Case Report

Intralobar Pulmonary Sequestration Presenting as Chronic Non-productive Cough

Ravindra Gudavalli[1], Carol C. Farver[2], David P. Mason[3] and Umur Hatipoglu[1]

Departments of Pulmonary and Critical Care Medicine, Respiratory Institute[1], Anatomic Pathology, Pathology and Laboratory Medicine Institute[2] and Cardiothoracic Surgery, Cardiovascular Institute[3], Cleveland Clinic Foundation, Ohio, USA

Abstract

We report a case of a 60-year-old female who was known to have intralobar pulmonary sequestration and her only symptom was chronic cough. She had no history of infections and surgical resection led to complete resolution of her chronic cough. **[Indian J Chest Dis Allied Sci 2015;57:23-25]**

Key words: Pulmonary sequestration, Chronic cough, Surgery.

Introduction

Pulmonary sequestration (PS) is a rare anomaly representing 0.2% to 6.5% of all pulmonary malformations. Intralobar PS, first described by Pryce in 1946, accounts for about 75% of PS case reports in the literature.[1] Recurrent episodes of infection are the most common complication associated with intralobar PS and surgical resection is usually recommended even in asymptomatic patients. Chronic non-productive cough is an uncommon presentation of PS. We recently diagnosed a patient presenting with chronic non-productive cough to have an intralobar PS.

Case Report

A 60-year-old female presented with non-productive cough of 5 months duration. Her symptoms worsened after a bout of upper respiratory infection. She was treated initially with oral antibiotics and inhaled steroids but without any improvement. Cough was present throughout the day but had no nocturnal component. She had been treated in the past with inhaled steroids for possible variant asthma but had no symptomatic benefit. She had a remote 6-pack years of smoking history.

Past medical history of the patient was significant for a pre-operative provisional diagnosis of intralobar PS; established incidentally on a pre-operative chest radiograph 6 years ago. Subsequent imaging indicated stability and no further investigations were performed. She had never been hospitalised for any respiratory infections. She was recommended to have elective surgery but she deferred.

Physical examination revealed clear lungs with no abnormal signs. Spirometry was normal. A subsequent methacholine challenge test was negative. A computed tomography scan of the chest revealed a 36 mm x 23 mm low attenuation lesion supplied by a large artery arising from the descending thoracic aorta (Figure 1).

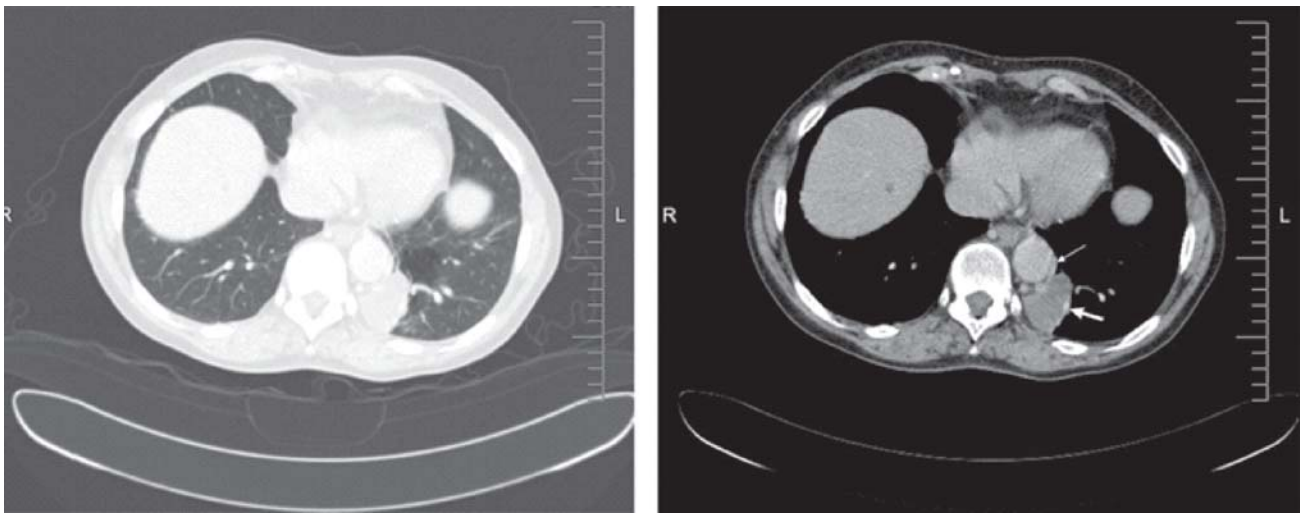

Figure 1. Lung (left) and mediastinal (right) windows of the computed tomographic images demonstrating feeding artery originating from aorta (thin arrow) and intralobar sequestration in the left lower lobe (thick arrow).

