

Case Series



Computed tomography findings of atypical intralobar pulmonary sequestration: A case series

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Abstract

Introduction: Bronchopulmonary sequestration is a rare congenital abnormality of the lung that has two different types as intralobar and extralobar. In this study, we aimed to present six cases of intralobar sequestration with atypical findings in terms of feeding, drainage, and localization.

Methods: Patients diagnosed with intralobar pulmonary sequestration in our clinic between 2015 and 2019 were evaluated retrospectively. Demographical features and atypical computed tomography (CT) findings of the patients were presented by literature.

Results: Among 45 patients with intralobar sequestration, 6 (13.3%) (5 males and 1 female) with a mean age of 43.5±25.4(0-78) years old had atypical pulmonary findings on CT images. Atypical features regarding arterial supply was detected in 8.9%, venous drainage in 2.2%, location in 4.4%, radiological appearance in 4.4% and co-existing lesion in 2.2% of the patient with intralobar sequestration.

Conclusion: Typical and atypical features of pulmonary sequestration must be well-known for differential diagnosis of solid or cystic pulmonary lesions.

Keywords: Pulmonary sequestration, Intralobar, Computed tomography, Atypical findings

Introduction

Pulmonary sequestration is a congenital dysplastic lung tissue, which is not associated with the tracheobronchial tree and pulmonary arteries. Arterial supply usually occurs through the thoracic aorta. Pulmonary sequestration has two different types. Intralobar pulmonary sequestration (75%-90%) is located in the lung tissue and does not have its own visceral pleura. It is usually seen as a heterogeneous solid or cystic mass.¹⁻³ Left lower lobe of the lung is the most frequent location of intralobar pulmonary sequestration and it is supplied from thoracic aorta.⁴ Extralobar pulmonary sequestration (10%-25%) completely separates from normal lung tissue and has its own pleura.^{4,5}

The typical features of intralobar pulmonary sequestration include arterial supply from the thoracic aorta by solitary branch, venous drainage via pulmonary veins, left lower lobe involvement, and heterogeneous solid mass appearance. The lesion may be seen as a cystic mass including air when it is infected.^{2,6}

In this study, we aimed to present atypical computed tomography (CT) findings of intralobar sequestration by reviewing each feature of all our patients.

Methods

In this retrospective research study, patients with a histopathological proven diagnosis of intralobar pulmonary sequestration who had also a presurgery contrast-enhanced CT scan in our clinic between 2015 and 2019 were evaluated based on the hospital database and radiology department records. Demographic and medical data of each patient were recorded. CT images were obtained by a multislice CT device with 16 detectors (Somatom Emotion 16-slice; CT2012E-Siemens AG Berlin and Munich, Germany). Patients were injected with 1-1.5 cc/kg of non-ionized intravenous contrast agent iohexol (Amersham Health, Ireland) or iopromide (Schering, Germany) at a rate of 3 mL/s via an automated syringe (CT 9000 ADV Liebel-Flarsheim). In thorax CT examination, images were obtained from the distal neck to the upper abdomen at the 25th second after contrast agent administration. The examination was performed by holding the patient's breath. Axial and multiplanar reformatted images with 3 mm slice thickness were evaluated by two radiologists with 10 to 12 years of experience on thorax radiology. All consensus was reached again by these two radiologists. Images were interpreted for atypical features of sequestration tissue including location,

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arterial feeding, venous drainage, radiological appearance, and accompanying lesions. The variables were expressed as number and mean \pm standard deviation.

Results

An intralobar pulmonary sequestration was detected in overall 45 patients. All these patients had the diagnosis of intrapulmonary sequestration by histopathological examination after thoracotomy with resection of the lesions. Among these 45 patients, 6 had atypical findings regarding location, arterial supply, venous drainage, radiological appearance, and co-existing lesion. The mean age of the 6 patients (13.3%) with atypical pulmonary sequestration was 43.5 ± 25.4 (0-78) and the male/female ratio was 5 (5/1).

Case 1

The first case was a 53-year-old male who presented with cough and shortness of breath. The lesion was located in the middle lobe of the right lung and arterial feeding was provided from the abdominal aorta (Figure 1). The venous drainage was via the pulmonary vein. The lesion had an infected cystic appearance with fluid-filled bronchial structures and consolidated areas. The atypical findings of this case were the location, arterial supply, and also infected cystic appearance.

