

Bronchopulmonary Sequestration: A Case Report

Mostafa Negmeldin*

Consultant of Respiratory Medicine, Altnagelvin Hospital, Northern Ireland, UK

***Corresponding Author:** Mostafa Negmeldin, Consultant of Respiratory Medicine, Altnagelvin Hospital, Northern Ireland, UK.

Received: December 26, 2018; **Published:** January 28, 2019

Abstract

Background: Bronchopulmonary sequestration, sometimes referred to simply as pulmonary sequestration, is a rare congenital abnormality of the lower airways. It consists of a non-functioning mass of lung tissue that lacks normal communication with the tracheobronchial tree and receives its arterial blood supply from the systemic circulation [1]. BPS can present in several ways, Intra lobar sequestrations (ILS) which are located within a normal lobe and lack their own visceral pleura. Most ILS occur in the lower lobes, but they can occur anywhere within the thorax [2]. Approximately 60 percent are located in the posterior basal segment of the left lower lobe [3]. Extra lobar sequestrations (ELS) are located outside the normal lung, and have their own visceral pleura, with a pedicle that contains the vascular connections. They vary in size, but usually are relatively small compared with the normal lobes. The vast majority is in the left hemi thorax, and the most common location is between the left lower lobe and hemi diaphragm [4].

Case Presentation: A 33-year-old Egyptian lady presented with recurrent lower respiratory tract infections for which she received repeated courses of antibiotics with good response. However, she had suffered socioeconomically from these attacks for a long time. Her clinical picture and radiological findings were suggestive of left-sided intra lobar pulmonary sequestration. Therefore, she was referred to the thoracic surgeon for excision and the diagnosis of bronchopulmonary sequestration was confirmed.

Conclusion: Pulmonary sequestration is a rare congenital pulmonary disorder defined as an area of dysplastic and non-functioning pulmonary tissue with an anomalous systemic blood supply [5]. It has been classically described in two forms, intra lobar sequestrations located within the visceral pleura and surrounded by normal lung, and extra lobar sequestrations which have a separate pleural covering [6].

Keywords: *Bronchopulmonary Sequestration; Visceral Pleura; Intra Lobar Sequestrations (ILS); Extra Lobar Sequestrations (ELS)*

Introduction

Pulmonary sequestration is a rare congenital malformation characterized by a mass of non-functioning lung tissue separated from the normal bronchopulmonary tree and vascularized by an aberrant systemic artery [7,8]. It represents between 0.15 and 6.45% of all pulmonary malformations. No chromosomal abnormality could be identified in any of the patients presenting with such a malformation [9]. Two types of pulmonary sequestration are recognized, depending on whether or not the malformation possesses its own pleural covering. Intra lobar sequestration is an abnormal region within the normal pulmonary parenchyma without its own pleural covering. Extra lobar sequestration corresponds to a true accessory lung, with its own pleural envelope [1,9-11]. There are numerous differences between these two types. Intra lobar sequestrations are not associated with other cardio-pulmonary anomalies, but extra lobar sequestrations may be found in association with cardiac, or more frequently, diaphragmatic anomalies in 50% of cases [12,13]. The fact that intra lobar and extra lobar sequestrations can be found simultaneously suggests that the two forms might share a common embryo pathogenic basis [14].

Clinically, the malformation remains generally silent as long as there is no secondary infection or communication with the bronchial tree or gastrointestinal tract. The diagnostic investigation of choice remains aortography, but it has to be demonstrated whether digital subtraction angiography or MRI angiography will prove more efficient and less invasive [15,16]. Pulmonary angiography and bronchography can help with the diagnosis, but are seldom diagnostic by themselves [17-19]. The mainstay of treatment has always been surgical excision [20].

Case Presentation

This 33-year-old woman presented to our outpatient clinic in June 2018 with cough, greenish sputum and fever. Also, she reported frequent episodes of chest infections which adequately responded to antibiotics. Other system review showed no alarming symptoms. Her examination was unremarkable apart from infra scapular left-sided crepitations while the CXR showed left-sided mass like lesion extending through the middle and lower lung zones. A chest CT scan with contrast revealed a left well defined lower pole lobulated mass measuring 9X6.8 cm with multiple cystic areas of low attenuation and heterogeneous enhancement while it takes its arterial blood supply from a branch of the descending aorta and gives its venous drainage to the left pulmonary vein, a picture is highly suggestive of intrapulmonary sequestration. After that, the diagnosis was confirmed after surgical excision.

