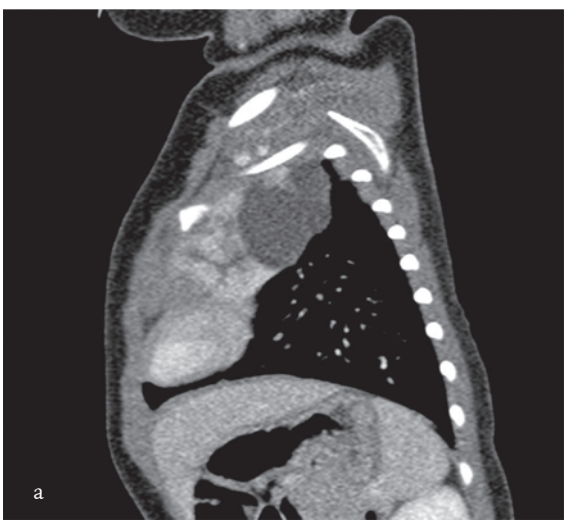
A pathological intrathoracic mass in a prenatal screening ultrasound examination is an indication for the postnatal sophisticated imaging diagnostic measures. The lesion may be assessed on computed tomography (CT), angiography or magnetic resonance imaging (MRI). Size, vascular supply, and communication with the tracheobronchial tree or the alimentary tract should be taken into account. As PS can be associated with other congenital anomalies, and a general evaluation is obligatory[1]. To avoid the risk of potential complications, surgical removal of the PS is the most common treatment option.

We report a child with a prenatally diagnosed intrathoracic mass that was determined to be ELPS, with a rare localization in the upper mediastinum, as an example of a diagnostic challenge in pediatric surgery.

Case Report

An intrathoracic mass was diagnosed in a routine sonography screening of a single fetus of a 36-year-old woman, gravida 3 para 3, at gestational age of 29 weeks. A heterogeneous cystic and solid lesion in the left lung area was monitored further. A cystic adenomatoid lung malformation was suspected. A female infant was born spontaneously at 39 gestational weeks, weighing 2660 g, with Apgar scores of 9 at 1 minute and 10 at 5 minutes. The newborn showed no respiratory distress. Postnatal chest CT at nine days of age revealed a cystic lesion in the superior mediastinum measuring 28x25x20 mm. Despite an inconclusive image, an enterogenic cyst was suspected. The newborn had no other congenital malformations. Postnatal echocardiography showed regular anatomic structure of the heart and great vessels; moreover, there were no pathological findings on the transfontanel cranial sonography.

The patient showed no pathological symptoms in further observation; nevertheless, a chest contrast-enhanced CT was performed at the age of four months. It showed a significant increase in the size of the mediastinal mass, which measured 40x27x25 mm at that time. It was composed of cystic and solid components. The upper cystic part of the tumor extended from the C7 level and adhered to the esophagus and left subclavian artery with fluid-filled cysts up to 2 cm in diameter. The solid part of the mass was located along the thymus gland edge. Systemic blood supply was not identified definitely (Fig. 1a, b). At that point, suspicion turned to a thymus gland cyst or teratoma.



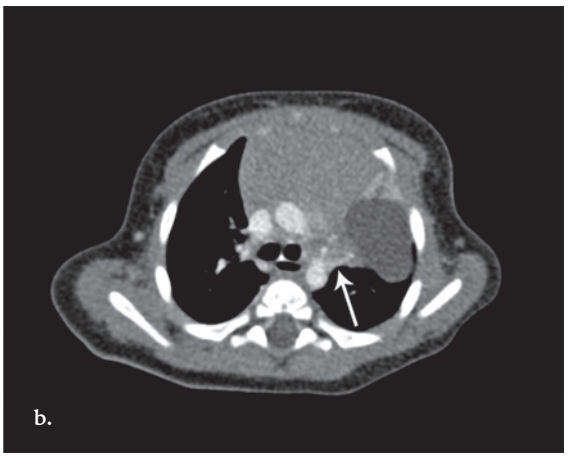


Fig. 1 a, b. Contrast-enhanced CT scans at the age of four months: a heterogeneous mass shaping the upper lobe of the left lung. *Arrow* demonstrates a band of connective tissue possibly containing the artery arising from the thoracic aorta.

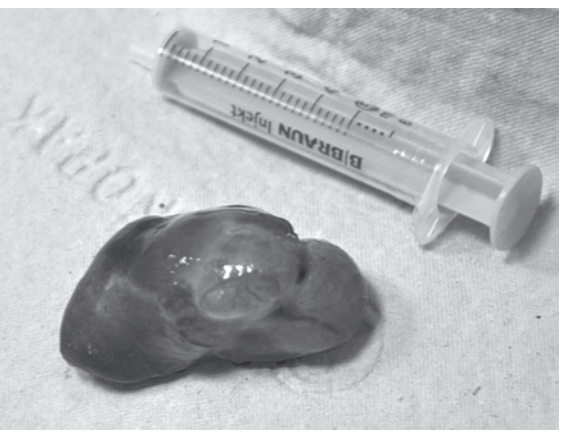


Fig. 2. The resected specimen with its own pleural covering.

At the age of six months, the infant underwent a surgical excision of the lesion in the Department of Pediatric and Oncological Surgery, Pomeranian Medical University, Szczecin, Poland, as a cooperation of both pediatric and general thoracic surgeons. A left posterolateral thoracotomy was performed through the fifth intercostal space. The heterogeneous soft mass with its own visceral pleura was seen located next to the upper lobe of the left lung and thymus gland. A small anomalous vessel arising from the descending aorta and

entering the mass was identified. The lesion was resected after ligation of that vessel. The resected mass was covered with its own pleura (Fig. 2). Based on the intraoperative features of the vascular supply and structure of the mass, the surgeons suspected ELPS. Histopathological examination of the resected specimen confirmed the diagnosis. The CT scans were carefully reevaluated after the surgery. In the final section of the aortic arch, a thin band of connective tissue was visualized, reaching a strongly vascularized tumor (Fig. 1b), possibly containing the vessel found during the surgery.