Case 2

The second case was a 38-year-old male who had night sweat and wheezing. The lesion was fed by two arterial branches originated from the thoracic aorta and drained to the azygos vein (Figure 2). The lesion had a heterogeneous solid mass appearance. The atypical findings of this case were venous drainage and two arterial branches arising from the thoracic aorta.

Case 3

This case was a 49-year-old asymptomatic female. The arterial feeding was provided by truncus celiacus and venous drainage was via the pulmonary vein (Figure 3). The lesion had a heterogeneous solid mass appearance. The atypical finding in this case was arterial supply.

Case 4

The fourth case was a 43-year-old male who had complaints of cough and shortness of breath. The lesion with a heterogeneous solid mass appearance was located in the lower lobe of the left lung, fed from the thoracic aorta and drained to the pulmonary vein. It was associated with a neighboring bronchogenic cyst (Figure 4). The atypical finding in this case was an adjacent bronchogenic cyst.

Case 5

The fifth case was an asymptomatic newborn female in whom the sequestration was located in the lower lobe of

the right lung and the arterial feeding was from the truncus celiacus (Figure 5). The lesion had a typical heterogeneous mass appearance with venous drainage via the pulmonary vein. The atypical finding in this case was arterial supply origin.

Case 6

The last case was a 75-year-old male who presented

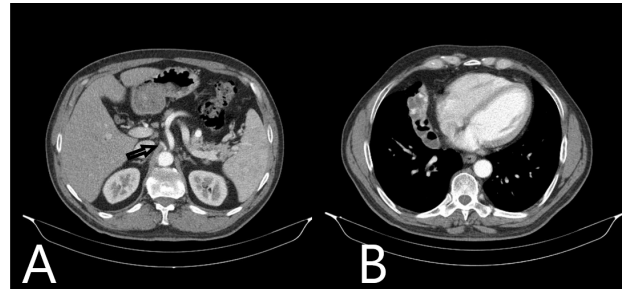


Figure 1. A thin artery arising from abdominal aorta medial to truncus celiacus (A) feeding a heterogenous sequestration tissue with air-fluid levels (B) in middle lobe of right lung.

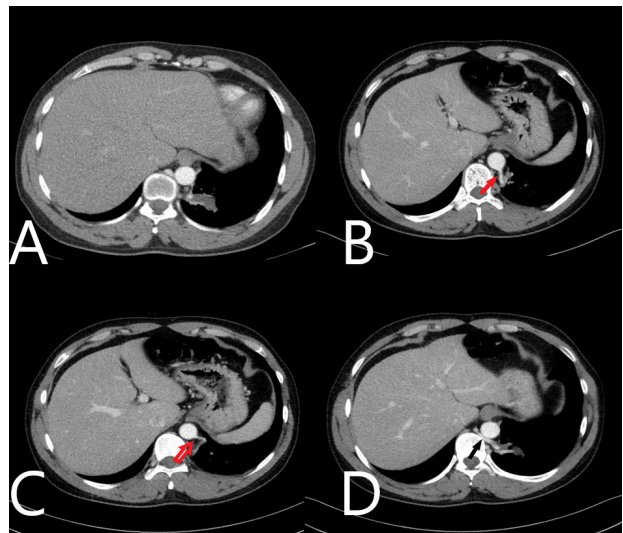


Figure 2. A sequestration in lower lobe of left lung (A) which has two arteries from abdominal aorta (B, C) (red arrows) and draining to azygos vein (D) (black arrow).

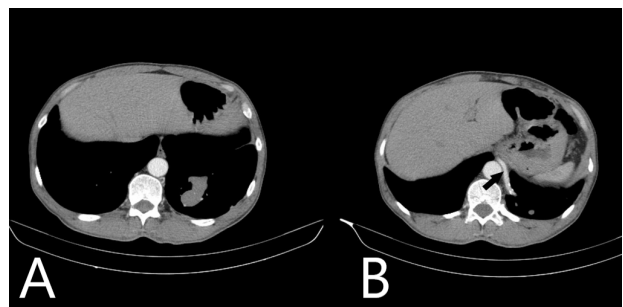


Figure 3. A sequestration in lower lobe of left lung (A) that was fed by a branch originated from truncus celiacus (B) (black arrow).

with fever and cough. The sequestration tissue with a heterogeneous infected cystic appearance was located in the lower lobe of the left lung which was fed from the thoracic aorta and drained to the pulmonary vein. The atypical findings in this case was an infected cystic appearance which could be confused with the pneumonic infiltration (Figure 6).

