Pulmonary Sequestrations: Issues Related to Diagnosis, Possible Complications, and Comorbidities

Pulmoner Sekestrasyonlar: Tanısal Zorluklar, Olası Komplikasyonlar ve Komorbiditeler

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Özet

Amaç: Bronkopulmoner sekestrasyonlar nadir görülen konjenital anomalilerdir. Asemptomatik kalabilecekleri gibi intrapulmoner hematom, hemotoraks ve aspergilloma gibi komplikasyonlar gelişebilir, veya karsinoid tümör, adenokarsinoma, skuamoz hücreli karsinoma, sarkom veya pulmoner blastom gibi malign transformasyonlar gösterebilirler. Bronkopulmoner sekestrasyonlar, diğer konjenital anomaliler ile de beraberlik gösterebilirler. Çalışmamızın amacı sekestrasyonlarda tanı konulması sırasında karşılaşılan zorluklar, eşlik eden konjenital anomaliler ve gelişebilecek komplikasyonların vurgulanmasıdır. Gereç ve Yöntem: 2003-2015 yılları arasında kliniğimizde opere edilen 19 bronkopulmoner sekestrasyonlu olgunun bilgileri retrospektif olarak değerlendirilmiştir. Bulgular: Lezyonlar 16 hastada sol tarafta, 3 hastada sağ tarafta yerleşmekteydi ve 15 hastada intralober iken 4 hastada ekstraloberdi. Başvuru sırasında bir hastada ampiyem ve pnömotoraks, bir hastada hemoptizi, iki hastada plevral efüzyon ve iki hastada tekrarlayan pnömoni şikayetleri mevuttu. İntralober sekestrasyonlu bir olguda eş zamanlı olarak orta kranial fossada 4 cm çaplı araknoid kist tespit edildi. Ekstralober sekestrasyonlu bir olguda postoperatif histopatolojik incelemede multipl tümörlet alanları ve karsinoid tümör bulundu. Postoperatif bir hastada şilotoraks ve bir hastada transuda plevral efüzyon gelişti. Ortalama hastanede kalış süresi 7,9 (3-26) gündü ve mortalite gelişmedi. Tartışma: Pulmoner sekestrasyon tespit edilen hastalarda gelişebilecek komplikasyonlar ve malign transformasyonlar nedeniyle alınacak olan cerrahi kararı geciktirilmemelidir ve eşlik eden konjenital anomaliler açısından dikkatli olunmalıdır.

Anahtar Kelimeler

Sekestrasyon; Cerrahi Tedavi; Konjenital Anomaliler; Malign Transformasyon; Karsinoid Tumor

Abstract

Aim: Bronchopulmonary sequestrations are a rare congenital anomaly. Although they may remain asymptomatic, complications such as intrapulmonary hematoma, hemothorax, and aspergilloma may occur, or they may undergo malignant transformation to become a carcinoid tumor, adenocarcinoma, squamous cell carcinoma, sarcoma, or pulmonary blastoma. Other congenital anomalies may also be associated with bronchopulmonary sequestrations. Our study aimed to emphasize the difficulties in diagnosis, accompanying congenital anomalies, and possible complications. Material and Method: We retrospectively evaluated data obtained from 19 patients with bronchopulmonary sequestration who underwent surgery in our clinic between 2003 and 2015. Results: Lesions were located on the left side in 16 cases and on the right side in three cases, and 15 cases were intralobar, while four were extralobar. One patient had empyema and pneumothorax, one patient had hemoptysis, two patients had pleural effusion, and two had recurrent pneumonia during admission. A patient with intralobar sequestration had a coexistent 4 cm diameter arachnoid cyst in the right middle cranial fossa. Multiple tumorlet areas and carcinoid tumors were postoperatively determined in an extralobar sequestration case. Postoperatively, one patient had chylothorax and one had transudate pleural effusion. The average length of stay in the hospital was 7.9 (3-26) days, and there was no mortality. Discussion: The possibility of complications and the risk of malignant transformation in patients with bronchopulmonary sequestration mean that the decision regarding surgical resection should not be delayed, and attention should be paid to concomitant congenital abnormalities.

