



## Case Report

## Radiology aspect of intra-lobar pulmonary sequestration, lung cancer-associated, and hybrid lesions: A case report

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## ABSTRACT

**Background:** Pulmonary sequestration is a mass of lung tissue disconnected from the bronchial tree, which derives its blood supply from one or more systemic vessels.

**Case presentation:** 3 participants were diagnosed with intra-lobar pulmonary sequestration (IPS) where the diagnosis was obtained using CT-Scan. The diagnosis is also supported by the results of Thoracic CT-Angiography and other examinations.

**Discussion:** The CT-Scan image of the IPS thorax needs to be considered carefully, especially in low research settings.

**Conclusion:** This report is expected to help in diagnosing IPS which is a rare case so that misdiagnosis can be minimized.

## 1. Introduction

Pulmonary sequestration is a mass of lung tissue disconnected from the bronchial tree, which derives its blood supply from one or more systemic vessels, commonly the thoracic or abdominal aorta. There are two types (intra and extra-lobar sequestration). Intra-lobar sequestration (ILS) is more common (75%) than ELS. ILS is surrounded by normal lung; that is, it shares the pleural investment with the surrounding normal lung, and usually drains into the pulmonary venous system. 60% of the cases are located in the left lower lobe and they relatively frequently become infected. ELS usually has its pleural covering and systemic venous drainage to the azygos or portal venous systems. The ELSs are located at the left base in 77% of cases. ELSs can be located below the diaphragm and mimic neuroblastoma or adrenal hemorrhage. ELS is associated with multiple congenital malformations in 65% of cases, including other types of bronchopulmonary foregut malformation [1,2]. There is considerable overlap between pulmonary sequestration and Congenital Pulmonary Airway Malformation (CPAM) and in some cases, pulmonary sequestration can also be accompanied by lung malignancy. Based on the description above, we are interested in reporting several cases of pulmonary sequestration in Indonesian patients. we report our case using surgical case report (SCARE) 2020 guidelines [3].

## 2. Case presentation

The number of case series in this report is 3 participants. Details of the differences between the three participants can be seen in Table 1.

In case 1, A 58-years old female patient, with the chief complaint is abdominal pain and lump in the upper abdomen, rarely cough, no sputum or bloody cough, there is no shortness of breath and fever, there are no eating and bowel problems. The patient was referred suspected of abdominal mass. At first abdominal ultrasound was performed and the result was hetero-echoic solid mass at the superior of the right liver with size  $\pm 11 \times 11.9$  cm and abdominal organs appear normal, from this finding we suggest performing a chest CT-Scan with contrast administration. From chest CT-Scan, there is heterogen-density mass with necrotic and calcification component with size  $\pm 13.3 \times 10.2 \times 11.4$  cm intra-thoracic intrapulmonary of the right inferior lobe, disconnected from the bronchial tree, with systemic feeding artery (five from thoracic aorta and two from abdominal aorta) and right pleural effusion (Fig. 1). From the multidisciplinary discussion, this case is concluded as intra-lobar pulmonary sequestration (IPS), and surgery was planned for this patient. In preparation, the patient also underwent arteriography and embolization, to evaluate vascularization and embolization to minimize bleeding during surgery.