Features of the patients are summarized on Table 1.

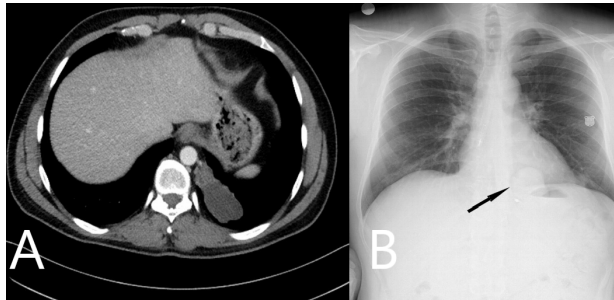


Figure 4. A sequestration in lower lobe of left lung (A) and adjacent a bronchogenic cyst (Arrow) located posterolateral to the sequestration tissue (A, B)

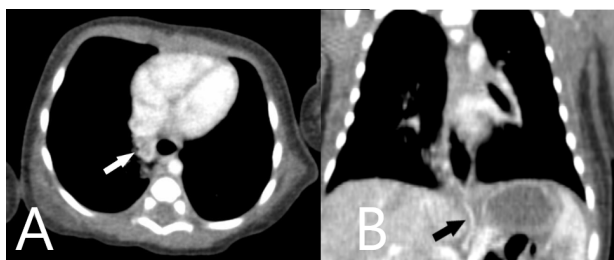


Figure 5. A sequestration tissue in lower lobe of right lung (A) that has arterial supply from truncus celiacus (B) (black arrow)

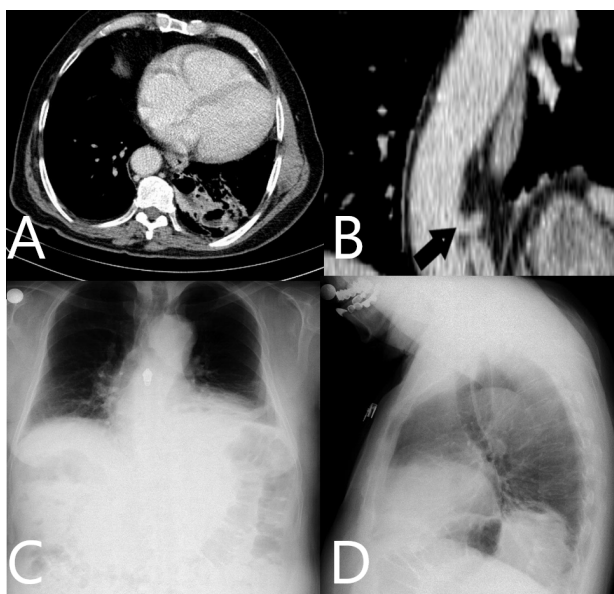


Figure 6. The sequestration tissue in lower lobe of left lung (A) that had arterial supply from thoracic aorta (B) with air-fluid levels. The lesion is located in posterolateral of left lung (C-D)

Complaints of the patients

Three of our 6 patients with atypical intralobar pulmonary sequestration, had cough and dyspnea and one of them had night sweating. Cough and dyspnea was seen in half of the patients (50%) while sweating in 16.7%. Two of our patients (33.3%) were asymptomatic.

Radiological algorithm

In two of our cases, the CT scans were performed after detecting lesions on PA chest radiography (Figures 4B, 6C, and 6B) In the newborn patient, CT was performed because of the suspicion of sequestration detected on antenatal ultrasound. The remaining three cases were found incidentally on initial CT scans.

The diagnosis of sequestration

Intralobar pulmonary sequestration was diagnosed clinically and radiologically in all of the cases and all of them were confirmed by surgery and histopathological examinations.

Location of lesions

Sequestration tissue was observed in the lower lobe of the left lung in 43 (95.6%) of overall 45 patients, in the middle lobe of the right lung in one case (case 1) (2.2%) and in the lower lobe of right lung in the remained one (case 5) (2.2%). Lower lobes involvement was detected in 97.8% (44/45) and left lung involvement in 95.6% (43/45). No bilateral involvement was seen.

