

# Extralobar pulmonary sequestration: a case report

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**ABSTRACT:** *Extralobar pulmonary sequestration: a case report. A. Yilmaz, G. Bektemur, G.H. Ekinci, E.A. Ongel, M. Kavas, O. Haciomeroglu, M. Demir, B. Burunsuzoglu.*

We described the case of a 36-year-old Turkish female with an extralobar pulmonary sequestration who suffered from chest and back pain for five years without any evidence of pulmonary infection. A chest X-ray showed an area of opacity behind the cardiac silhouette in

the lower area of the left hemithorax. A CT scan of the thorax with intravenous contrast showed a 9 x 7 cm in size ovoid mass with necrosis in the lower left lobe. It revealed two aortic branches directed toward the pulmonary opacity. She subsequently underwent surgery and the anomalous tissue was removed by mass excision. The patient was diagnosed with extralobar pulmonary sequestration. *Monaldi Arch Chest Dis 2013; 79: 2, 90-92.*

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Pulmonary sequestration is defined as a portion of abnormal lung tissue that does not communicate with the tracheobronchial tree through a normally located bronchus and has an abnormal vascular supply [1]. It was first described by Rektorzik in 1861, but the term "sequestration" was introduced by Pryce in 1946 in a 7-case report [2, 3]. This extremely rare disorder accounts for 0.16% to 6.4% of all pulmonary congenital malformations [3]. Depending on its location, pulmonary sequestration is generally subdivided into intralobar and extralobar sequestration [4]. Extralobar pulmonary sequestration accounts for 14%-25% of cases [2]. It is most frequently diagnosed in the first 6 months of life, but is seen incidentally, though rarely, in older children and adults [5]. We present an adult case with extralobar pulmonary sequestration.

## Case Report

A 36-year-old Turkish female, a non-smoker, was admitted to our department with complaints of chest and back pain for five years. Her past history was unremarkable. A chest X-ray (CxR) showed an area of opacity behind the cardiac silhouette in the lower area of the left hemithorax (figure 1). On admission, her resting pulse rate was 86 beats per min., blood pressure 100/70 mmHg, and respira-

tory rate 18 breaths per min. Her respiratory system and general examinations revealed no significant abnormalities. Full blood count and biochemical tests were normal. Erythrocyte sedimentation rate was 20 mm/h. During hospital stay, the patient was given a course of antibiotics, but opacity in the left lower area persisted. A Computed Tomography (CT) scan of the thorax with intravenous contrast showed a 9 x 7 cm in size ovoid mass with necrosis in the lower left lobe. It revealed two aortic branches directed toward the pulmonary opacity (figure 2), which is consistent with a suspicion of pulmonary sequestration. Bronchoscopic examination revealed a normal endobronchial appearance. Bronchial lavage examination was negative for acid-fast bacilli. Her thoracic Magnetic Resonance (MR) angiography showed an arterial supply from descending thoracic aorta. Pulmonary functional tests were within normal limits. The patient was referred to surgery, and a left-sided thoracotomy was performed. The lesion was located within the pleural space between the diaphragm and the lower lobe. Four arteries arising from the descending aorta were identified. The anomalous pulmonary tissue was removed by mass excision. Pathologic examination of surgical material obtained diagnosis of extralobar pulmonary sequestration. The patient still remains in excellent condition after nearly two years from the operation.



Fig. 1. - Chest-x ray shows an area of opacity behind the cardiac silhouette in the lower zone of the left hemithorax.

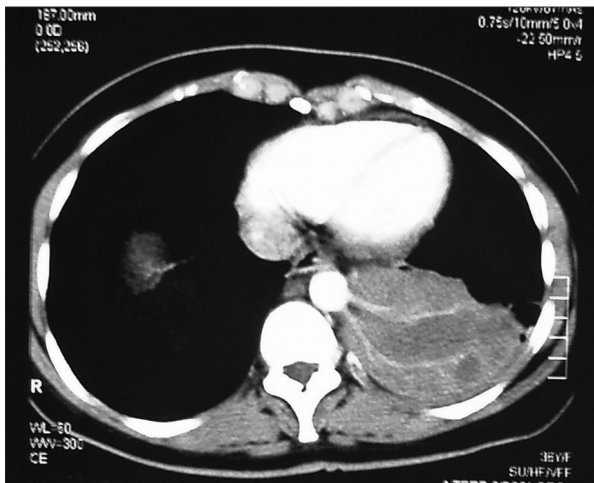


Fig. 2. - Computed Tomography scan of thorax with intravenous contrast showed a 9x7 cm in size ovoid mass with necrosis in the left lower lobe and two aortic branches directed toward the pulmonary opacity.

## Discussion

Pulmonary sequestration is an extremely rare congenital abnormality of the lower respiratory system, consisting of about 0.16-6.4% of all pulmonary malformations [3]. It exhibits three major features: an aberrant blood supply of a large caliber artery, a bronchial anomaly and dysgenesis of the parenchyma [6]. The etiology of pulmonary sequestration is unknown, but it is usually considered to be a foregut malformation [1, 5]. Anatomically pulmonary sequestration is generally classified into intralobar and extralobar sequestration. The intralobar type is located within the normal lung parenchyma and shares the viscera pleura of the parent lobe of the lung, whereas the extralobar type is located outside the normal lung and has its own visceral pleura. Intralobar sequestration has normal pulmonary venous return, while extralobar sequestration is associated with aberrant pulmonary venous drainage [5, 7].