[Received: October 12, 2012; accepted after revision: March 6, 2014]

Correspondence and reprint requests: Dr Umur Hatipoglu, Department of Pulmonary and Critical Care Medicine, Respiratory Institute, Cleveland Clinic, 9500 Euclid Avenue, A90 Cleveland, Ohio-44195, USA; E-mail: hatipou@ccf.org

The patient underwent a left lower lobe segmentectomy. The histopathology of the resected specimen showed chronic organising pneumonia with areas of honey-comb change and non-necrotising granulomas (Figure 2). Special stains for bacterial or fungal pathogens were negative. The granulomatous inflammation was deemed non-specific.

After the surgical resection of the interlobal sequestration, her cough resolved completely. The patient remained symptom-free during 3 years of clinical follow-up.

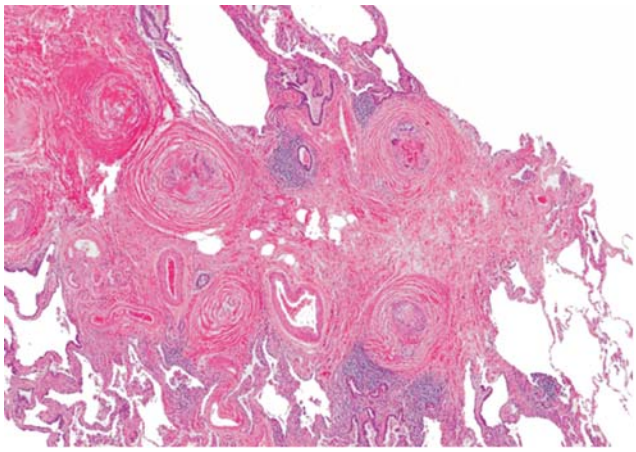

Figure 2. Histopathological photograph showing non-necrotising granulomas in the setting of chronic organising pneumonia. (Haematoxylin and eosin ×10).

Discussion

Pulmonary sequestration is an uncommon condition wherein a portion of lung tissue has absent or abnormal communication with the tracheobronchial tree and has an abnormal vascular supply.[2] Two types of PS have been described: intralobar sequestration (ILS) and extralobar sequestration (ELS). The ILS is contained within the visceral pleura with venous drainage into the pulmonary veins. The ELS is separated from the normal lung and is outside the visceral pleura with venous drainage into a systemic vein. The arterial supply in both ILS and ELS is from a systemic artery. Controversy exists with regards to the origin and development of PS and various theories have been proposed. The congenital origin of the ILS[3] has been questioned by Stocker and Malczak[4] who studied the pulmonary ligaments of children without any congenital or pulmonary vascular anomalies and demonstrated systemic arteries within the pulmonary ligament in 10 of 11 cases. Bronchial obstruction, pneumonia, pulmonary artery occlusion, pleuritis, and parasitisation of pulmonary ligament have been proposed to explain the acquired origin theory of the development of ILS.[4,5]

The ILS accounts for 75% of PS cases and is typically located in the medial or posterior basal segments of the left lower lobe. Our patient presented with a left lower lobe posterior basal segment ILS. Savic *et al*[1] described 133 ELS and 400 ILS cases in their review of the literature. The ILS is usually diagnosed in pediatric or adolescent patients though more than 20 cases have been reported in the literature of patients older than 40 years.[6,7] In the series by Petersen *et al*,[6] 40% (6/15) of these adult patients were asymptomatic at the time of diagnosis while in symptomatic patients cough and haemoptysis were the most common presenting symptoms. In the two largest single institution case series,[8,9] most adult patients with ILS were symptomatic at the time of presentation. Symptomatic patients have recurrent episodes of bronchitis or pneumonia. Our patient was diagnosed in her late 50's, had no history of recurrent infectons and presented only with non-productive cough. Complications associated with PS described in the literature include fatal haemoptysis,[10] cardiovascular complications,[11] fungal, bacterial and mycobacterial including atypical infections,[12-14] and malignant degeneration.[15] The severe complications associated with ILS necessitate surgical removal.