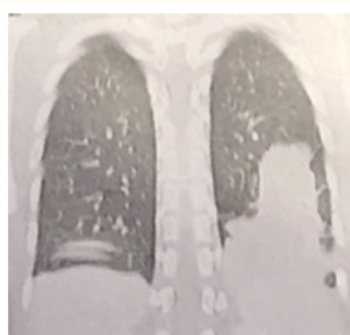


Figure 1: Coronal section for the patient showing the left-sided mass like lesion.

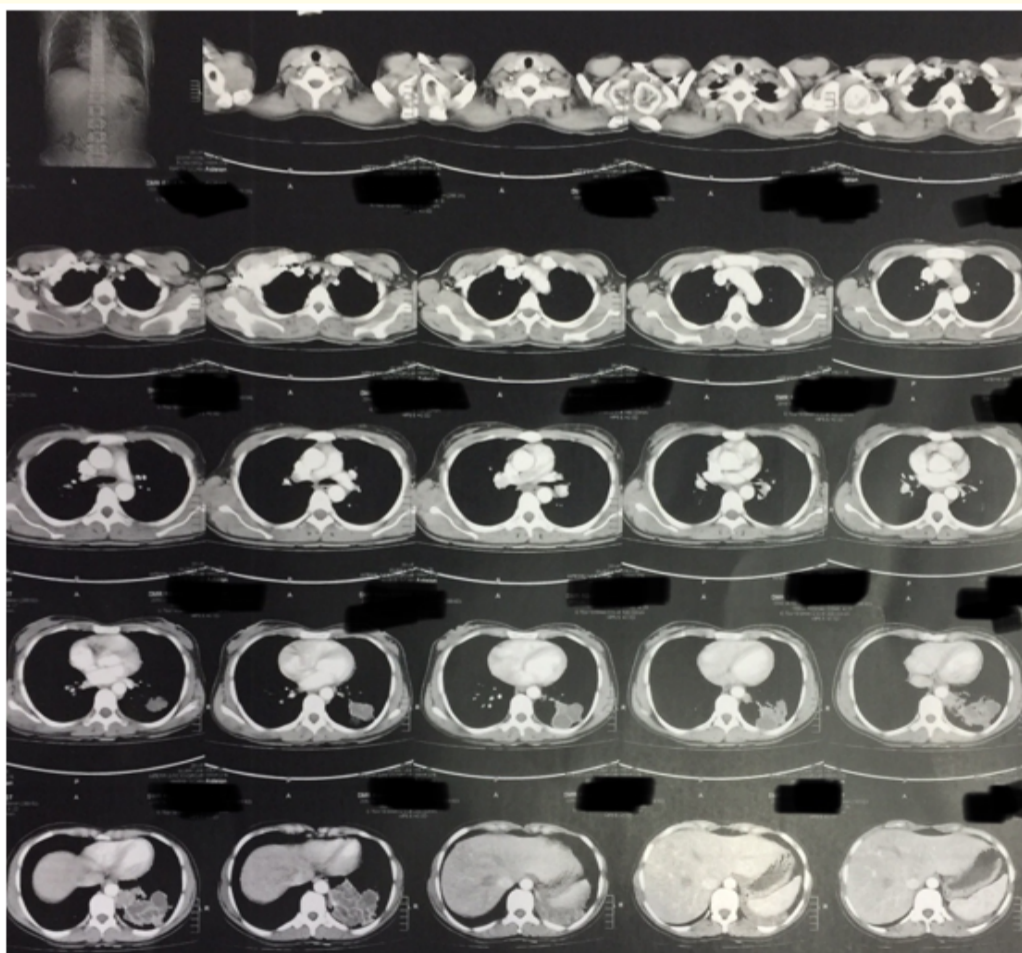


Figure 2: CT chest showing the bronchopulmonary sequestration in the left lower lobe.