Arterial supply

A total of 4 (8.9%) atypical arterial supply was observed in the patients. In two (4.4%) patients (cases 3 and 5), the artery of sequestration tissue arose from celiac truncus and in one (2.2%) patient (case 1) from the abdominal aorta. In one patient (2.2%) (case 2), two nourishing arteries originating from thoracic aorta were observed. One artery arising from thoracic aorta was seen in remained 41 patients (91.1%).

Venous drainage

Among 45 patients, in only one (2.2%) (case 2) the sequestration tissue drained to azygos vein while in remained 44 ones (97.8%) drained to pulmonary veins.

Radiological appearance

In two patients (cases 1 and 6) (4.4%), the lesions had a cystic dominant appearance which was due to a probable infectious process while in remained 43 ones (95.6%), typical heterogeneous solid mass appearance was observed.

Co-existing lesions

In one patient (2.2%) (Case 4), a bronchogenic cyst adjacent to sequestration tissue was noted since in the other 44 (97.8%) patients no co-existing congenital lesion

Table 1. The radiological features of the patients

	Age of diagnosis	Clinical complaint	Localization	Arterial origin	Venous drainage	Computed tomography findings
Case 1	53	Cough and shortness of breath	Middle lobe of the right lung	Abdominal aorta	Pulmonary vein	Infected sequestration tissue with fluid-filled bronchial structures and consolidated areas (Figure 1A and 1B)
Case 2	38	Night sweats and wheezing	Lower lobe of the left lung	Two separate branches from the thoracic aorta	Azygos vein	Heterogeneous, solid appearance sequestration tissue (Figure 2A -2D)
Case 3	49	Asymptomatic	Lower lobe of the left lung	Truncus celiacus	Pulmonary vein	Heterogeneous, solid appearance sequestration tissue (Figure 3A and 3B)
Case 4	43	Cough and shortness of breath	Lower lobe of the left lung	Thoracic aorta	Pulmonary vein	Heterogeneous, solid appearance sequestration tissue and coexistence of bronchogenic cyst in the neighborhood of sequestration. ((Figure 4A and 4B)
Case 5	Newborn	Asymptomatic	Lower lobe of the right lung	Truncus celiacus	Pulmonary vein	Heterogeneous, solid appearance sequestration tissue (Figure 5A and 5B)
Case 6	78	Cough and fever	Lower lobe of the left lung	Thoracic aorta	Pulmonary vein	Infected sequestration tissue with cystic areas, fluid values and solid appearances (Figure 6A -6D)

within lung parenchyma was seen.

Discussion

Pulmonary sequestration is a congenital abnormality involving dysplastic lung tissue that does not have a relation with the tracheobronchial tree and pulmonary arteries. It is a rare congenital abnormality which accounts for 0.15%-6.4% of all congenital lung malformations.⁷ Diagnosis is often difficult because of the various clinical presentations described. It was first described by Pryce in 1946.⁸ Two different types of pulmonary sequestration have been described as intralobar and extralobar sequestration. Intralobar pulmonary sequestration is observed in normal lung tissue and it does not have its own pleura. Extralobar pulmonary sequestration is completely separated from normal lung tissue and it has its own pleura. Approximately 75% of all sequestration cases are intralobar pulmonary sequestration. Nearly 98% of intralobar pulmonary sequestration cases are located in the lower lobes and generally located in the left lower lobe.⁴ In this study, we did not include the patients with extralobar pulmonary sequestration. Of all 45 patients, 44 (97.8%) had an intralobar sequestration in lower lobes similar to the literature. Also, the left lobe involvement (95.6%) was very significant in our study.

Approximately 50%-60% of intralobar pulmonary sequestrations are detected in younger patients in their second decade and rarely diagnosed in the patient older than 50 years old.⁹⁻¹¹ One of our cases was a newborn who was diagnosed after surgery in the neonatal period. Others were diagnosed at the age of 53,38,49,43 and 75 years, respectively. As consistent with the literature, the mean age of our patients with atypical pulmonary sequestration was higher than of those with pulmonary sequestration reported previously in the literature. We think that this difference may be due to the atypical findings which are

not well-known.

Several imaging modalities are used for the diagnosis of bronchopulmonary sequestration.² Chest X-ray is an important diagnostic imaging modality.¹² Bronchopulmonary sequestration is seen as a well-defined homogenous opacity or solitary lung nodule on the chest radiographs.^{4,13} In our two cases, sequestration tissue was first detected as parenchymal opacities on direct X-ray (Figures 4B, 6C and 6D).