Keywords

Sequestration; Surgical Treatment; Congenital Abnormalities; Malignant Transformation; Carcinoid Tumor

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Introduction

A bronchopulmonary sequestration (BPS) is a portion of lung tissue that lacks a normal connection with the tracheobronchial tree and has an abnormal vascular supply. Although aberrant blood supply of the lung was first reported by Huber in 1777, the term "sequestration" was described by Price in 1946 [1]. It is a rare congenital malformation with an estimated incidence of 0.15% to 6.45% [2], and is divided into two types, intralobar sequestration (ILS) and extralobar sequestration (ELS). An ELS is enclosed within its own pleural membrane, whereas an ILS shares the pleural membrane of the normal lung.

Some previous studies have shown that BPS can be associated with other congenital anomalies. Common anomalies that may be concomitant with BPS are oesophagobronchial diverticulum, diaphragmatic hernia, deformities of the skeletal system, heart and great vessel anomalies, and lung, renal, and cerebral anomalies [3].

Although BPS may remain silent, complications such as intrapulmonary hematoma, hemothorax, and aspergilloma may occur, or they may undergo malignant transformation [4, 5].

In most cases of ILS, venous drainage occurs via the pulmonary vein to the left atrium, although connections to the vena cava, azygous vein, or the right atrium may also be found. In ELS, venous drainage takes place via the systemic circulation through the azygous or hemiazygous vein or the vena cava [6].

Material and Method

Between January 2003 and August 2015, 19 patients (eleven males, eight females) with a mean age of 35.1 (2-65) years (Table I) underwent lobectomy or segmentectomy for BPS. This series of cases was retrospectively analyzed according to age, gender, clinical features, diagnostic procedures, lesion localizations, arterial supply, and surgical findings and techniques, associated anomalies, and postoperative complications were evaluated. All patients were preoperatively evaluated by chest computed tomography (CT) and underwent fiberoptic bronchoscopic examination. Thoracotomy was performed for 18 patients and one patient underwent video-assisted thoracoscopic surgery (VATS). Sequestration lobe excision was performed in all ELS cases. Of the ILS cases, 13 underwent lobectomy and one underwent segmentectomy.

Table I. The distribution of bronchopulmonary sequestrations.

	ILS	ELS	Total
Gender			
Male	9	1	10
Female	5	4	9
Location			
Right	1	2	3
Left	13	3	16

ILE = intralobar pulmonary sequestration; ELS = extralobar pulmonary sequestration.

Results

Of the 19 BPS cases reviewed, 26.3% of the patients were asymptomatic, and the most common complaints were cough (30.8%), chest pain (15.4%), dyspnea (11.5%), fever (11.5%), sputum expectoration (11.5%), and hemoptysis (3.8). Fifteen cases were misdiagnosed, due to complaints and radiological findings.

A 53-year-old male with ILS had headache; a coexistent congenital arachnoid cyst of 4 cm in diameter was detected in the right middle cranial fossa via cranial CT. Left lower lobe superior segmentectomy was performed without any postoperative complications.

Multiple tumorlet areas, neuroendocrine cell hyperplasia, and a 4 mm typical carcinoid tumor were found in the case of a 52-year-old female who had left lower ILS and who underwent a left lower lobectomy (Fig.1).



Figure 1: Surgical images of a 52-year-old woman with intralobar bronchopulmonary sequestration.

A 61-year-old female was misdiagnosed for two years as having asthma, due to dyspnea and wheezing. She was evaluated with CT, as she had had no improvement in her condition despite using bronchodilators; ELS was found. Symptoms were reduced following resection (Fig.2).

