Case Report

Intralobar Pulmonary Sequestration with Aspergilloma and a Large Aberrant Artery in a 35 Year Old Man

Manouchehr Aghajanzadeh MD[1*], Ali Alavy MD[2], Gilda Aghajanzadeh MD[3], Sara Massahnia RN[3]

1. *Dept. of Thoracic Surgery, Razi Hospital, Guilan University of Medical Sciences, Rasht, Iran.*
2. *Dept. of Pulmonology, Razi Hospital, Guilan University of Medical Sciences , Rasht, Iran.*
3. *Respiratory Diseases & TB Research Center of Guilan University of Medical Sciences, Razi Hospital, Rasht, Iran.*

(Received 10 Jan 2010; Accepted 29 Feb 2010)

Abstract

A 35 year old man presented with a 4 month history of cough, hemoptysis, malaise, and fever (>38°C) and moderate (6 kg) weight loss. Investigations with CT scan of the chest revealed a cavitary lesion in the lower lobe of the left lung suggesting the presence of a pulmonary aspergilloma, the remaining lung fields being clear. There was no history of any pulmonary infections. Surgical excision of the affected lobe was performed .During surgical exploration we found that the affected lobe receives its blood supply by a large artery from the thoracic aorta. During follow-up his clinical status is excellent.
J Cardiovasc Thorac Res 2010; Vol.2 (1): 43-46

Keywords: *Aspergilloma ý Pulmonary Sequestration ý Aberrant Artery*

**Corresponding Author: Manouchehr Aghajanzadeh MD, Departement of Thoracic Surgery, Razi Hospital, Guilan University of Medical Sciences, Rasht, Iran Tel: +98 131-5550028 Fax: +98 9119063079 Email: smassahnia@yahoo.com*

Introduction

Bronchopulmonary sequestration (BPS) sometimes referred to simply as pulmonary sequestration, is a rare congenital malformation of the lower respiratory tract. 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]. Sequestrations are classified anatomically, as follows: Intralobar sequestration (ILS, also known as intrapulmonary sequestration), in which the lesion is located within a normal lobe and lacks its own visceral pleura; Extralobar sequestration (ELS, also known as extrapulmonary sequestration), in which the mass is located outside the normal lung and has its own visceral pleura. Both types are composed of normal lung tissue, including airway and alveolar elements. A rare variant of sequestration is bronchopulmonary-foregut malformation (BPFM). In this anomaly, the sequestered lung tissue is connected to the gastrointestinal tract. Rare complications include heart failure due to excessive flow through the aberrant artery, massive hemothorax and massive hemoptysis [2-4]. Cases have been reported of fibrous mesothelioma and carcinoma Nocardia asteroides infection and aspergillum Arising within Intralobar sequestration (ILS) [5-7]. Intralobar pulmonary sequestration associated with asymptomatic aspergillosis is a rare case. We describe the case of a 35-year-old man with undiagnosed intralobar pulmonary sequestration with aspergilloma.

A 35-year-old man presented with a 4-month history of cough, hemoptsis, malaise, and fever (>38°C). Moderate (6 kg) weight loss was also reported. He was a current heavy smoker (40 packs/year). His past medical history included only a depression treated with tricyclic drugs. On admission he had no respiratory distress. Pulse was regular at 78 beats/min and blood pressure was 140/80 mm Hg. Physical examination was normal. There was neither cyanosis nor digital clubbing. With the patient breathing room air, arterial blood-gas analysis showed a pH of 7.40, an arterial carbon dioxide tension of 37 mm Hg, and an arterial oxygen tension of 77 mmHg. Blood laboratory studies disclosed no further abnormalities. Chest radiography and computed tomographic scanning revealed a left-sided basal cavitary pattern (Fig 1).

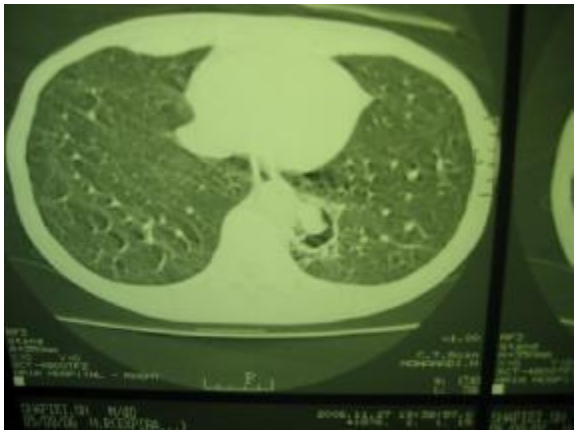

Fig 1- Computed tomographic scan of the thorax showing a cavitary lesion in the left lower lobe

Broncscopy with aspiration was not diagnostic. With diagnosis of a symptomatic pulmonary aspergilloma, the patients was candidated for thoracotomy. The patient underwent left postero-lateral thoracotomy. At exploration, a gross mass was found in the left lower lobe. Careful separation of adhesions between the lobe and the diaphragm and chest wall allowed the identification of an aberrant large artery of descending thoracic aorta origin. The artery was double ligated and divided (Figure 2, 3).