Traditionally, the definitive diagnosis of pulmonary sequestration is made by demonstrating the aberrant systemic artery in an invasive arteriographic examination.^{4,13} Today, with the development of CT technology, three-dimensional images can be obtained and diagnosis can be made easily with contrast-enhanced CT. Therefore, CT is the best noninvasive imaging method for the detection of parenchymal abnormalities.¹³ In all of our cases, the diagnosis was made by contrast-enhanced multislice CT and confirmed by the surgery.

In a study by Savic et al, it was reported that air-fluid levels were seen in 26% of the patients with intralobar sequestration.⁴ In our study, we detected air fluid level in two cases (4.4%). Intralobar pulmonary sequestration is usually seen as a homogeneous or heterogeneous solid mass in the tomographic examination. It may rarely contain cystic changes.^{4,13} Savic et al stated that 13.7% of the patients with intralobar pulmonary sequestration had also co-existing abnormalities.⁴ Their rate was marked higher than our results probably due to the fact that they included not only chest structures but also all system abnormalities including musculoskeletal and nervous systems. In our series, there was a co-existing adjacent (a bronchogenic cyst) lesion in the lung in only one patient (2.2%).

Pulmonary sequestration is also included in the differential diagnosis of 'middle lobe syndrome'. In this

respect, it is important to look at the arterial origin of heterogeneous lesions seen in the middle lobe. In our study, it was possible to confuse sequestration tissue with middle lobe syndrome in case 1, but the preliminary diagnosis was made by demonstrating the arterial supply from the abdominal aorta. For this reason, the detailed imaging of all solid or cystic intrapulmonary lesions is essential.

According to the study performed by Savic et al, the artery of intralobar sequestrations originates from thoracic aorta in 73.9%, and from abdominal aorta in 18.7% of the patients. Truncus celiacus and its branches (splenic and hepatic arteries) may rarely be reported as the origin of arterial supply of intrapulmonary sequestration.⁴ Multiple arterial feeding was reported in 17.4% of the patients with intralobar sequestration.⁴ In our study, arterial feeding occurred from the abdominal aorta in one case (2.2%), and from the celiac artery in two cases (4.4%). In one case, there were two arteries arising from thoracic aorta and entering the sequestration tissue. In our study, multiple feeding artery ratio was 2.2% (n=1). For the other 41 patients (91.1%), the solitary artery of the sequestration tissue arose from the thoracic aorta.

Pulmonary drainage of intralobar sequestration is mostly through the pulmonary veins. In less than 5% of the cases, venous drainage is via systemic circulation through the vena azygos, hemiazygos, intercostal veins and inferior-superior vena cava.¹⁴ In our study, similar to the literature, venous drainage via azygos vein was detected in only one case (2.2%) while the sequestration tissue drained to pulmonary veins in 97.8% (n=44) of the patients.

The primary treatment modality for bronchopulmonary sequestration is surgery, but it carries the risk of infection.^{15,16} Preoperative imaging is important in terms of providing information about location, arterial nutrition, and venous drainage for surgical planning. Transcatheter endovascular embolization has been successfully applied since 1998 as another treatment modality.¹⁷ Transcatheter endovascular embolization treatment is an alternative to surgery in neonates and children as it is a minimally invasive method.¹⁸ Accurate preoperative diagnosis is important as it will change the surgical approach. All of our cases were successfully treated with surgery without any complication or residue.

Although we have evaluated the patients with lobar sequestration over a wide time period, the small number of cases can be considered as a limitation of our study.

Conclusion

Contrast enhanced CT is a very effective method for the diagnosis of pulmonary sequestration which provides important vascular and structural features of the lesions. For an accurate evaluation and differential diagnosis, typical and atypical CT findings of pulmonary sequestration must be well-known. Since pulmonary

sequestration can mimic various lesions in all age groups, as in our patients, the arterial supply of all lesions in especially lower lobes must be carefully evaluated.

Conflict of Interest

Authors declare no conflict of interest in this study.

Ethical Approval

Authors declared that procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008. The study was approved by a university ethics committee at the date of 22/05/2020 with a number of 2020/03-47.

Author's Contribution

SO: Editing, revision, manuscript writing process, interpretation of the data. MO: Review article, manuscript writing process and interpretation of data. MBA, FD, ID,CG: Review article and interpretation of data. FS: Interpretation of data.

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