The postoperative course was uncomplicated, and the patient was discharged on the seventh postoperative day. She remains asymptomatic in good health and has not required treatment for five months postoperatively.

Discussion

The incidence of PS is 0.15%-1.8%, making it the second most common congenital lung anomaly[3]. It is linked with a high incidence of associated anomalies, such as diaphragmatic hernia, tracheoesophageal fistula, congenital heart disease, foregut duplication, and aneuploidy. The risk of such coexistence reaches 50% in ELPS cases. There is a high male predominance of PS, reaching 3:1 in patients with ELPS[1]. A prenatal diagnosis makes it possible to initiate early postnatal evaluation considering precise features of the mass, its vascular supply and potential coexisting anomalies. A prenatal sonography of the presented female infant revealed a cystic intrathoracic mass, which was suspected to be an enterogenic cystic lesion. Despite the high statistical risk, there was no evidence of other malformations. In more than half of ELPS cases, the lesions are located between the lower lobe of the lung and the diaphragm. Atypical localizations of ELPS include, for example, intradiaphragmatic, abdominal and intrapericardial cases[4,5]. Our patient presented an unusual ELPS localization in the upper mediastinum next to the thymus. Therefore, a thymus gland cyst or teratoma was suspected.

Radiological visualization of the aberrant artery of an unknown intrathoracic lesion directs the suspicion to PS. Prenatal ultrasound screening is the method of choice for detecting congenital lung anomalies. However, most of those anomalies cannot be completely characterized *in utero*, as there is high incidence of nonspecific imaging findings[6]. Zhang et al.[7] in a large retrospective study, described 68 PS cases diagnosed prenatally with clear identifiable systemic arterial blood supply visualized in Doppler ultrasound. Moreover, in the description of the prenatal diagnosis, it was emphasized that PS can be seen as a well-defined echogenic mass in the lower chest or the suprarenal region of the abdomen. In our case, as the intrathoracic mass was situated in the upper mediastinum, PS was not suspected at first. There was no data regarding systemic blood supply found in the prenatal screening. Therefore, the heterogeneous mass was monitored with classic contrast-enhanced CT after birth, and at the age of four months, with no evidence of aortic blood supply. This is in compliance with historical data by Ikezoe et al.[8], in which the rate of demonstration of the anomalous feeding artery using only CT reached 67%. If the arteries of a sequestration can be identified in conventional images and three-dimensional (3D) reconstructions, an additional angiography should be performed[9].

While arterial supply does not vary in the two types of PS, a proper evaluation of the venous drainage can be helpful in distinguishing ELPS from ILPS. In intralobar cases, the drainage is usually via the inferior pulmonary vein, while in extralobar lesions it is systemic. Lee et al.[10] underlined that this kind of differential diagnosis can be challenging, as it may be impossible to visualize venous drainage or to diagnose in the presence of a small vessel with unusual course. Different authors consider MRI angiography as an efficient and noninvasive method to precisely demonstrate the arterial supply of a sequestration to successfully differentiate PS from other lung and chest lesions[11,12]. Regardless of the certainty of the preoperative identification of the aberrant vessels, a successful outcome of the operation can be obtained with a careful surgical dissection[13].

The heterogeneous character of the intrathoracic mass should draw attention to PS. However, it can be misleading at the same time. Depending on the proportion of cystic components, type of material filling them, such as fluid, air or necrotic material, different assumptions can be drawn preoperatively[8]. Congenital mediastinal

malformations include PS, bronchogenic cysts, teratomas, lymphangiomas, hemangiomas, and neuroenteric cysts[14].

A surgical resection is controversial in asymptomatic patients. Therefore, possible complications of conservative management of PS should be kept in mind, which include risk of recurrent infection, hemorrhage, pneumothorax, malignant transformation, and consequences of pressure on other organs[15]. Bollouhey et al.[14], in a large study about congenital mediastinal malformations, stressed the absence of an increase in size of the lesion as a requirement to avoid surgical intervention. A conservative observation is a good option when the natural history of the PS involves a natural regression. This process is suggested to be a result of constriction of a small arteriole of the feeding artery[12]. In our case, a significant growth of the lesion was confirmed in the following CT with no signs of respiratory dysfunction or other complications prior to the surgery and in the postoperative course. Resection of an ELPS includes ligation of the systemic vessels, and can be performed via a thoracoscopic approach[16-18]. Curros et al.[19] and Lee et al.[20] reported successful effects of the embolization of the anomalous vessel, which led to regression of the lesions as confirmed in the follow-up CT. In our case, as a result of the uncertain preoperative diagnosis and radiological description, a classic thoracotomy was performed. This approach enabled a successful resection with efficient dissection of the feeding artery arising from the aorta.

In conclusion, a prenatal diagnosis of an intrathoracic mass enables its early postnatal evaluation. ELPS should be considered in the differential diagnosis of mediastinal malformations in children. It is crucial to perform surgery early enough to avoid occurrence of possible complications arising from a PS. It can be difficult to establish the precise features of the arterial supply in the preoperative diagnostic procedures. Furthermore, histopathological confirmation of ELPS should be a factor to evaluate preoperative radiological suggestions. Therefore, our case should raise the awareness of radiologists, pediatric surgeons and other consultants involved in the diagnostic process of mediastinal lesions in children.

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