

Intralobar pulmonary sequestrations in adults

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Abstract

Intralobar sequestration accounts for 75% of pulmonary sequestrations. It is characterized by the presence of nonfunctional parenchymal lung tissue, receiving systemic arterial blood supply. We conducted a retrospective medical records review of all patients evaluated and treated in our pulmonary department of military hospital of Tunisia with diagnosis of PS from January 2007 through December 2015. Among them, we report 5 cases of intralobar pulmonary sequestrations operated. There are three women and two men; the mean age is 27.6 years. The sequestration was intralobar in all cases. Clinical presentations were chest pain and productive cough in three cases. Chest X-ray showed left basal opacity in three cases, bilateral basal reticulonodular opacities in one case and round hydric opacity in the right lower lobe in one other case. Computed tomography was performed and revealed an aberrant systemic artery born from the lateral side of aorta supplying a left lower lobe sequestration in four cases and a right lower lobe mass in only one case. The confirmation was operative in all cases and histologic only in three cases. All patients were treated by lobectomy. Only one case presented with a pulmonary sequestration combined with tuberculosis and he was treated firstly by antituberculous chemotherapy. The results were excellent with a favorable clinical course and the mortality was nil.

Keywords: Pulmonary sequestration, Diagnostic, Treatment, Clinical course

Introduction

Pulmonary sequestration is a rare congenital abnormality of the lower airway. It consists of a nonfunctioning mass of lung tissue that lacks normal communication with the tracheobronchial tree and that receives its arterial blood supply from the systemic circulation [1]. There are two types of sequestrations: Extralobar (EPS) and intralobar pulmonary sequestration (IPS). EPS become symptomatic early in life and is often identified on prenatal ultrasound, nevertheless IPS is more commonly identified later in life secondary to recurrent infection [2]. Sequestration is most readily diagnosed by CT or MRI [3]. The confirmation is operative and histological. The standard treatment is resection of the segment or lobe that contains the sequestered tissue; the prognosis is generally favorable [4]. Through our five case reports we discuss the diagnostic difficulties by emphasizing the contribution of modern imaging and surgical treatment.

Case report 1:

A fifty three year old male, smoker (75 pack-years) without past medical history, was admitted to the hospital complaining of hemoptysis, productive cough and left chest pain. Clinical examination was normal. Laboratory values were normal. BK

search in sputum and tumor markers were negative. The chest X-ray revealed bilateral basal reticulonodular opacities and a large left hilum. The bronchoscopy showed an infiltrated bronchial mucosa and expanded spurs at both upper lobes. Bronchial lavage was normal. Bronchial biopsy confirmed the lack of signs of malignancy.

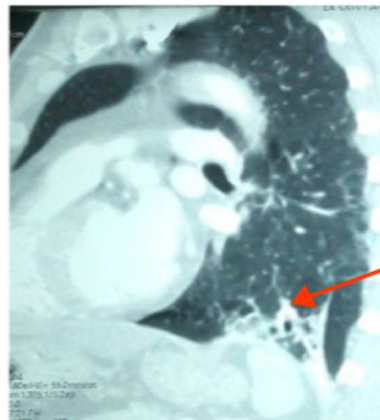


Figure1: Computed tomography scan in 53- year- old male with arrow pointing to an intralobar pulmonary sequestration of the left lower lobe.

Computed tomography (figure 1) was performed and an intralobar

pulmonary sequestration of the left lower lobe was identified and a one centimeter aberrant systemic artery originating from the abdominal aorta supplying the sequestered lobe was noted. Preoperative pulmonary function tests demonstrated an FEV1 of 2.33 liters (73.8% predicted) and FCV of 2.57 liters (65.3% predicted). The patient underwent a left lower lobectomy via a left posterolateral thoracotomy. The aberrant arterial supply was identified. His postoperative course was simple and he was discharged on hospital day ten without complications. He was hospitalized after a month for left encysted pleural effusion regressing by rest and pleural kinesitherapy. The patient at follow up reported a parietal chest pain resistant to symptomatic treatment.

Case report 2:

A seventeen year old female, without medical past history, presented with the complaint of a chronic, productive cough, purulent sputum and dyspnea on exertion. On examination, left rhonchi were found at auscultation. Laboratory values were normal. BK search in sputum and tumor markers were negative. The chest X-ray noticed left posterobasal opacity. The bronchoscopy was normal. Initially, the diagnostic of nontuberculous infectious pneumonitis was retained and the patient was treated with antibiotics but there was no radiologic cleaning. Computed tomography was, then, performed and demonstrated an aberrant systemic artery born from the lateral side of aorta supplying a parenchymal lower lobe posterobasal condensation concluding to a left lower lobe sequestration (Figure 2).