A left lower lobectomy was performed without further difficulty. Histopathologic study revealed the wall of an aspergilloma, a partially eroded lining of metaplastic squamous epithelium, overlying chronic inflammation, lung parenchyma with evidence of atelectasis and bronchectasis and cystic changes (Figure 3). During follow-up the patient was asymptomatic, with no radiographic signs of disease. He has also regained his body weight.

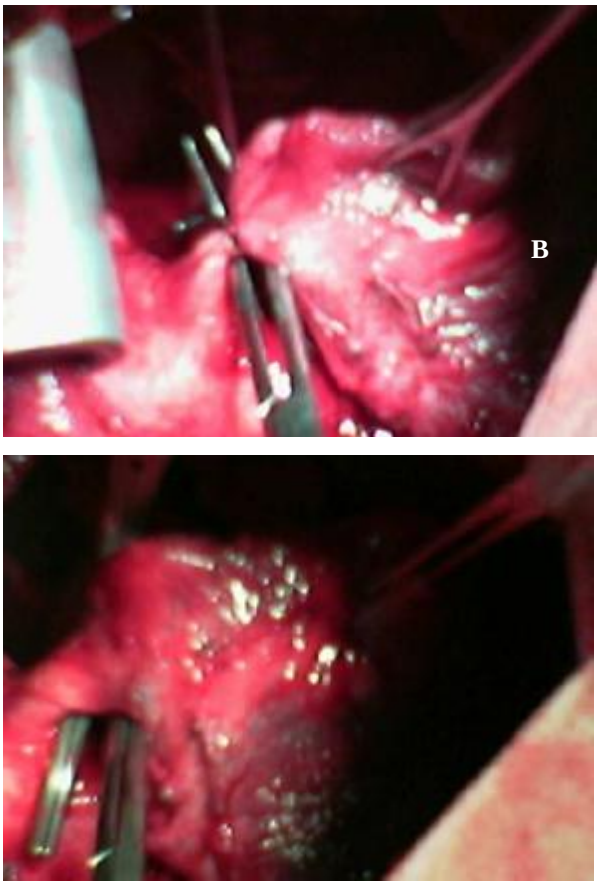

Fig 2- (A, B):Show Large Aberrant Artery and Involved Lobe.

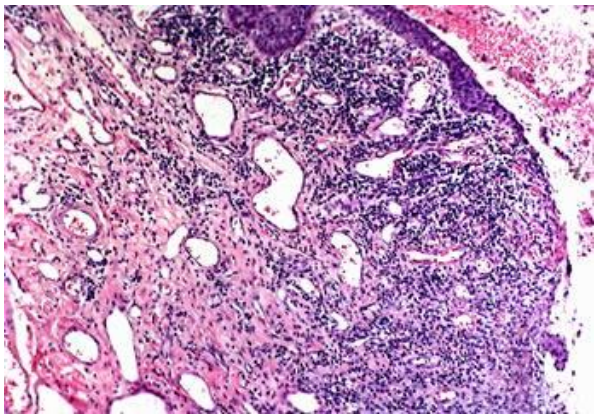

Fig 3 pathology of the patient- The wall of the aspergilloma shows a partially eroded lining of metaplastic squamous epithelium, overlying chronically inflamed granulation tissue and scar. Note the large vascular channels with little smooth muscle in their walls. These branches of the bronchial artery can be a source of bleeding.

Discussion

A pulmonary sequestration refers to nonfunctioning lung tissue with no direct communication with the tracheobronchial tree and with an anomalous blood supply from the systemic vessels. Prycein first described this entity, and subdivided it morphologically into ILS and ELS. ILSs comprises 75% of sequestrations, and are encountered in young adults, usually by the age of 20 years[8] .The ILS shares the visceral pleura with the normal lung tissue, and receives its blood supply from the aorta in 94% of cases and it drains into the pulmonary venous system in 95% of cases. Extralobar sequestration is almost exclusively discovered in infancy, due to early onset of symptoms[4]. The sequestered lung has its own visceral pleura, and may even occur outside the thorax. The ELS blood supply is usually systemic, from branches of the aorta (80% of cases); however, the venous drainage is mainly via the azygos-hemiazygos system (80% of cases) [2-4]. The association of ELS with other congenital anomalies (65% of the cases), such as bronchopulmonary foregut malformation, supports an embryonic origin of this entity[1]. In contrast, ILS may be acquired, with endobronchial supply to the infected area and subsequent hypertrophy of small systemic arteries that supply this area[7,9]. As our case which had a large and hypertrophied artery. Obstruction triggers recurrent pneumonias. Both forms are usually located in the posterobasal segment of the left lower lobe and in the posterior vascular costodiaphragmatic sulcus between the left lower lobe and the hemidiaphragm, but may occur anywhere in the thorax, or even below the diaphragm[1,2,8]. Pathologic characteristics include extensive fibrosis and chronic inflammation and cystic changes replacing the lung parenchyma[8] . Contrast CT of the chest reveals the complex character of the lesion with its anomalous blood supply. Preoperative aortogram may confirm the diagnosis and identify the vascular supply, in order to avoid catastrophic intraoperative bleeding[10]. The standard treatment is resection of the segment or lobe that contains the sequestered tissue[8,11]. Other options are ligation or embolization of the Vascular pedicle. Some authors advocate conservative