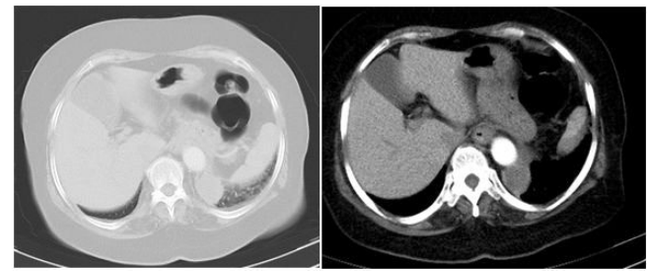


Figure 2: Thorax BT images of a 61-year-old woman with extralobar bronchopulmonary sequestration.

A 15-year-old male had had a cough for two years, and hemoptysis occurred in the two weeks prior to admission. The bronchoscopic examination was normal, and a left lower lobectomy was performed. Postoperative pleural serous effusion occurred and was treated with thoracentesis.

Two female pediatric patients, aged two and five years old, had recurring pneumonia. Radiologic examination showed ILS in the two-year-old and ELS in the five-year-old. The patient with ILS underwent segmentectomy via thoracotomy and was discharged on the 7th day without any complications. The child with ELS underwent a sequestration lobectomy via thoracotomy. She had postoperative chylothorax and required redo thoracotomy for ductus ligation. She was discharged on the 26th day.

A 16-year-old male patient with ILS, in whom left lower lobe consolidation was found in a chest CT, was followed up. In the second month, the size of the consolidation area and the subcarinal lymph node were increased, and pleural effusion was added to the CT findings. Transthoracic needle biopsy was per-

formed, and pneumothorax occurred. A left lower lobectomy was then performed for this patient, and he was discharged seven days postoperatively without any complications.

A 54-year-old male who had had chronic obstructive pulmonary disease for 12 years was admitted to the hospital with serious dyspnea, fever, and a cough. Pneumothorax and empyema were determined by chest X-ray and a tube thoracostomy was performed. After empyema treatment, the patient needed pulmonary rehabilitation due to decreased pulmonary function tests, after which he underwent resection. Left lower lobectomy was performed without any complications.

A 17-year-old female patient with ELS who had pleural effusion in the same hemithorax was referred to our clinic. Left sequestration lobectomy was performed without any postoperative complications.

Chest radiography was used for all patients and all the patients were preoperatively evaluated with CT. The CT findings revealed bronchiectasis in eight cases, two of which showed air-fluid levels.Patchy infiltrations with perilesional emphysematous changes were observed in seven cases, and a solid mass lesion was found in four cases. PET CT was also used in two of the latter cases, while CT angiography was used in one case (Fig.3), and MR angiography was used in the remaining case. Fiberoptic bronchoscopy showed normal findings in all patients. A transthoracic biopsy was performed in three cases and pneumothorax occurred in one patient. Pleural effusion was determined in three cases, one of which was complicated by empyema and spontaneous pneumothorax.

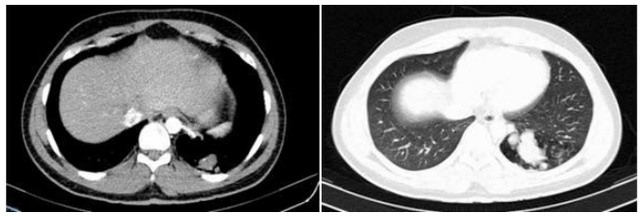


Figure 3. BT angiography shows arterial supply from the abdominal aorta in a 26-year-old man with intralobar sequestration.

Lesions were located on the left side in 16 cases (84.2%); only three lesions were located on the right side (15.8%) (TableI). ILS was detected in 15 patients (78.8%) and ELS was found in four cases (21.1%).

In the ILS cases, left thoracotomy and lower lobectomy was performed in 10 patients, VATS and left lower lobectomy was performed in one patient, right thoracotomy and lower lobectomy was performed in two patients, left lower lobe superior segmentectomy was performed in one patient, and left upper lobe posterior segmentectomy was performed in the remaining patient. In the ELS cases, left thoracotomy and lobectomy was performed in three of the patients and right thoracotomy and lobectomy was carried out in the remaining patient (TableII). The average length of stay in hospital was 7.9 (3-26) days.