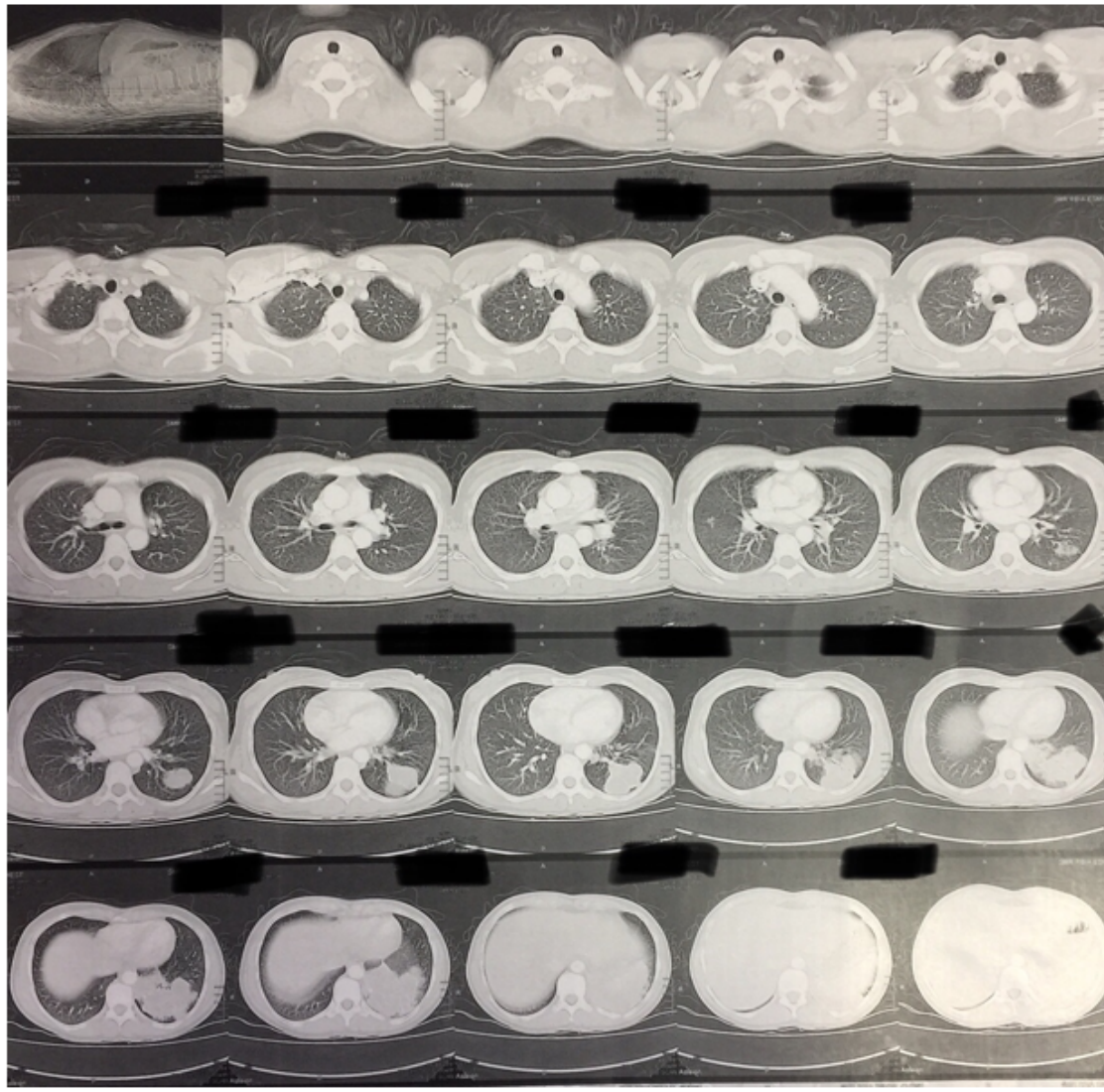


Figure 3: The parenchymal window showing the sequestration.

Discussion

Pulmonary sequestration arises from an abnormal pulmonary development along with pathological vascularization [21]. There are two types of sequestration recognized. Intra lobar pulmonary sequestration is usually contained within the visceral pleura of a pulmonary lobe and its vasculature drains into the pulmonary venous system. Extra lobar pulmonary sequestration is usually within a pleural sheath of its own and the venous drainage is to the azygos or hemiazygos system [22].