Figure 2: Aberrant arterial supply of a left lower sequestration noted by the red arrow.

Preoperative pulmonary function tests demonstrated an FEV1 of 1.99 liters (58% predicted) and FCV of 2.39 liters (61.6% predicted). The patient underwent a left thoracotomy and a Wedge resection of the destroyed parenchyma. Pathologic review of the surgical piece showed a dystrophic parenchyma characterized by acute inflammatory phenomena. She was subsequently discharged home without complications.

Case report 3:

A twenty eight year old female, without medical past history, was hospitalized suffering from chest pain radiating to the left arm and a history of recurrent respiratory infections. Clinical examination was normal. Laboratory values were normal with the exception of high CRP. BK search in sputum was negative. The chest X-ray showed a left heterogeneous basal opacity. The bronchoscopy showed a left bronchial mucosa infiltrated with hyperemia. Computed tomography and also MRI revealed an intralobar left

lower lobe sequestration with signs of super infection (Figure 3) and a systemic arterial born from the left side of aorta supplying the sequestered parenchyma was identified. Preoperative pulmonary function tests were essentially normal. The patient underwent a left lower lobectomy via a left posterolateral thoracotomy. Her postoperative course was remarkable for sequellar pleural effusion regressing spontaneously.

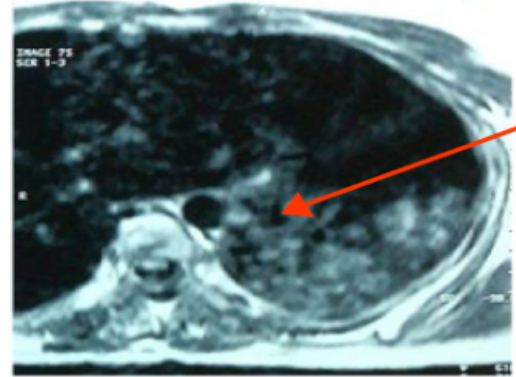


Figure 3: MRI noted an intralobar left lower lobe sequestration.

Case report 4:

Forty eight year female, was admitted to the hospital to explore a radiologic pulmonary opacity fortuitously discovered. Her past medical history was significant for a hypertension and recurrent unilateral epistaxis. Clinical examination was normal. Laboratory values were normal. The chest X-ray revealed a round opacity of hydric tonality in the right lower lobe. BK search in sputum, hydatid serology and tumor markers were negative. The bronchoscopy was normal. Computed tomography was performed and showed a peripheral tissue mass of the apical segment of the right lower lobe measuring three centimeter evoking at the first place a malignancy. Etiological investigations were negative (cervical echography, thyroid function tests, gynecological examination, ultrasound mammography and pelvic echography). Transthoracic biopsy was performed and it was non-contributory. During her hospitalization, a computed tomography of sinus was performed to explore the recurrent unilateral epistaxis and showed a mass of the left maxillary sinus. She was discharged on department of pneumology and transferred to the ENT department and be operated for the mass. Pathologic review concluded to a schwannoma. The patient was hospitalized, then, after two months to complete the investigations. A new bronchoscopy was performed and revealed a stenosis of the right Nelson and the biopsies were negative. A new computed tomography showed a mass with hydric density of the right Fowler with increased volume compared to the previous computed tomography (Figure 4).

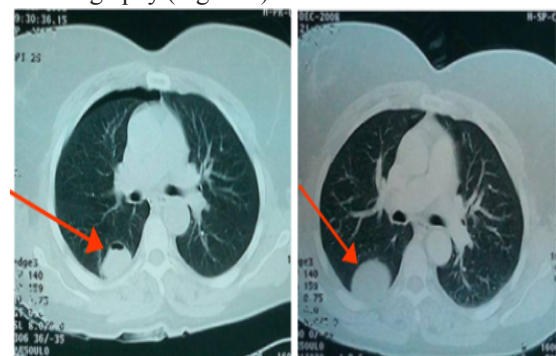


Figure 4: Computed tomography scan with arrow pointing to the

increased volume of the mass in the right Fowler after 2 months.

Preoperative pulmonary function tests were normal. The patient underwent a right posterolateral thoracotomy. Surgical exploration identified a fluctuant cystic tumor, thin walled, at the posterior extremity of the oblique fissure measuring five centimeter and the puncture brought thick chocolate liquid which was supplied by a thin systemic artery; concluding then to a pulmonary sequestration. A cystectomy and a resection of the prominent dome were performed. Pathologic review concluded to an intralobular pulmonary sequestration was retained. She was subsequently discharged home without complications.