Case 2, a 38-years old female patient, with a complaint of

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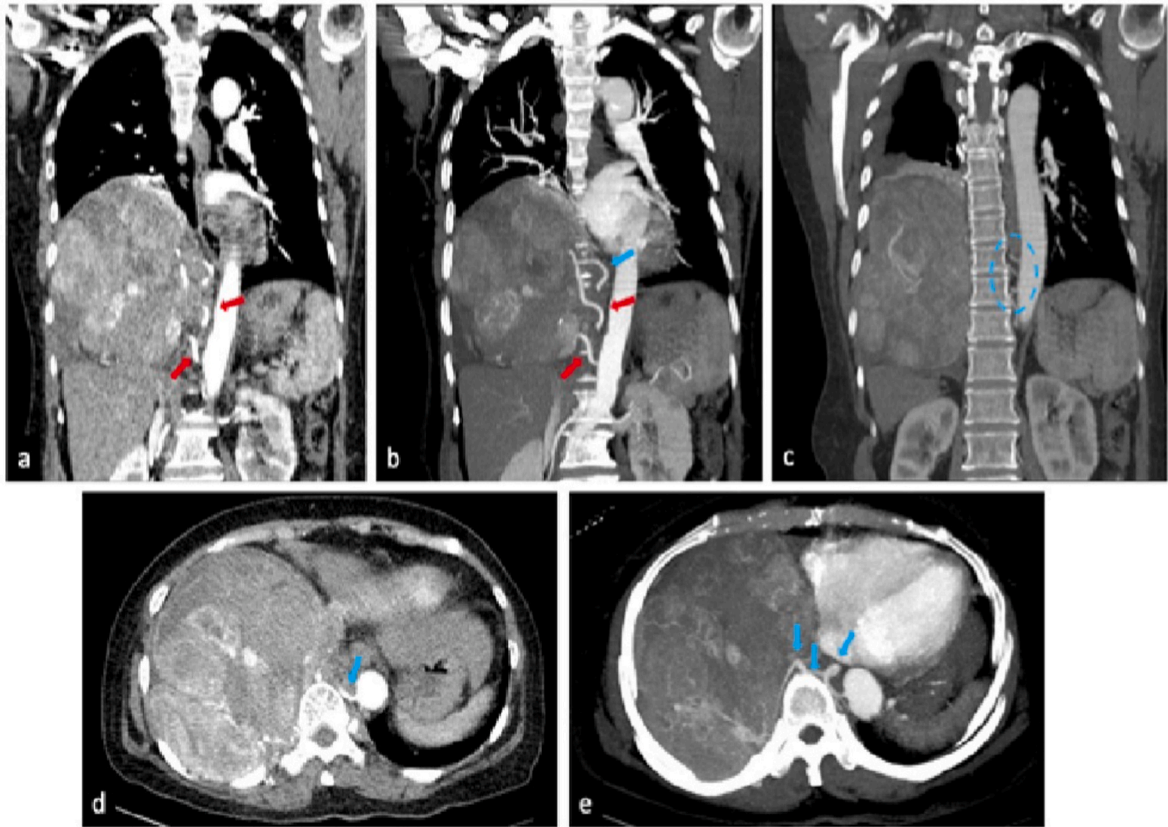
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**Table 1**  
Comparison of each patient intra-lobar pulmonary sequestration.

No	Age	Gender	Diagnosis	Location	Arterial supply	Associated feature and anomalies	Radiology manifestation	Therapy
1	58-year-old	Female	ILS	right inferior lobe	thoracic aorta and abdominal aorta	right pleural effusion	Mass	Embolization
2	38-year-old	Female	ILS and Adenocarcinoma	left inferior lobe	aortic arch and thoracic aorta	blebs and left pleural effusion	Mass	Embolization and oral chemotherapy
3	2 days	Female	ILS and CPAM	right inferior lobe	thoracic aorta	pneumonia, ASD and PDA	Multiple cavities	Supportive therapy

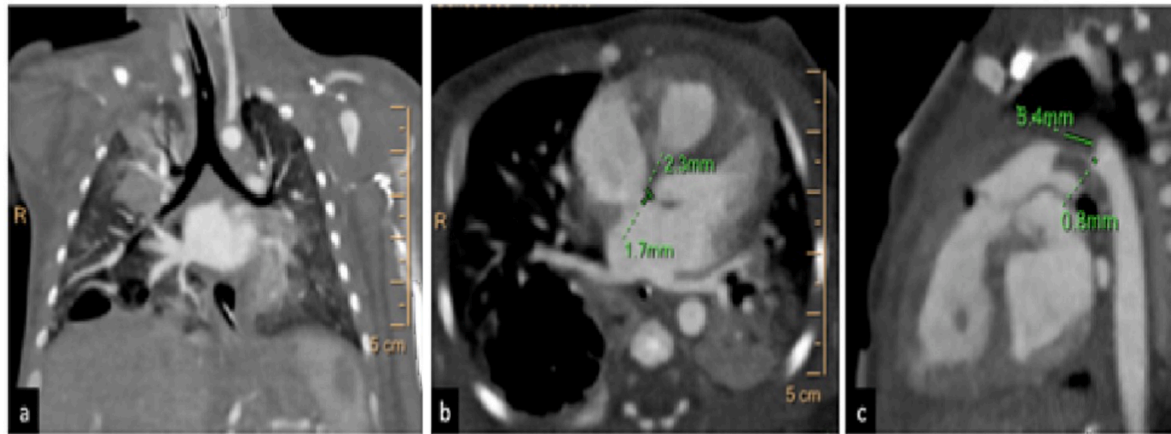
Note: ILS = intra-lobar pulmonary sequestration; CPAM = congenital pulmonary airway malformation; ASD = atrial septal defect; PDA = patent ductus arteriosus.



