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Lung Sequestration: A Rare Cause for Pulmonary Symptoms in Adulthood

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Key Words

Bronchopulmonary malformation · Lung sequestration · Hemoptysis · Congenital pulmonary disorder · Aberrant blood supply

Abstract

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Background: Lung sequestration is a rare congenital pulmonary disorder and is usually diagnosed in children with recurrent pulmonary infections. Lung sequestrations are not commonly found to be a cause of respiratory symptoms in adults. **Objectives:** It was the aim of this study to show that pulmonary sequestration is rare in advanced age and can be accompanied by severe pulmonary symptoms. *Methods:* We conducted a case series analysis of patient characteristics, symptoms, diagnosis and treatment of 11 adults with a lung sequestration at the Thoraxklinik Heidelberg between 2001 and 2009. Results: From 2001 to 2009, intralobar lung sequestration was diagnosed and treated in 11 adults aged 19 to 58 years with an average age of 39.9 \pm 11.3 years and a male:female distribution of 5:6. In 3 patients (27.3%), the predominant symptom was hemoptysis. Recurrent pulmonary infections occurred in 1 patient (9.1%); pneumonia and lung abscess were detected in 2 patients (18.2%). In 3 cases

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Fax +41 61 306 12 34 E-Mail karger@karger.ch www.karger.com © 2011 S. Karger AG, Basel 0025-7931/11/0825-0445\$38.00/0 Accessible online at: www.karger.com/res (27.3%), dry cough was the predominant symptom, and in only 2 cases (18.2%), lung sequestration was asymptomatic. Eight patients (72.7%) were diagnosed by imaging techniques prior to surgery. In 3 cases (27.3%), diagnosis was made intraoperatively and by pathological examination. Surgical intervention included 7 lobectomies (63.6%), 3 wedge resections (27.3%) and 1 (9.1%) segmentectomy. **Conclusion:** Lung sequestration in adults is rare, but it can cause severe pulmonary symptoms. In cases of recurrent pulmonary infections of identical localization or recurrent hemoptysis, lung sequestration should be considered in order for the diagnosis to be made rapidly. Surgical resection is the treatment of choice. Copyright © 2011 S. Karger AG, Basel

Introduction

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In 1777, Huber first described the aberrant blood supply to the lung [1]. It is defined as functionless lung parenchyma without connection or with abnormal connection to the tracheobronchial system and has an abnormal blood supply. Lung sequestration is very rare and only represents 0.15–6.4% of congenital pulmonary abnor-

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Fig. 1. a CT scan with contrast medium. Coronal section. Intrapulmonary lung sequestration in the left lower lobe. Systemic blood supply originating from the aorta thoracica descendens. **b** CT scan with contrast medium. Transverse section. Intrapulmonary lung sequestration in the left lower lobe. The ground-glass opacity in the left lower lobe results from recent hemoptysis.



Table 1. Patient characteristics and symptoms

Patient No.	Sex	Age at diagnosis years	Symptoms	
1	m	19	hemoptysis	
2	m	29	hemoptysis	
3	m	35	pneumonia	
4	f	36	hemoptysis	
5	f	36	dry cough, pneumonia	
6	f	37	lung abscess	
7	m	41	recurrent pulmonary infections	
8	f	46	dry cough, hemoptysis	
9	f	47	asymptomatic	
10	f	54	dry cough, pneumonia in childhood	
11	m	59	asymptomatic	

malities [2]. The most important presenting symptoms are recurrent pulmonary infections or productive cough. Severe hemoptysis as the first symptomatic manifestation has been reported very rarely [3]. In most cases, lung sequestrations are diagnosed in childhood or adolescence. In addition to taking a detailed history and physical examination, imaging procedures such as CT or MR angiography are important to delineate aberrant blood supply [4–6]. In confirmed cases, surgical resection is the treatment of choice.

The aim of this case series analysis is the evaluation of symptoms, diagnostic and treatment modalities of lung sequestration in adults. This retrospective study demonstrates that lung sequestration in advanced age is rare and can be accompanied by severe pulmonary symptoms.

Methods

The study protocol of our retrospective case series analysis was presented to and approved by our local ethics committee. All patients diagnosed with lung sequestration at the Thoraxklinik Heidelberg between February 2001 and June 2009 were included. The cases were identified by reviewing surgical and pathological reports as well as by corresponding diagnosis coding. In addition to the assessment of age and symptoms, the method of diagnosis and treatment were evaluated. Lung sequestration was diagnosed by typical radiologic findings, surgery or histological examination.

Results

Patient Characteristics and Symptoms

Lung sequestration was diagnosed in 11 adults, aged 19–58 years, with an average age of 39.9 \pm 11.3 years and a male:female distribution of 5:6. Their presenting symptoms are shown in table 1. In only 2 cases (18.2%) lung sequestration was asymptomatic.