Postoperative complications were observed in two patients. Pleural effusion occurred in one patient with left lower ILS. A five-year-old female patient had postoperative chylothorax and was followed up with tube thoracostomy for 16 days. Right redothoracotomy and ductus ligation were required due to continuing chylous drainage.

Table II. Surgical procedures used for bronchopulmonary sequestrations

Incision	Procedure	ILS	ELS
Left thoracotomy	Lobectomy	10	-
	Sequestration lobe excision	-	3
	Segmentectomy	1	-
Right thoracotomy	Lobectomy	3	-
	Sequestration lobe excision	-	1
	Wedge Resection	-	1

ILE = intralobar pulmonary sequestration; ELS = extralobar pulmonary sequestration.

Discussion

BPSs are uncommon congenital malformations of the lung and can be classified as intralobar and extralobar. They consist of nonfunctioning lung tissue that lacks normal communication with the tracheobronchial tree, and they receive arterial blood supply from the systemic vascular structures. Congenital malformations of the lower respiratory system are rare, and BPS constitutes 0.15 to 6.4% of these [1]; various publications have revealed that even tertiary care referral centers diagnose fewer than one case per year [7, 8].

BPS is anatomically classified in two types. The first of these is ILS, which lacks its own visceral pleura, and is located within a normal pulmonary lobe. The second type is ELS, in which the lesion has its own pleura and is located outside the normal lung parenchyma. BPS frequently has aberrant connections to bronchi, lung parenchyma, or the gastrointestinal tract, and recurrent infections are more often observed in ILS than in ELS cases.

ILS constitutes 75 to 90% of BPS [1, 9], and the ELS/ ILS ratio is higher in children than in adults. Wei found that BPS is mainly located in the lower lobes, especially in the left lower lobe (71.53%) [10]. Almost 90% of ELS occur in the left hemithorax, and 10% of them may present below the diaphragm. Bilateral sequestrations are uncommon, and only three cases had previously been reported; two of these were ILS and one was mixed ILS and ELS [10, 11]. In the present study, 78.9% of cases were ILS and 84.2% of our cases were located in the left lower lobe.

ILS is often diagnosed in the first and second decades of life, especially after the age of two years [12]. Other congenital malformations can be particularly identified with ELS, such as diaphragmatic hernia, pulmonary hypoplasia, vertebral anomalies, congenital heart diseases, and colonic duplication [6, 7]. Most of our patients had been diagnosed in their fourth decade of life. In one patient, an intracranial congenital arachnoid cyst was found during preoperative evaluation and was followed up by neurosurgeons.

BPS is associated with various symptoms. However, most ELS cases are asymptomatic and are incidentally found on routine radiographic controls. They may be misdiagnosed as lung cancer, pulmonary cysts, or mediastinal tumors. In symptomatic cases, especially patients with ILS, the symptoms mimic pneumonia (cough, purulent sputum, fever, etc.) [13]. ILS usually presents with signs of recurrent pneumonia in adolescence or adulthood. Otherwise, ELS presents with respiratory distress or feeding difficulties in early life and is frequently associated with other congenital abnormalities [14]. In our series, 73.7% of the patients were symptomatic, and the most common symptoms

were cough, chest pain, and dyspnea.

The most useful diagnostic methods for BPS are CT angiography (CTA), magnetic resonance angiography (MRA), and digital subtraction angiography (DSA), which may show the aberrant arterial supply [6, 7]. We performed CTA for one patient and MRA for another. PET CT was used for two patients for differential diagnosis of solid masses and low FDG uptakes were observed (1.79 and 2.54).

For prenatal diagnosis, ultrasonography (USG) can be used. USG may show a well-defined echodense homogeneous mass in ELS cases [14]. In color flow doppler USG, a systemic artery from the aorta to the fetal lung is pathognomonic. Prenatal magnetic resonance imaging (MRI) may help to distinguish BPS from other lesions (congenital pulmonary airway malformation, congenital diaphragmatic hernia, or other less common lesions). In a study of pregnant women, Adzick found that the majority of lesions were detected in routine second-trimester screening (between 18 and 36 weeks' gestation). Although in utero regression was demonstrated, postpartum imaging is still required [15].

