Received: 22.04.2024
Accepted: 12.11.2025
Published: 13.01.2026

Tomasz Pytrus, Karolina Michalska

Pulmonary sequestration – a case report

Sekwestracja płucna – opis przypadku

2nd Department of Paediatrics, Gastroenterology and Nutrition, Wroclaw Medical University, Wrocław, Poland
Correspondence: Karolina Michalska, 2nd Department of Paediatrics, Gastroenterology and Nutrition, Wroclaw Medical University, Marie Curie-Skłodowskiej 50/52, 50-372 Wrocław, Poland, e-mail: k.b.michalska@gmail.com

https://doi.org/10.15557/PiMR.2025.0036

ORCID iDs
1. Tomasz Pytrus https://orcid.org/0000-0001-7609-8479
2. Karolina Michalska https://orcid.org/0009-0003-0808-6451

Abstract

The paper presents a case study of an infant with symptoms of gastroesophageal reflux, recurrent pneumonia, and tracheal narrowing observed during laryngoscopic examination, in whom intralobar sequestration was suspected during the diagnostic process. The article further provides an overview of current knowledge concerning the aetiology, clinical presentation, therapeutic approaches, and complications of pulmonary sequestration. Pulmonary sequestration is a rare congenital anomaly of the respiratory system in which non-functional lung tissue develops during prenatal development without communication with the bronchial tree. It is typically supplied by systemic arteries, most commonly from the descending aorta. Pulmonary sequestration is classified into two types: intralobar and extralobar. Common symptoms include respiratory distress, feeding difficulties, recurrent pneumonia, chronic cough, and haemoptysis. Lobectomy remains the treatment of choice; however, in asymptomatic patients, and in the absence of established guidelines, conservative management with periodic monitoring may be considered as a favourable alternative.

Keywords: pulmonary sequestration, intralobar sequestration, extralobar sequestration

Streszczenie

Praca zawiera opis przypadku niemowlęcia z objawami refluksu żołądkowo-przełykowego, kilkukrotnymi zapaleniami płuc i widocznym w badaniu laryngoskopowym zwężeniem tchawicy, u którego w toku diagnostyki wysunięto podejrzenie sekwestru wewnątrzpłatowego. Ponadto w artykule podsumowano aktualną wiedzę na temat etiologii, objawów klinicznych, leczenia i powikłań sekwestracji płucnej. Sekwestracja płucna to rzadka wada wrodzona układu oddechowego, w której w trakcie rozwoju prenatalnego tworzy się niefunkcjonalna tkanka płucna niemająca komunikacji z drzewem oskrzelowym i zaopatrywana przez tętnice systemowe, najczęściej aortę zstępującą. Wyróżnia się sekwestrację wewnątrz- i zewnątrzpłatową. Do typowych objawów sekwestracji można zaliczyć zaburzenia oddychania, problemy z karmieniem, nawracające zapalenia płuc, przewlekły kaszel, krwioplucie i inne. Chociaż leczeniem z wyboru jest lobektomia, to wobec braku jednoznacznych wytycznych w przypadku braku objawów można niekiedy przyjąć postawę wyczekującą. Wówczas dobrym rozwiązaniem może okazać się okresowa kontrola.

Słowa kluczowe: sekwestracja płucna, sekwestracja wewnątrzpłatowa, sekwestracja zewnątrzpłatowa

INTRODUCTION

This study aimed to present the case of a boy with recurrent pneumonia and symptoms including gastroesophageal reflux, in whom intralobar sequestration was suspected, and to provide a review of the literature on pulmonary sequestration.

CASE REPORT

A ten-month-old boy was admitted to the gastroenterology department due to symptoms of gastroesophageal reflux, such as regurgitation, hiccups, and cough. He was born at term by spontaneous vaginal delivery, in good general condition (Apgar 10/10) with a birth weight of 3,580 g. He was breastfed until two months of age; due to severe regurgitation, back arching, and skin changes, he was initially fed a casein hydrolysate formula and later, because of lack of improvement, an amino-acid-based formula, which brought relief. The boy was evaluated by a gastroenterologist two months before admission. He was treated with a proton pump inhibitor, and according to his mother, the regurgitation stopped after treatment. He had no other concerning gastrointestinal symptoms, and his weight gain was satisfactory. He passed stools once daily, formed or mushy, without pathological admixtures. The boy is currently undergoing dietary expansion; no adverse events have been observed.

He had three episodes of pneumonia in the past (on the seventh day of life and at one and six months of age), for which he was twice treated with antibiotics in an outpatient setting. In addition, the boy's medical history included an ENT consultation, during which laryngomalacia was suspected, leading to hospitalisation in the otolaryngology department. Laryngoscopy at that time revealed no structural abnormality of the larynx, although a mild external narrowing of the tracheal lumen was noted. A cardiology consultation was recommended, which showed no abnormalities and excluded congenital vascular rings. During hospitalisation in the gastroenterology department, laboratory tests were normal. Abdominal ultrasound (US) revealed an obtuse angle of His; otherwise without significant deviations from normal. US of the neck, lungs, and mediastinum did not show any obvious obstruction or focal lesion that could compress the trachea. Consequently, it was decided to extend the diagnostic evaluation with computed tomography (CT) of the chest, which demonstrated a small, triangular area of atelectasis in the medial portion of segment 10 of the left lung (in contact with the diaphragm), measuring 1.3 × 0.6 cm. Findings were suggestive of intrapulmonary sequestration (Fig. 1).