antibiotic treatment with surgical intervention only in cases of repeated infections; however, this strategy is rarely used because of the potential of bleeding complications from repeated infections[8,10,12]. Infection is the major complication of pulmonary sequestration and the sequestered tissue is frequently infected with bacterial pathogens.[12] Infection may present as an acute or chronic respiratory illness, with clinical and radiographic findings consistent with pneumonia or abscess. An aspergilloma usually arises in a pre-existing cavity in the lungs. A number of infections and other conditions can produce these cavities including tuberculosis, sarcoidosis, neoplasms, cystic fibrosis other fungal infections such as histoplasmosis or coccidioidomycosis, nocardia or invasive aspergillosis. Thus, any condition that causes cavitation in the lungs may subsequently be associated with the development of an aspergilloma. There is only few previous reports of aspergilloma species infection with intralobar bronchopulmonary sequestration[11,12]. One of them was a 65-year-old woman with intrapulmonary sequestration, anomalous systemic arterial supply to the left lower lobe and aspergillosis who underwent left lower lobectomy and ligation of an anomalous artery by Video-Assisted Thoracoscopic surgery (VATS) [11]. During 15 year of my practice I operated three cases of intralobar pulmonary sequestration without preoperative diagnosis. Two of them with symptoms and signs of bronchectasis in right lower lobe and other with left lower lobe mass and their blood supply in two cases was from thoracic aorta and in another case from abdominal aorta.

Conclusion

Our patient demonstrated that aspergillos infection can occur in the intralobar pulmonary sequestration. In such conditions an underlying lung malformation, such as pulmonary sequestration, should be suspected. Prompt imaging with attention to vascular anatomy will lead to early diagnosis and treatment.

References

1. Gerle RD, Jaretzki A III, Ashley CA, Berne AS. Congenital bronchopulmonary-foregut malformation: Pulmonary sequestration communicating with the gastrointestinal tract. ***N Engl J Med*** 1968; 278: 1413-1419.
2. Levine, MM, Nudel, DB, Gootman, N, et al. Pulmonary sequestration causing congestive heart failure in infancy: a report of two cases and review of the literature. ***Ann Thorac Surg*** 1982; 34:581.
3. Avishai, V, Dolev, E, Weissberg, D, et al. Extralobar sequestration presenting as massive hemothorax. ***Chest*** 1996; 109:843.
4. Rubin, EM, Garcia, H, Horowitz, MD, Guerra, JJ Jr. Fatal massive hemoptysis secondary to intralobar sequestration. ***Chest*** 1994; 106:954.
5. Paksoy, N, Demircan, A, Altiner, M, Artvinli, M. Localised fibrous mesothelioma arising in an intralobar pulmonary sequestration. ***Thorax*** 1992; 47:837.
6. Gatzinsky, P, Olling, S. A case of carcinoma in intralobar pulmonary sequestration. ***Thorac Cardiovasc Surg*** 1988; 36: 290.
7. Muhammad Shibli, MD, Cliff Connery, MD, Janet M. Shapiro, MD Intralobar and Extralobar Bronchopulmonary Sequestration Complicated by Nocardia asteroids. ***Infection South Med J*** 2003: 96: 78-80.
8. Pryce DM. Lower accessory pulmonary artery with intralobar sequestration of lung: A report of seven cases. ***J Pathol*** 1946; 58: 457-467.

9-Stocker JT, Malczak HT. A study of pulmonary ligament arteries: Relationship to intralobar pulmonary sequestration. ***Chest*** 1984; 86: 611-615.

10.Frazier AA, Rosado de Christenson ML, Stocker JT, Templeton PA. Intralobar sequestration: Radiologic-pathologic correlation. ***Radiographics*** 1997; 17: 725-745.

11. Sato H, Watanabe A, Yamaguchi T, Harada N, Yamauchi A, Inoue S, Abe T. Pulmonary Sequestration Associated With Asymptomatic Aspergillosis. ***Ann Thorac Cardiovasc Surg***. 2005; 11:41-44.
12. Tanaka M., Miyamoto H., Sakao Y., Harada R., Hata E. A case report of intralobar sequestration associated with lung aspergillus. Nippon Kyobu Geka Gekkai Zasshi 1995; 43: 366-370.