Case report 5:

Twenty one year male, smoker (2 pack-years), without past medical history presented with the complaint of chest pain, a chronic productive cough, night sweats, fever at 39 degree and deterioration of general status. Interrogation revealed a tubercular contagion. On examination, the patient was febrile at 39 degree. A crackly sound in the left lung base was found at auscultation. Laboratory values showed an inflammatory syndrome (hyper leukocytosis at 10000, high CRP at 77). The chest X-ray revealed excavated left basal opacity. An infectious necrotizing pneumonia was evoked and the patient was treated with triple antibiotic with clinical and biological improvement. BK search in sputum and cytobacteriological sputum were negative. The bronchoscopy showed a diffuse inflammation of the whole bronchial tracts. BK search in bronchial liquid was negative. A computed tomography was performed and identified a left basal parenchymatous condensation siege of multiple cystic and hydric cavities; corresponding to a left basal intralobular pulmonary sequestration. Two systemic arteries born from the left lateral side of aorta were supplying the sequestered parenchyma (Figure 5).

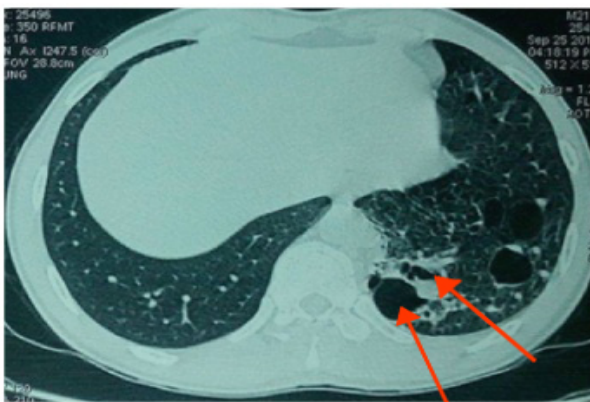


Figure 5: CT view of an intralobar left basal pulmonary sequestration siege of multiple cystic and hydric cavities noted by the red arrows.

In front of the clinical presentation and the tubercular contagion, a super infection by a specific germ, especially tuberculosis, was strongly suspected. The patient was treated with antituberculous chemotherapy during six months. A new computed tomography was performed and revealed a partial regression of the parenchymous condensation inside the sequestration with disappearance of fluid retention at the cystic cavities. The patient underwent a left lower lobectomy via a left posterolateral thoracotomy. His postoperative course was remarkable for a persistent left pneumothorax requiring

a thoracic drainage twice. He was discharged home day twenty six without complications.

Discussion

Pulmonary sequestration is a relatively rare congenital malformation, most diagnosed prenatally or during childhood. Few reports of initial diagnostic are occurring during adulthood [5].

It was firstly described byRektorzik in 1861, but Pryce in 1946 gave it a precise definition and classified it in two groups: intralobular (ILS) and extra lobular sequestration (ELS) [6].

Whereas ILS is contained within normal lung parenchyma, ELS is separated from normal lung and has its own visceral pleura. Almost always, arterial supply to the pulmonary sequestration was from a systemic artery and venous drainage is to pulmonary veins in ILS and to a systemic vein in ELS [7].

Intralobar sequestration theories debate in defining the disease as either a congenital or acquired condition. Theories of the acquired aetiology state that chronic pulmonary infections of a lung tissue disconnected from the normal bronchial tree can cause hypertrophy of a regional systemic artery, hence the aberrant arterial supply. Despite being separated from the bronchopulmonary tree, infections may spread to the sequestered segment from adjacent aerated lung tissue via accessory inter-alveolar connections and canals of Lambert. These theories are supported by the low incidence of other congenital anomalies in association with intralobar sequestration, as opposed to the extralobar type [8].

This rare abnormality has an incidence between 0.15% and 6.45% among all pulmonary malformations and it was found in 1-2% of pulmonary resection [9]. An intralobar sequestration is the most common form of the disease where the lung malformation remains within the visceral pleura of its lobe, whereas the extralobar type corresponds to a true accessory lung, with a separate pleural envelope and an aberrant venous drainage [10].

Frequently, ILS sit in the posterior basal region of the lower lobe. The location in the middle lobe is very rare, as well as that at the upper lobe. The bilateral location is an exception and when it exists, it imposes a particular therapeutic strategy.

This condition can be seen at any age especially during the first two decades. Men seems to be more affected than females [11].

Patients can be totally asymptomatic and, then, the diagnostic can be revealed fortuitously by a radiologic pulmonary opacity. More frequently however, the patient can present with the complaint of various respiratory symptoms such as recurrent pulmonary infections, chest pain or chronic cough, as is the case with our patients (table 1).

