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# A rare case of coexistent intralobar and extralobar pulmonary sequestration

**Case Report**

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**Abstract:** In the pathology of respiratory tract, sequestration presents as a non-functional lung tissue with no communication with tracheobronchial tree. It represents a rare congenital bronchopulmonary and vascular malformation, which occurs at a frequency of 0.1 to 6%. Intralobar and extralobar sequestrations are extremely rare congenital anomalies, especially if they occur at the same time in a patient. Proper diagnosis and appropriate surgical therapy, in the absence of associated anomalies, provide an excellent prognosis. In this paper, we are describing the simultaneous presence of intralobar sequestration (ILS) in the lower lobe of the left lung and extralobar sequestration (ELS) which is positioned on the aortic arch, in a 53 years old man. Two years post surgery, there is no recurrence or any patomorphological and functional disorders in the respiratory tract.

**Keywords:** *Bronchopulmonary sequestration • Congenital abnormalities • Lung • Surgery*

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## 1. Introduction

Term sequestration comes from a latin word sequestratio, which literally means separation or setting aside from the whole. In the pathology of respiratory tract, pulmanory sequestration represents a non-functional lung tissue without communication with the tracheobronchial tree. It represents a rare congenital bronchopulmonary and vascular malformation that occurs at a frequency of 0.1 to 6% [1,2]. Pulmonary sequestration was first described in detail by Pryce in the 1946 when intralobar (ILS) and extralobar form (ELS) of sequestration were precisely defined [3]. In the last 50 years some 900 separate publications have been presented with both forms of lung anomalies, while the simultaneous existence of the ILS and ELS has so far been presented only in 8 publications [4,5].

In this paper, we are describing the simultaneous presence of ILS in the lower lobe of the left lung and the ELS which is positioned on the aortic arch, in a 53 years old man. Two years after surgery, the patient is without recurrence and without any pathomorphological and functional disorders in the respiratory tract.

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## 2. Case report

A 53-year-old man was hospitalized at the Department of Thoracic Surgery, Clinical Center Kragujevac in Serbia due to strong cough with haemopthysis. On admission, the patient was conscious, oriented, fully mobile, with normal vital signs, with no signs of peripheral lymphadenopathy, which gave the impression of an easy patient. The patient was fully examined clinically and radiographicaly, and laboratory tests were performed. Internist findings were in the orderly manner and respiratory function within the reference values. Radiography (RTG) of the chest showed infiltrative changes in the left lower lobe of the lung. Bronchoscopy examination resulted in hyperemia of the mucus in trachea and principal bronchus, indicating extramural compression in the left and right main bronchus, whereas other lobular bronchi and bronchial subsegments were easily visualised. Pathohistological and cytological examination of material taken during byoptic fiberaspiration showed a squamous metaplasia and a mild, non-specific, inflammatory infiltration.

The chest multisliced computerized tomography (MSCT) was done with IV contrast and it showed left paraortic and supradiaphragmic non-homogeneous honeycomb structure, resizing 42 x 69 x 35 mm with micro-calcifications (dense content, up to a solid structure), attenuation of 20 to 43 hazard units (HU, Figure 1a,b). In comparison with the previous MSCT findings, the lesion was stationary. The abnormal parenchymal condensation zone was in the left apically paraaortic area. There was no pleural effusion. Several individual lymphnodes were seen in the right paratracheally area, approximately 9 mm in diameter. The board of the department of thoracic surgery opted for an operative treatment.

After adequate preoperative preparation, the patient underwent left posterolateral thoracotomy through V intercostal space under the general endotracheal anesthesia. There was an adhesion between the pleura surfaces in the lungs and above the top of the pericardium, and with partly dull, partly sharp preparation, they were separated with adequate hemostasis. After mobilization of the left lung with the exploration in the left lower lobe of the lung, a brown-red lesion was found, positioned centrally, which macroscopically resembled air-free lung tissue (Figure 2a). Another such lesion with the same characteristics, 5 cm in diameter, was found on the aortic arch (Figure 2b). Due to such intraoperative findings, it was decided for the left lower lobectomy to be performed, which began with a preparation of the lower pulmonary vein and it was finished with the removal of the lower lobe. The extirpation of the aortic arch lesion followed the triple binding and cutting of the two inlet arterial blood vessels, which originated from the aortic arch. The operation was completed by placing stitches, in reverse order, following placement of the two chest drainage tubes through the skin holes and connection to underwater drainage system. Both tissue preparations were sent off for pathohistological analysis. Macroscopically, both lesions were veiled in a smooth and glossy membrane, at the intersection they represented homogeneous material of brown-red color and of soft consistency. Lesion in the lung lobe was localized in its lower part, close to the visceral pleura and was at the largest diameter ~60 mm, while the removed mass from the aortic arch was 50 mm in diametar. Microscopically, observing the number of paraffin sections which were taken from both lesions, there were irregularly spaced elements of the lung parenchyma, bronchioles surrounded by rare and irregular, locally dilated alveoli, multiple blood vessels with thickened wall and with numerous lymph vessels and capillaries, and focal cartilage that was not continuous with bronchial wall were noticed. Stroma was partly changed, partly edematous and loose, saturated with blood, and inflammatory infiltrates, dominated by the lymphocytes with some rare neutrophilic leukocytes. (Figure 3a,b). Based on this histological picture, the diagnosis of ELS and ILS was made.