In spite of its rarity, pulmonary sequestration should be considered in presence of certain features on a chest X-ray, prompting further investigations in order to avoid encountering an unexpected pathology during surgery. Diagnosis is best confirmed by CT-scan and retrograde aortography. Except for exceptional inoperable cases, intra lobar asymptomatic forms should be excised prophylactically, because of infection hazard [21].

Conclusion

Pulmonary sequestration is a congenital lesion in which a systemic artery supplies a cystic portion of the lung in which its bronchus usually does not communicate with the remainder of the bronchial tree [22]. Given the probability of developing infectious complications, the sequestered tissue should be excised. Excision of the sequestered lung tissue is a safe procedure with a low complication rate. Thoracoscopy seems now to find its place in the surgery of this disease [21].

Bibliography

1. Landing BH and Dixon LG. "Congenital malformations and genetic disorders of the respiratory tract (larynx, trachea, bronchi, and lungs)". *American Review of Respiratory Disease* 120.1 (1979): 151-185.
2. Shanti CM and Klein MD. "Cystic lung disease". *Seminars in Pediatric Surgery* 17.1 (2008): 2-8.
3. Stocker JT, et al. "Cystic and congenital lung disease in the newborn". *Perspectives in Pediatric Pathology* 4 (1978): 93-154.
4. Houda El M., et al. "Antenatal diagnosis of extralobar pulmonar sequestration". *Pan African Medical Journal* 19 (2014): 54.
5. Felker RE and Tonkin ILD. "Imaging of pulmonary sequestration". *American Journal of Roentgenology* 154.2 (1990): 241-249.
6. Savice B., et al. "Lung sequestration: report of seven cases and review of 540 published cases". *Thorax* 34.1 (1979): 96-101.
7. Kravitz RM. "Congenital malformations of the lung". *Pediatric Clinics of North America* 41.3 (1994): 453-473.
8. Jansen D., et al. "Bronchopulmonary sequestration with an aneurysm of the aberrant artery". *Annals of Thoracic Surgery* 60.1 (1995): 193-194.
9. Gdaniez K., et al. "Clinical symptoms and therapy of lung separation". *Progress in Pediatric Surgery* 21 (1987): 86-97.
10. Sade MR., et al. "The spectrum of pulmonary sequestration". *Annals of Thoracic Surgery* 18.6 (1974): 644-658.
11. Boumghar M., et al. "Sequestration pulmonaire intralobaire et surinfection tuberculeuse". *Schweizerische Medizinische Wochenschrift* 109 (1979): 1460-1464.
12. Avishai V., et al. "Extralobar sequestration presenting a massive hemothorax". *Chest* 109.3 (1996): 843-845.
13. Cukier A., et al. "Scimitar sign with normal pulmonary venous drainage and systemic arterial supply". *Chest* 105.1 (1994): 294-295.
14. Savic B., et al. "Lung separation: report of seven case review of 540 published cases". *Thorax* 34 (1979): 96-101.
15. Jeanbourquin D., et al. "Place de l'arteriographie dans le bilan sequestrations pulmonaires". *Annales de Chirurgie Thoracique et Cardio-Vasculaire* 487.3 (1994): 275-276.
16. Donovan CB., et al. "Bronchopulmonary with MR angiographic evaluation". *Angiology* 45.3 (1994): 239-244.
17. Louie HW., et al. "Pulmonary sequestration: 17 year experience at UCLA". *American Surgeon* 59.12 (1993): 801-805.
18. Merlier M., et al. "Aspects anatomo-cliniques des sequestrations pulmonaires". *Annales de Chirurgie Thoracique et Cardio-Vasculaire* 9 (1970): 511-514.

19. Grewal RG and Yip CK. "Intralobar pulmonary sequestration and mediastinal bronchogenic cyst". *Thorax* 49.6 (1994): 615-616.
20. Cerruti MM., *et al.* "Bilateral intralobar pulmonary, sequestration with horseshoe lung". *Annals of Thoracic Surgery* 55.2 (1993): 509-510.
21. N Halkic., *et al.* "Pulmonary sequestration: a review of 26 cases". *European Journal of Cardio-Thoracic Surgery* 14.2 (1998): 127-133.
22. James W Kilman., *et al.* "Pulmonary Sequestration". *Archives of Surgery* 90 (1965): 648-657.

Volume 8 Issue 2 February 2019

©All rights reserved by Mostafa Negmeldin.