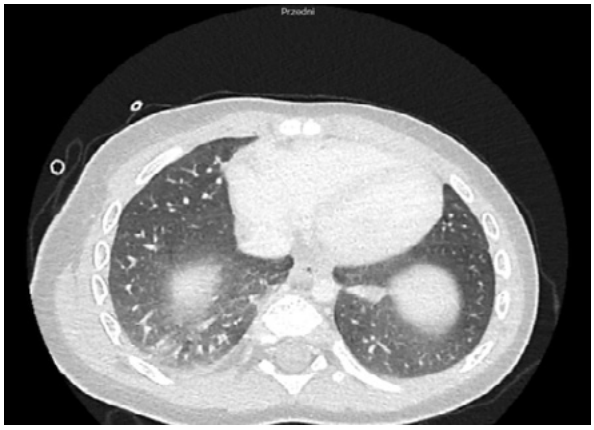

Fig. 1. Computed tomography of the chest

DISCUSSION

Pulmonary sequestration is a rare developmental anomaly of the respiratory system, characterised by non-functional lung tissue that has no connection with the bronchial tree and is supplied by at least one abnormal systemic artery. The anomaly most commonly involves the lower lobes of the lungs[1]. The aetiology of sequestration is not fully understood. Most likely, it arises from an additional abnormal tracheobronchial bud between the fourth and eighth weeks of foetal life[2–4]. Other hypotheses suggest pulmonary parenchyma degeneration, and some even regard sequestration as an acquired disease resulting from abnormal development, for example due to infection[3]. Pulmonary sequestration is the second most common congenital lung anomaly and accounts for less than 6% of all congenital pulmonary malformations. In 30% of cases, it is detected incidentally[2]. On imaging studies, sequestration appears as mass-like, cystic, bronchiectatic, or cavitary lesions[3]. Histologically, the pulmonary parenchyma shows signs of inflammation or fibrosis. Occasionally, the findings may also suggest emphysema[2]. Two types are distinguished: intralobar sequestration (ILS), which is covered by a shared visceral pleura, and extralobar sequestration (ELS), which is separated from normal lung tissue by its own pleura. ILS accounts for 76–85% of all sequestration cases and is most often located in the lower lobe of the left lung[2]. It is supplied by systemic arteries, most commonly the thoracic aorta and less frequently the abdominal aorta[1]. Venous drainage is usually into the pulmonary vein[5]. ILS occurs equally frequently in both sexes. ELS is most commonly located between the diaphragm and the lower lung lobes, more often affecting the left lung[2,5]. Atypical locations have also been described, such as mediastinal, pericardial, diaphragmatic, or abdominal. It may also occur bilaterally[6,7]. In ELS, arterial blood supply is mainly derived from the thoracic or abdominal aorta, and less commonly from the coeliac trunk, splenic artery, subclavian artery, or intercostal arteries[2]. Venous drainage occurs into systemic veins. ELS occurs more frequently in males[2]. In more than 60% of patients with ELS, other developmental anomalies coexist, most commonly congenital diaphragmatic hernia or congenital heart defects[1,3,4].

DOI: 10.15557/PiMR.2025.0036

Symptoms

ELS usually presents clinically in the neonatal period or early infancy – in 60% of cases within the first six months of life[1,2,4]. Typical symptoms include respiratory distress, feeding difficulties, and congestive heart failure. Prenatal examinations may also reveal foetal hydrops. ELS is only sporadically infected, as it is separated by pleural infiltration[2,5].
ILS rarely causes symptoms before the age of two[1], and much more often remains asymptomatic. The most common clinical manifestations of ILS include recurrent pneumonia, chronic cough, chest pain, haemoptysis, dyspnoea, and pneumothorax[6,7].

Diagnosis

Diagnostic evaluation should begin with a chest X-ray. However, small pathological lesions may not be detectable on X-ray. US is recommended as the first-line examination for patients with suspected supradiaphragmatic changes in childhood. For more detailed diagnostic assessment, CT angiography or magnetic resonance angiography (MR angiography) may be employed[1,2]. The multiplanar imaging capabilities of CT and magnetic resonance allow better visualisation of parenchymal lung lesions and the anatomical course of systemic arterial and venous supply. When non-invasive diagnostic methods are insufficient, arteriography should be considered[1]. Prenatal diagnosis can be obtained by performing a Doppler ultrasound examination from the 16th week of foetal life[1]. It should be emphasised that the detection rate of sequestration in prenatal examinations is increasing[7]. Pulmonary sequestration should be considered in every case of congenital pulmonary adenomatoid malformation, congenital diaphragmatic hernia, and in children presenting with symptoms such as recurrent respiratory tract infections, respiratory failure, haemoptysis, and congestive heart failure without an identifiable cardiac cause[1,4]. Asymptomatic ELS is usually detected incidentally during routine diagnostic evaluations in adult patients[8,9].