Extralobar pulmonary sequestration accounts for 14-25% of the cases. Males are affected ap-

proximately four times more often than females. Sixty-one percent of patients present when they are younger than six months of age. It is rare to find it in adults [2, 3, 5]. Extralobar pulmonary sequestration is frequently located between the left lower lobe and diaphragm. Rare infradiaphragmatic, intraabdominal, intrapericardial, mediastinal or upper thoracic region locations have been reported [6, 8]. The patients are usually asymptomatic and are generally discovered by chance during routine CxR [8]. Over half of these patients have associated pulmonary anomalies such as bronchogenic cysts, lung hypoplasia, congenital cystic adenomatoid malformation, congenital diaphragmatic hernia [5]. We presented an adult female patient with extralobar pulmonary sequestration. The lesion was located within the pleural space between the diaphragm and the lower lobe in our patient. She suffered from chest and back pain localized to the side of the lesion. She had no significant evidence of infection and had no previous history of pulmonary infection. Our patient had no associated congenital anomalies.

Ideally, the diagnosis could be obtained without surgery by imaging. Imaging studies have two principal objectives: to rule out other pathologies and to confirm the presence of an anomalous arterial supply [2, 9]. A CxR is the first step in the diagnosis of pulmonary sequestration, like most other thoracic diseases [8]. Extralobar sequestration manifests as a single, well-defined, homogeneous, triangular opacity typically located in the posterior basal segment of the lower left lobe. CT of the chest characteristically demonstrates extralobar sequestration as a homogeneous well-circumscribed mass of the soft tissue attenuation. Aberrant vessels should be visualized for the definitive diagnosis of pulmonary sequestration. The diagnosis of pulmonary sequestration can be confirmed by multichannel CT scans of the chest with an intravenous contrast and reconstruction. In such cases, arteriography is unnecessary although it is gold standard for the diagnosis of pulmonary sequestration [5, 8, 9]. In our case, a CxR showed an area of opacity behind the cardiac silhouette in the lower area of the left hemithorax. A CT of the thorax with intravenous contrast revealed a mass lesion in the posterior basal segment of the lower left lobe and two aortic branches directed toward the mass lesion. Her thoracic MR angiography showed an arterial supply from descending thoracic aorta. The blood supply of extralobar sequestration comes directly from the thoracic or abdominal aorta in approximately 80% of the cases [5]. The differential diagnosis of pulmonary sequestration includes bronchial atresia, cystic adenomatoid malformation, intrapulmonary bronchogenic cyst, and arteriovenous fistula. These conditions are differentiated through the finding of an anomalous systemic arterial supply in bronchopulmonary sequestration [1].

Although pulmonary sequestration is a benign disease, severe complications such as hemothorax, fatal hemoptysis, cardiovascular complications, superimposed infections, torsion, and be-

nign and malignant tumours have been reported. If the diagnosis of pulmonary sequestration is certain, these complications necessitate the removal of the pulmonary sequestration lesion. If the diagnosis is uncertain, surgery is the rule of thumb for diagnosis and treatment in pulmonary sequestration [1, 8]. The treatment of extralobar pulmonary sequestration consists of excision of the sequestered lung tissue. Surgical resection is easily performed as the lesion is distinct and separate from the adjacent normal lung tissue. Identification and ligation of anomalous vessels should be carried out carefully during surgical procedure [5, 7, 8]. These may be achieved via open thoracotomy or video-assisted thoracic surgery [7]. In our case, the anomalous pulmonary tissue was removed by mass excision.

In conclusion, extralobar pulmonary sequestration is a rare congenital disorder. Imaging methods such as CT of the chest with intravenous contrast and arteriography can identify the systemic blood supply. Prognosis is excellent after surgical treatment.

## References

1. Huang EY, Monforte HL, Shaul DB. Extralobar pulmonary sequestration presenting with torsion. *Pediatr Surg Int* 2004; 20: 218-20.
2. Ulys A, Samalavicius NE, Cicenias S, *et al.* Extralobar pulmonary sequestration. *Int Med Case Rep J* 2011; 4: 21-3.
3. Aryal G, Pathak V. Bronchopulmonary sequestration presenting as recurrent pneumonia. *WMJ* 2011; 110: 240-2.
4. Prasad R, Garg R, Verma SK. Intralobar sequestration of lung. *Lung India* 2009; 26: 159-61.
5. Rosado-de-Christenson ML, Fraizer AA, Stocker JT, Templeton PA. Extralobar sequestration: Radiologic-pathologic correlation. *Radiographics* 1993; 13: 425-41.
6. Cserni T, Kiss A, Jozsa T, Szölösi Z, Nagy B. Extralobar pulmonary sequestration in the right upper thoracic region. *Respiration* 2007; 74: 215-9.
7. Taylor JA, Laor T, Warner BW. Extralobar pulmonary sequestration. *Surgery* 2008; 143: 833-4.
8. Gezer S, Taştepe I, Sirmali M, *et al.* Pulmonary sequestration: A single-institutional series composed of 27 cases. *J Thorac Cardiovasc Surg* 2007; 133: 955-9.
9. Pugliese JG, Bartholo TP, dos Santos HTA, *et al.* Usefulness of chest CT in the diagnosis of pulmonary sequestration. *J Bras Pneumol* 2010; 36: 260-4.



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