Diagnostic Workup

In 8 of the 11 patients (72.7%), lung sequestration was diagnosed by imaging techniques prior to surgery. The diagnosis is based on the identification of the aberrant arterial vessel originating from the aorta. In 5 of these 8 preoperatively diagnosed lung sequestrations, the descending thoracic aorta provided the abnormal arterial blood supply (fig. 1). In 3 patients, the arterial supply originated infradiaphragmatically, in 1 case from the left gastric artery and in 2 cases from the abdominal aorta (in 1 of these latter patients, with an intrapulmonary aneurysm of the aberrant vessel) (fig. 2). In the last of the 8 preoperatively diagnosed lung sequestrations, endobron-

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Fig. 2. a CT angiography. Intrapulmonary lung sequestration in the right lower lobe with aneurysm of the aberrant arterial vessel originating from the aorta abdominalis. **b** CT angiography. Coronal section. Intrapulmonary lung sequestration in the right lower lobe with aneurysm of the aberrant arterial vessel.

chial ultrasound (EBUS) allowed visualization of the aberrant vessels (fig. 3). In the remaining 3 of the 11 patients (27.3%), lung sequestration was diagnosed intraoperatively and by the pathologist. In 1 of these patients, a pulmonary mass was detected by CT, initially raising suspicion of a bronchial carcinoma. Only by surgical intervention and histological examination malignancy could be excluded and lung sequestration was confirmed. In the second patient, surgery was performed due to pulmonary abscess formation, again detected by CT. In the third patient, the indication for lobectomy was resection due to recurrent pulmonary infections.

Treatment

All lung sequestrations in the 11 patients occurred in the lower lobes: in 6 patients (54.5%) in the left lower lobe, in 5 patients (45.5%) in the right lower lobe. An extrapulmonary sequestration was not found. In all patients therapy consisted of surgical resection. The extent of resection was chosen according to the size and type of the sequester and the associated malformations. Lobectomy was performed in 7 patients (63.6%), wedge resection in 3 patients (27.3%) and an anatomical resection of 1 segment in 1 patient (9.1%). In 1 patient, a combined thoracic and abdominal procedure was necessary. At first, transperitoneal ligation of the aberrant vessel between the celiac trunk and the superior mesentery artery was performed, followed by resection of the lower lobe during thoracotomy. Surgery and the postoperative course were unremarkable in 10 of 11 patients. Re-thoracotomy was necessary in 1 patient due to bleeding. None of the patients experienced any further complaints after the surgery (table 2).

Discussion

Lung sequestration is a rare congenital pulmonary malformation consisting of functionless lung tissue without connection or with an abnormal connection to the



Fig. 3. a Endobronchial ultrasound. The blood vessel (arrow) is surrounded by lung tissue. Typical ultrasound image of an atelectasis or inflammatory tissue with white echo spots. **b** Endobronchial ultrasound. Abnormal blood vessel (arrow).

Patient No.	Diagnosis	Localization	Origin of the aberrant arterial vessel	Therapy
1	CT + EBUS		descending thoracic aorta	lobectomy
3	CT + MRA	LLL	descending thoracic aorta	lobectomy
4 5	CTA MRT	RLL LLL	abdominal aorta, aneurysm descending thoracic aorta	lobectomy lobectomy
6 7	intraoperative CT + MRA	RLL LLL	descending thoracic aorta abdominal aorta	lobectomy wedge resection
8	CT	LLL	descending thoracic aorta	lobectomy
9 10	MRA intraoperative	RLL LLL	left gastric artery descending thoracic aorta	wedge resection segment 10 resection
11	intraoperative	RLL	descending thoracic aorta	wedge resection

Table 2. Diagnosis, localization of sequestration, origin of the aberrant arterial vessel and therapy

EBUS = Endobronchial ultrasound; LLL = left lower lobe; MRA = MR angiography; CTA = CT angiography; RLL = right lower lobe.

tracheobronchial system and with abnormal blood supply. Venous drainage is accomplished by pulmonary veins. It is also described that lung sequestration can develop postnatally following recurrent pulmonary infections and bronchial obstruction [7]. Lung sequestration must be distinguished from Scimitar syndrome defined by partial anomalous pulmonary venous connection [8].

There are two forms of lung sequestration: intrapulmonary sequestration, representing 85% of all lung sequestrations, and extrapulmonary sequestration. In 1979, Savic et al. [9] presented an analysis of 533 cases of lung sequestration with a predominance of 400 intralobar sequestrations versus 133 extralobar sequestrations. Intrapulmonary sequestration is surrounded by normal lung parenchyma, and in 90% of cases, is located in the lower lobe. However, extrapulmonary sequestrations are separated from the normal lung tissue. They are mostly found between the diaphragm and lower lobes, in 90% on the left side. Only in rare cases they are located infradiaphragmatically. Extrapulmonary lung sequestrations are more frequently associated with other congenital pulmonary or cardiac malformations and are 4 times more common in males than in females. Both forms of lung sequestration have an abnormal arterial blood supply: in 74%, the aorta provides the arterial blood supply, and in 96%, venous drainage is accomplished by pulmonary veins [2]. In our retrospective case series analysis of adults, only intrapulmonary sequestrations were found. Concomitant malformations could not be detected.