Table 1: demographics and characteristics of our patients with intralobar pulmonary

	Age	Gender	Presentation	Imaging-arterial supply identification	Location of intralobar sequestration	Treatment
Patient 1	23 years	male	Hemoptysis Pulmonary infection Chest pain	Identified (CT scan)	Left lower lobe	lobectomy
Patient 2	18 years	female	Pulmonary infection	Identified (CT scan)	Left lower lobe	Wedge resection
Patient 3	28 years	female	Pulmonary infection Chest pain	Identified (CT scan)	Left lower lobe	lobectomy
Patient 4	48 years	female	Pulmonary infection Chest pain	Identified (CT scan)	Right lower lobe	lobectomy
Patient 5	21 years	male	Pulmonary infection Chest pain	Non identified	Left lower lobe	lobectomy

Other clinical presentations are more severe such tuberculosis combined with the pulmonary sequestration (12)and even massive hemoptysis [13].

Chest X-ray is the first step in the diagnostic of pulmonary sequestration. There are three typical imaging manifestations of ILS: a solitary nodule or mass, a cystic or multicystic lesion, or consolidation [14]. The computed tomography is generally sufficient for diagnostic in the most adult cases. It may visualize the mass, abnormal vessels, and associated anomalies, if present. In our experience, the chest CT scan was clearly sufficient to make the diagnosis plus delineate the anatomic features notable for operative planning. Angiography used to be the gold standard for demonstrating arterial supply and venous drainage and is the traditional method of diagnosis in pulmonary sequestration [15]. Currently, Multidetector computed tomography angiography (MDCT) is the modern method for detecting of pulmonary sequestration. There are two principle objectives of MDCT angiography for the investigation of a suspected case of pulmonary sequestration: Firstly, to confirm the presence of an anomalous systemic arterial supplying the sequestered lung and secondary to distinguish pulmonary sequestration from other lung opacities [16].Magnetic resonance imaging is very useful in demonstrating the aberrant arteries and underlying parenchymal changes [14]. Antenatal diagnosis of pulmonary sequestration can be made as early as 18 weeks of gestation by ultrasonography [17].

Lung cancer associated with sequestration in adults deserves special mention. Few reports of malignant neoplasms being involved in or near sequestered segments have been mentioned in the literature. In cases with simultaneous involvement, resection is obviously the mainstay of treatment. In contrast, and giving the very high risk of postoperative mortality, the preservation of the sequestered lung should be considered in patients with low respiratory function and no signs of respiratory infection [18].

Management of asymptomatic pulmonary sequestration is controversial [19].

However, simple conservative treatment, such antibiotics, cannot

eliminate the symptoms and hence, most authors recommend a chirurgical treatment of pulmonary sequestrations because of the inevitable risk of recurrent infections and possible spontaneous complications, especially hemothorax, sometimes fatal. This surgery must be proposed far acute episodes [20].

Surgical resection is the most effective method of treatment and ILS often requires lobectomy. In the other hand, ELS requires an elective resection (sequestrectomy) after careful control of its pedicle.

Surgical resection includes traditional surgery as well as the minimally invasive video-assisted thoracoscopic surgery (VATS). The minimal invasive VATS is a safe procedure with several advantages such as smaller surgical openings, quicker recovery, and cosmetically appealing incisions [21].

Lobectomy is generally accepted as an effective surgical approach for pulmonary sequestration. However, given that pulmonary sequestration is a benign disease, lobectomy may result in a loss of healthy lung tissue. Minimal resection with minimal invasiveness can preserve more pulmonary function. If the sequestered lesion is small and localized, a partial resection, resulting in superior postoperative lung function, may be an alternative to lobectomy. Recent reports have revealed the feasibility of wedge resection by VATS [22,23].

Another approach can be performed in particular cases: Endovascular embolization. It is safe and minimally invasive and can be used for pediatric patients with mild pulmonary infections or patients with only hemoptysis that are unable diagnosis and treatment for pulmonary sequestration to tolerate surgical resection [20].

Conclusion

Intralobar pulmonary sequestration is a rare disease with multiple nonspecific presentations. In the adult, chest CT scan is most helpful, and will provide information about the vascular anatomy that is crucial for operative treatment. Early surgical resection should continue to be the standard of treatment in these patients.