The postoperative course was uneventful, control RTG of the chest showed normal findings and on day 10 after the operation, patient was discharged from the hospital in a good general condition.

## 3. Discussion

Pulmonary sequestration is a rare congenital broncho-pulmonary-vascular malformation, which represents about 0.15 to 6.4% of all the anomalies of respiratory tract [1-7]. Pathomorphological, it is a non-functional lung tissue that has no communication with traheobronchial tree. It was first described in the mid last century by Pryce and colleagues, clearly differentiating sequestration in two different forms: intralobar and extralobar [3]. ILS is located within the functional lung parenchyma and does not have its own visceral pleura. ELS is the lung tissue that has its own visceral pleura, wthat is not in communication with traheobronchial tree, or with functional lung tissue, making it clearly separate from the accessory lung entity [8]. Generally, they are more frequently encountered to the left, rarely to be bilaterally localized. ELS may be located outside the thorax, in 80% of the cases encountered in men and is usually diagnosed in infancy [2,4-7,9].



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Blood supply of the sequestrum is separated from lungs vascular system. In 94% cases of the ILS is supplied with blood directly from the thoracic or abdominal aorta [4]. Rarely, inlet artery can be an aberrant vessel arising from the intercostal arteries, celiac, anonymous artery, subclavia, even from pericardioprenical arteries [10]. ELS is usually supplied with blood from the aorta, which has occured in this case, but also from other blood vessels, i.e. their aberrant branches, such as bronchial artery, circumflex coronary artery, lienal, renal or gastric arteries [4,10]. In fact, considering that the ELS may be located outside of the chest (usually in the upper parts of the abdomen), the artery that nourishes it usually starts from a large blood vessel that nourishes another organ or organ system. Venous drainage of ILS takes place mostly through azigos veins or lung vein systems in the left (95%), and very rarely in the right atrium [4,10,11]. This method of blood supply is a minor form of arterial-venous shunt.

Precisely, vascularization of the ILS and ELS can explain their origin. Namely, Pryce was describing the structure of the arteries nourishing the ELS, and came to two conclusions to which the attention was given later. These are firstly, that the arteries which nourish the sequestered lungs have a larger amount of elastic fibers in the media wall, and secondly, that there is no branch of pulmonary artery which is connected with the sequestered lung tissue itself [3]. On the basis of this, thesis is made on the embryonic origin of ELS, because this kind of artery structure matches the embryonic type of arteries, thus it is suspected that disruption of oxygen supply in the lung tissue during embryonic development can lead to the formation of aberrant arteries, which should compensate for this deficiency. The embryonic origin is also speculated due to frequent association of ELS (58%) with the congenital malformations of the anterior abdominal wall and gastrointestinal tract [6,11,12]. For example, even in 20-30% of the cases, ELS is associated with diaphragmatic hernia [13].

In contrast to ELS, ILS can be acquired. The obstruction of the bronchial tree at the level of terminal bronchioles may result in functional disorders of the pulmonary circulation, resulting in hypertrophy of small subsegmental arteries which nourish the traheobronchial tree. At the same time there is a discontinuity of a traheobronchial tree in this area, mostly because of recidivant infection, and thus all conditions for the occurrence of ILS are created [10,14,15].

The presence of ILS and ELS in one patient at the same time is extremely rare phenomenon. To date, only eight cases were described; in 7 of which ILS and ELS were placed unilaterally and in one of them bilaterally [4,5,9]. In our patient, ILS was localized in the lower lobe of the left lung, and ELS was on the aortic arch.

Clinical manifestations of ILS and ELS can be long absent. In most cases, symptoms are corresponding to a chronic lung infection and therefore bronchiectasis is often suspected, especially in the case of ILS [10]. In some cases, differential diagnosis includes lung tumors for ILS, and with the ELS, especially if located near the diaphragm, the existence of mediastinal tumors, intra-abdominal tumors, aneurysms of the aorta and others can be suspected [10,15,16]. These are the reasons why patient must be carefuly monitored, with detailed analysis and further examinations, with contrast-radiographic diagnostic methods (CT, MSCT, angiography), sovereign to complete the final diagnostics, as well as for the choice of treatment methods [4,7].

The treatment of choice for the ILS and ELS is surgical resection. The second choice for treatment is antibiotic and symptomatic therapy, with surgical treatment at a later stage. However, this kind of approach to the pathology is still rarely used because the frequent infections of the sequestered lung tissue can lead to fatal bleeding of abberant vessel [1,3]. The very essence of surgical treatment of pulmonary sequestration is based on finding and ligature of the intake, i.e. aberrant blood vessels. When it comes to the ELS, finding the aberrant artery and ligation is relatively easy, while in the case of ILS anatomical resection of one lung lobe containing the ILS must be done. In case of coexisting ILS and ELS, both of them are worked on in a single act of resection, usually through the postero-lateral thoracotomy, which we also applied as a surgical approach [14]. In recent years video-assisted thoracoscopy is increasingly used as modern technique, with relatively comfortable operating mode [17]

## 4. Conclusion

ILS and ELS represent extremely rare congenital anomalies, especially if they occur at the same time in a patient. Proper diagnosis and appropriate surgical therapies, in the absence of associated anomalies, can provide excellent prognosis.



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