Diagnosis of sequestration is initially suspected based on chest radiograph.[16] Retrograde angiography has been the "Gold standard" for defining the vascular supply and diagnosing PS but the newer non-invasive techniques such as magnetic resonance angiography or computed tomographic angiography may demonstrate the aberrant vessels and underlying parenchymal changes and obviate the need for the invasive testing in most cases. Delineation of the anomalous vasculature prior to surgery by non-invasive or invasive angiography minimises the occurrence of fatal intraoperative haemorrhagic complications.

Surgery is indicated in most patients with ILS particularly those who are symptomatic.[16] The optimal management of asymptomatic patient is unclear but most clinicians recommend surgical resection since these lesions may be associated with serious complications. A segmentectomy or lobectomy is usually indicated for symptomatic patients with ILS although pneumonectomy is rarely needed. Major complications reported are intra-operative haemorrhage, broncho-pleural fistula and empyema. Our patient had an uneventful recovery after resection of sequestered segment with resolution of her chronic cough.

References

1. Savic B, Birtel FJ, Tholen W, Funke HD, Knoche R. Lung sequestration: report of seven cases and review of 540 published cases. *Thorax* 1979;34:96–101.
2. Gottrup F, Lund C. Intralobar pulmonary sequestration: a report of 12 cases. *Scand J Respir Dis* 1978;59:21–29.
3. Boyden EA. Bronchogenic cysts and the theory of intralobar sequestration: new embryologic data. *J Thorac Surg* 1958; 35:604–16.
4. Stocker JT, Malczak HT. A study of pulmonary ligament arteries: relationship to intralobar pulmonary sequestration. *Chest* 1984;86:611–5.

5. Bolca N, Topal U, Bayram S. Bronchopulmonary sequestration: radiologic findings. *Eur J Radiol* 2004;52:185–91.
6. Petersen G, Martin U, Singhal A, Criner GJ. Intralobar sequestration in the middle-aged and elderly adult: recognition and radiographic evaluation. *J Thorac Cardiovasc Surg* 2003;126:2086–90.
7. Van Raemdonck D, De Boeck K, Devlieger H, Demedts M, Moerman P, Coosemans W, *et al*. Pulmonary sequestration: a comparison between pediatric and adult patients. *Eur J Cardiothorac Surg* 2001;19:388–95.
8. Halkic N, Cuenoud PF, Corthesy ME, Ksontini R, Boumghar M. Pulmonary sequestration: a review of 26 cases. *Eur J Cardiothorac Surg* 1998;14:127–33.
9. Gezer S, Tastepe I, Sirmali M, Findik G, Türüt H, Kaya S, *et al*. Pulmonary sequestration: a single-institutional series composed of 27 cases. *J Thorac Cardiovasc Surg* 2007; 133:955–9.
10. Hofman FN, Pasker HG, Speekenbrink RG. Hemoptysis and massive hemothorax as presentation of intralobar sequestration. *Ann Thorac Surg* 2005;80:2343–4.
11. Fabre OH, Porte HL, Godart FR, Rey C, Wurtz AJ. Long-term cardiovascular consequences of undiagnosed intralobar pulmonary sequestration. *Ann Thorac Surg* 1998;65:1144–6.
12. Yatera K, Izumi M, Imai M, Ikegami T, Miyazaki N, Kido M. Intralobar sequestration with tuberculous infection confined to the sequestrated lung. *Respirology* 2005;10:685–8.
13. Freixinet J, de Cos J, Rodriguez de Castro F, Julia G, Romero T. Colonisation with aspergillus of an intralobar pulmonary sequestration. *Thorax* 1995;50:810–11.
14. Shiota Y, Arikita H, Aoyama K, Horita N, Hiyama J, Ono T, *et al*. Pulmonary sequestration associated by mycobacterium intracellulare infection. *Intern Med* 2002;41:990–2.
15. Gatzinsky P, Olling S. A case of carcinoma in intralobar pulmonary sequestration. *Thorac Cardiovasc Surg* 1988;36:290–91.
16. Yucel O, Gurkok S, Gozubuyuk A, Caylak H, Sapmaz E, Kavakli K, *et al*. Diagnosis and surgical treatment of pulmonary sequestration. *Thorac Cardiovasc Surg* 2008; 56:154–7.