References

1. Landing BH, Dixon LG (1979) Congenital malformations and genetic disorders of the respiratory tract (larynx, trachea, bronchi, and lungs). *Am Rev Respir Dis* 120:151-185.
2. Oermann CM, Redding G, Weisman LE, et al. (2010) Bronchopulmonary sequestration.
3. Walker CM, Wu CC, Gilman MD, Godwin JD, Shepard JA, et al. (2014) The imaging spectrum of bronchopulmonary sequestration. *Curr Probl Diagn Radiol* 43:100-114.
4. Hirai S, Hamanaka Y, Mitsui N, Uegami S, Matsuura Y (2007) Surgical treatment of infected intralobar pulmonary sequestration: a collective review of patients older than 50 years reported in the literature. *Annals of thoracic and cardiovascular surgery* 13:331-334.
5. Hertenberg C, Daon E, Kramer J (2012) Intralobar pulmonary sequestration in adults: three case reports. *Journal of Thoracic Disease* 4:516-519.
6. Atilla Pekcolaklar, Necati Citak, Muzaffer Metin, Abdulaziz Kok, Alper Celikten, et al. (2010) The rare congenital anomaly of pulmonary sequestration experience and review of literature. *Indian Journal of Thoracic and Cardiovascular Surgery* 26:251-254.
7. Gezer S, Tastepe I, Sirmali M, Findik G, Turut H, et al. (2007) Pulmonary sequestration: a single-institutional series composed of 27 cases. *The Journal of thoracic and cardiovascular surgery* 133:955-959.
8. Miller JE, Cotes DJ, Chinn MR (2006) Lung function: theory and measurement in health and disease (6th ed.). Malden, Mass. Blackwell Pub P. 27.
9. Gustafson RA, Murray GF, Warden HE, Hill RC, Rozar GE (1989) Intralobar sequestration a missed diagnosis. *The Annals of thoracic surgery* 47:841-847.
10. Maurice Radermecker, De Bast Y, Delphine Nguyen Dang, Benoît Ghaye, Raymond Limet (2005) Séquestration pulmonaire intralobaire. *Revue Médicale de Liège* 60:839-841.
11. Tashtoush B, Memarpour R, Gonzalez J, Gleason JB, Hadeh A (2015) Pulmonary Sequestration: A 29 Patient Case Series and Review. *J Clin Diagn Res* 9:AC05-8.
12. Huang X, Xu X, Yu C, Fan R, Lu Y (2012) Pulmonary sequestration with tuberculosis confined to the sequestered lung. *Annals of Thoracic and Cardiovascular Surgery* 18:51-55.
13. Ferland N, Couture C, Provencher S (2015) Near-fatal haemoptysis as presentation of a giant intralobar pulmonary sequestration. *Eur Respir Rev* 24:155-156.
14. Walker CM, Wu CC, Gilman MD, Godwin JD, Shepard JA (2014) The imaging spectrum of bronchopulmonary sequestration. *Current problems in diagnostic radiology* 43:100-114.
15. Sancak T, Cangir AK, Atasoy C, Ozdemir N (2003) The role of contrast enhanced three-dimensional MR angiography in pulmonary sequestration. *Interactive cardiovascular and thoracic surgery* 2:480-482.
16. Long Q, Zha Y, Yang Z (2016) Evaluation of pulmonary sequestration with multidetector computed tomography angiography in a select cohort of patients: A retrospective study. *Clinics (Sao Paulo)* 71:392-398.
17. Gajewska-Knapik K, Impey L (2015) Congenital lung lesions: Prenatal diagnosis and intervention. *Seminars in pediatric surgery* 15:156-159.
18. Okamoto T, Masuya D, Nakashima T, Ishikawa S, Yamamoto Y (2005) Successful treatment for lung cancer associated with pulmonary sequestration. *The Annals of thoracic surgery* 80:2344-2346.
19. Prasad R, Garg R, Verma SK (2009) Intralobar sequestration of lung. *Lung India* 26:159-161.
20. Yun-Xi Wang, Jing Zhang, Yang Liu, Xiang-Yang Chu, Yong-Kang Nie, et al. (2016) Case Report Diagnostics and multimodal therapy for pulmonary sequestration. *Int J Clin Exp Med* 9:4734-4741.
21. Gonzalez-Rivas D (2012) Single-port video-assisted thoracoscopic lobectomy. *Minimally Invasive Thoracic and Cardiac Surgery: Springer* 105-111.
22. Lin ZW, Gu J, Xu ST, Ge D, Wang Q (2016) Video-Assisted Thoracoscopic Surgery for Intralobar Pulmonary Sequestration: Wedge Resection Is Feasible in Limited Peripheral Lesions. *Thorac Cardiovasc Surg* 64:456-460.
23. Kestenholz PB, Schneiter D, Hillinger S, Lardinois D, Weder W (2006) Thoracoscopic treatment of pulmonary sequestration. *European journal of cardio-thoracic surgery* 29:815-818.

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