On chest radiographs, sequestration typically appears as a uniform mass within the thoracic cavity or pulmonary parenchyma. Recurrent infection can lead to the development of cystic areas within the mass [14, 16]. Air-fluid levels due to bronchial communication have been observed in 26% of ILS cases [17].

The parenchymal abnormalities associated with BPS are best visualized using CT, although their appearance is variable. The most common finding is a solid mass. The lesion may be homogeneous or heterogeneous and may sometimes include cystic changes. Large cavitary lesions with an air-fluid level, a collection of many small cystic lesions containing air or fluid, or a well-defined cystic mass are rare findings. Peri-lesional emphysematous changes are characteristic and chest radiograph may not show this finding. In the present study, patchy infiltrations and bronchiectasis were observed in eight cases, two of which showed air-fluid levels; patchy infiltrations with perilesional emphysematous changes were found in seven cases; and a solid mass lesion was found in four cases via CT. PET CT was used in two patients with solid lesions and MR angiography was used in one patient. All the patients with solid lesions underwent transthoracic biopsy (two ILS patients and one ELS patient).

MRI can define the aberrant artery and venous drainage, especially if enhanced three-dimensional MR angiography is used. However, CT allows for sharper delineation of thin-walled cysts and emphysematous changes than does MRI [17].

ILS and ELS have different pathologic characteristics. In histopathologic examination of ILS, regions of inflammation, microcystic changes, and mucus accumulation are more often observed than in ELS. ELS may appear as normal parenchyma or show parenchymal abnormalities as congenital pulmonary airway malformation.

Some studies showed that arterial embolization is an effective and safe treatment for BPS and may be preferred for patients with a shunt [18]. Surgical resection is the recommended treatment approach for BPS, especially for patients with infection. ILS usually requires lobectomy or segmentectomy for complete resection, while excision of ELS is simpler because it has own isolated pleura. Thoracoscopic (VATS) resection is a safe and feasible option for PBS resection [19], and vascular structures should be identified and ligated carefully in all patients. We performed left lower lobectomy via VATS for an 18-year-old male patient who was subsequently discharged on the third postoperative day without any complications.

One of the most important aspects of resection is careful identification of vascular structures, because arterial supply of BPS may arise from the subdiaphragmatic aorta. In our series, 31.6% of patients had aberrant arterial supply from the abdominal aorta.

BPS-related associations and complications that have been reported are: bleeding into the tissue, pulmonary infarction, hemothorax, hemoptysis, pneumothorax, abscess, aspergilloma, hemangioma, and inflammatory myofibroblastic tumor [5, 20, 21]. In our series, pleural effusion was determined in three cases, one of which was complicated by empyema and spontaneous pneumothorax. A patient with ILS in the left lower lobe was admitted with severe hemoptysis.

Congenital anomalies such as oesophagobronchial diverticulum, diaphragmatic hernia, deformities of the skeletal system, heart and great vessel anomalies, and lung, renal, and cerebral anomalies can be associated with BPS [3]. Congenital arachnoid cyst was determined in one of our patients who had headache and the suspicion of malignancy preoperatively.

Some case reports about BPS showed that malignancy may occur with BPS synchronously. They were carcinoid tumors, tumorlets, adenocarcinoma, squamous cell carcinoma, sarcoma and pulmonary blastoma [22- 26]. This is important because patients with BPS are frequently treated for benign diseases such as a common cold or pneumonia. If the patient is asymptomatic, the condition may not warrant intervention and may persist for several years. However, malignant disorders without treatment can worsen a patient's condition and spread to other organs. In our study, typical carcinoid tumor and tumorlets were detected in one case after surgery. The tumor was 4 mm in diameter, and had not been found preoperatively.

In conclusion, following a diagnosis of BPS, surgical resection should not be delayed so that complications and malignant transformations may be avoided. Careful preoperative evaluation will decrease perioperative complications due to vascular injuries and accompanying anomalies.

Competing interests

The authors declare that they have no competing interests.

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