Treatment

The course of sequestration can vary – from regression of the pathological mass to the development of life-threatening symptoms. Because detailed data on its natural history are lacking, there are no clear guidelines supporting the absolute necessity of surgical treatment, particularly in asymptomatic cases or those diagnosed in foetal life[1,3]. Surgical intervention should be considered in symptomatic patients and in situations where the nature of the lesion remains uncertain. In asymptomatic individuals, periodic monitoring may be a suitable approach, allowing for postponement of surgery[3,9]. The treatment of choice for ILS is lobectomy performed via traditional thoracotomy, whereas for ELS, sequestrectomy is recommended. The VATS (video-assisted thoracoscopy) technique is currently gaining increasing popularity, as it is associated with a smaller postoperative wound, shorter drainage time, reduced duration of opioid use, shorter hospitalisation, and a better cosmetic outcome. Considering long-term complications, it also carries a lower risk of developmental deformities such as scoliosis or chest wall asymmetry[2,5,10]. Embolisation of the abnormal systemic vessels supplying the sequestration may also serve as an alternative to traditional thoracotomy[2,3]. Large lesions that may cause serious complications for the foetus require intrauterine intervention[11]. The prognosis is generally favourable in cases of pulmonary sequestration without associated pleural effusion or hydrops foetalis[12]. In the differential diagnosis, bronchial cysts, cystic adenomatoid malformations, focal bronchiectasis, congenital lobar emphysema, and retroperitoneal tumours in accessory abdominal lobe sequestrations should be considered[2].

Complications

Long-term complications of sequestration include gastroesophageal reflux, pneumonia, asthma, and growth disorders. In adults, the most frequently observed complication is haemoptysis due to high pressure originating from systemic vessels[2]. In the case of ILS, recurrent respiratory tract infections also occur, most commonly caused by *P. aeruginosa*. There are also reports of malignant tumour development within ILS[2].

CONCLUSIONS

The presented symptoms appear to be associated with the presence of intrapulmonary sequestration. Given the patient's good general condition and lack of symptoms, further observation was recommended, with follow-up imaging scheduled in 6–12 months. Expansion of the diagnostic work-up at a specialised pulmonology centre was also planned. In light of this clinical case, it should be emphasised that imaging diagnostics must be expanded in children with recurrent pneumonia, particularly when other causes – such as infectious or immunological disorders – have not been identified.

Conflict of interest

The authors do not declare any financial or personal links with other persons or organisations that might adversely affect the content of the publication or claim any right to the publication.

Author contribution

Original concept of study; collection, recording and/or compilation of data; critical review of manuscript; final approval of manuscript: TP. Analysis and interpretation of data; writing of manuscript: KM.

References

1. Corbett HJ, Humphrey GME: Pulmonary sequestration. Paediatr Respir Rev 2004; 5: 59–68.
2. Chakraborty RK, Modi P, Sharma S: Pulmonary sequestration. In: StatPearls [Internet]. StatPearls Publishing, Treasure Island, FL 2023 Jan–.
3. Mazurek H, Buchwald J, Pogorzelski A: Sekwestracja płucna. Pediatr Dypl 2013; 17: 56–60.
4. Pikwer A, Gyllstedt E, Lillo-Gil R et al.: Pulmonary sequestration – a review of 8 cases treated with lobectomy. Scand J Surg 2006; 95: 190–194.
5. Bonnard A, Malbezin S, Ferkdadji L: Pulmonary sequestration children. Is the thoracoscopic approach a good option? Surg Endosc 2004; 18: 1364–1367.
6. Gezer S, Taştepe I, Sırmalı M et al.: Pulmonary sequestration: a single-institutional series composed of 27 cases. J Thorac Cardiovasc Surg 2007; 133: 955–959.
7. Zhang N, Zeng Q, Chen C et al.: Distribution, diagnosis, and treatment of pulmonary sequestration: report of 208 cases. J Pediatr Surg 2019; 54: 1286–1292.
8. Yang L, Yang G: Extralobar pulmonary sequestration with a complication of torsion: a case report and literature review. Medicine (Baltimore) 2020; 99: e21104.
9. Wei Y, Li F: Pulmonary sequestration: a retrospective analysis of 2625 cases in China. Eur J Cardiothorac Surg 2011; 40: e39–e42.
10. Rothenberg SS: First decade's experience with thoracoscopic lobectomy in infants and children. J Pediatr Surg 2008; 43: 40–45.
11. Eber E: Antenatal diagnosis of congenital thoracic malformations: early surgery, late surgery, or no surgery? Semin Respir Crit Care Med 2007; 28: 355–366.
12. Litwińska M, Litwińska E, Szaflik K et al.: Management options for fetal bronchopulmonary sequestration. J Clin Med 2022; 11: 1724.

DOI: 10.15557/PiMR.2025.0036