The most common symptoms of lung sequestration include recurrent pulmonary infections and productive cough [3]. In 1989, a retrospective analysis of 10 patients with intralobar sequestration demonstrated that 90% presented with recurrent pulmonary infection, chronic cough and intermittent fevers [10]. Hemoptysis as the first manifestation of lung sequestration is rather rare. The most common causes for hemoptysis are lung cancer [11] and inflammatory processes like tuberculosis, bronchiectasis or bronchitis [10], depending on the local circumstances. In Europe, lung cancer is the most common cause; in the developing world, inflammatory processes are the main reason. In <1% of cases, lung sequestrations are responsible for expectoration of blood [13]. In 2007, Gezer et al. [3] published a case series of 27 children and adults suffering from lung sequestration. Twenty out of these 27 patients had symptoms preoperatively. The most common symptom was recurrent pneumonia in 50%, and hemoptysis occurred in only 10%. However, in our analysis, 27.3% of the patients suffered from hemoptysis, requiring rapid evaluation and treatment.

Intralobar sequestration is diagnosed at age ≤ 20 years in approximately 50-60% of cases and it is rarely found in patients >40 years [4]. Lung sequestration can be diagnosed antenatally by ultrasound at 18-19 weeks of gestation [14]. These antenatally diagnosed sequestrations often spontaneously involute or disappear completely. However, persistent lung sequestration may present with recurrent infection or cardiac failure later in childhood. After birth, diagnosis of lung sequestration requires imaging procedures to delineate aberrant blood supply in addition to taking the history and physical examination. Traditionally, the diagnosis of lung sequestration has been made by arterial angiography. Today, noninvasive imaging techniques, like CT or MR angiography [4-6], are used to demonstrate aberrant vessels and parenchymal disorders. A CT scan provides evaluation of an aberrant systemic artery in nearly 80% of cases [15], and MR angiography has shown its diagnostic potential in demonstrating the arterial supply and venous drainage of pulmonary sequestrations [16].

In 2001, von Raemdonck et al. [1] published the results of a retrospective analysis referring to lung sequestration in 28 children and adults. In his case series analysis, the intrathoracic abnormality had already been diagnosed by prenatal ultrasound in 14% of the patients. In 64%, the aberrant artery was identified preoperatively by arteriography or CT scan. In our analysis, similar results were found. Lung sequestration was diagnosed in 8 of 11 patients (72.7%) prior to surgery by imaging techniques with or without angiography, no longer using invasive arteriography. In 3 patients, who had a contrast-enhanced CT scan prior to surgical intervention, the diagnosis could only be confirmed intraoperatively. A pulmonary mass had been detected by CT scan in 2 cases, raising suspicion of bronchial carcinoma or abscess in these patients. They had presented with symptoms that can occur in pulmonary sequestration as well as in cancer or inflammatory processes. This demonstrates the necessity of the delineation of aberrant blood supply in order to confirm the diagnosis of sequestration. However, in a retrospective analysis, no statement about the section thickness of previous CT can be made. This shows that in some cases, the final diagnosis can only be made during diagnostic thoracotomy. As a new method for diagnosing lung sequestration preoperatively, endobronchial ultrasound has been described in 1 of the 11 patients, allowing for visualization of the aberrant vessels. However, so far, there are not sufficient data proving this to be a reliable diagnostic method.

The treatment strategy of lung sequestrations depends on the patients' symptoms. Surgery is indicated in patients reporting discomfort, recurrent infections or hemoptysis, but remains controversial in asymptomatic cases. Some authors prefer the watch-and-wait approach, whereas other authors consider prophylactic resection of lung sequestration appropriate to avoid complications [3, 17]. In this analysis, surgery was indicated in 9 out of 11 patients with complaints, and only 2 of the patients underwent surgery despite being free of symptoms. The surgical treatment of choice is lobectomy or segmentectomy. In our analysis, 7 patients (63.6%) underwent lobectomy. With great care and attention, all aberrant vessels have to be identified and ligated, especially those with infradiaphragmatic origin. Intraoperatively, the abnormal blood supply is often more complex than expected by imaging techniques prior to surgery. Missing such arterial vessels will result in severe bleeding complications. In individual cases, an embolization of the aberrant artery can be performed preopera-

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tively to facilitate the subsequent surgical resection [18]. Early diagnosis and treatment of lung sequestrations is important, because lung sequestrations persisting for a prolonged time may result in recurrent pulmonary infections complicated by superimposed infections such as tuberculosis [19], nontuberculous mycobacterial infections [20] or fungal infections [22]. Another severe problem, as shown in this case series analysis, can be hemoptysis, requiring immediate treatment. Heart failure due to an arteriovenous shunt [23–25] can be observed in the case of persisting lung sequestration. A further complication can be the formation of aneurysms within the aberrant vessels associated with a risk of rupture. Furthermore, cases of malignant tumors arising in intralobar lung sequestrations have been described [26].

Conclusion

This case series analysis demonstrates that lung sequestration in advanced age is rare but can be accompanied by severe pulmonary symptoms. In cases of recurrent pulmonary infections of identical localization or recurrent hemoptysis, lung sequestration should be considered in adults so that a diagnosis by imaging methods can be confirmed rapidly.

Financial Disclosure and Conflicts of Interest

All authors certify that there is no actual or potential conflict of interest.

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