**Fig. 1.** Chest CT-Scan with contrast administration. (a,b,c) showing the intra-thoracic intrapulmonary mass of right inferior lobe with feeding artery from the aorta; abdominal aorta (red arrow), thoracic aorta (blue arrow, dashed blue circle), (d,e) showing feeding artery from the thoracic aorta (blue arrow).



**Fig. 2.** Chest CT-Scan (a,b) showing the intra-thoracic intrapulmonary mass of left inferior lobe with largest feeding artery from the aortic arc (red arrow), (c) blebs on the apico-posterior segment of the left superior lobe (blue arrow).



**Fig. 3.** Thoracic CT-Angiography, mediastinal window. (a) consolidation with air-bronchogram at the right superior lobe and left inferior lobe, (b) small ASD, (c) small PDA tubular type (Krichenko Angiographic Classification).

hemoptysis, shortness of breath, weight loss of about 10 kg in a month, first Chest CT-Scan was performed, there is heterogen-density mass with necrotic and calcification component with size  $\pm 12.9 \times 11.6 \times 22.3$  cm intra-thoracic intrapulmonary of left inferior lobe, disconnected from the bronchial tree, with multiple feeding arteries from a systemic artery (thoracic aorta at the level 5th – 12th), lymph node enlargement  $\pm 1.5$  cm at right paratracheal, there is also blebs on the apico-posterior segment of left superior lobe and left pleural effusion. This case was also discussed in the multidisciplinary team and concluded as IPS and surgery was planned for this patient. Thoracic CT-Angiography this pulmonary sequestration got 4 feeding arteries from the thoracic aorta (largest feeding at aortic arch) and draining vein to left pulmonary vein. Embolization was performed by an interventional radiologist and only the largest feeding artery is embolized at the aortic arch. Biopsy was performed and histopathology revealed non-small cell carcinoma (adenocarcinoma) within pulmonary sequestration. Based on these findings we staged this mass as stage IIIC (T4N3M0), surgery was canceled and changed the therapy to oral chemotherapy (gefitinib). The last Chest CT-Scan evaluation was performed, size  $\pm 13.7 \times 12.5 \times 23.1$  cm, increased in largest diameter  $\pm 3\%$  and concluded as stable disease and fluid-filled blebs at the apico-posterior segment of the left superior lobe (Fig. 2).

Case 3, a 2-days old premature female newborn (7 months gestation age), born by cesarian section, present with suspected of pneumonia, congenital heart disease (atrial septal defect and patent ductus arteriosus), neonatal jaundice. Thoracic CT-Angiography was performed, there are multiple cavities lesions at the right inferior lobe, with the widest diameter  $\pm 1.3$  cm, these lesions got feeding arteries from a systemic artery (thoracic aorta) and related or connected to an inferior branch of the right bronchus (Fig. 3). There is consolidation with air-bronchogram at the right superior lobe and left inferior lobe, small ASD, and small PDA tubular type (Krichenko Angiographic Classification). This case was also discussed in the multidisciplinary team and concluded as hybrid lesions (CPAM and IPS) accompanied by other abnormalities, surgery cannot be performed on this patient, because her condition is unstable and only provide with supportive therapy. Three weeks later this baby died because of her condition complex condition.

### 3. Discussion

Pulmonary sequestration is the term used to describe a rare embryonic mass of lung tissue that has no bronchial communication with the normal tracheobronchial tree [4]. Pulmonary sequestration can be massive and life-threatening. It is thought to be due to increased capillary pressure secondary to elevated systemic pressures in the feeding arteries [5]. Multidetector computed tomography (MDCT) is the

technique of choice for evaluating pulmonary sequestrations as its higher spatial and temporal resolution is more sensitive in detecting small systemic vessels than MRI, especially in small children. CT depicts the feeding artery and the venous drainage also allows simultaneous evaluation of the lung parenchyma. MRI demonstrates a solid, well-defined, and hyperintense mass on T<sub>2</sub> weighted images, with a systemic feeding artery [6].

Sequestrations have been surgically resected to prevent recurrent pulmonary infections and to protect the normal surrounding lung parenchyma [5,7]. Embolization occluding the arterial supply, leading to spontaneous regression as a consequence of progressive fibrosis of the dysplastic lung tissue and feeding vessel culminating in vascular thrombosis and infarction, and is an alternative treatment method to conventional surgical resection. Moreover, the sequestration's vascular supply is often friable due to inflammation from repeated infection, and embolization has been demonstrated to reduce the risk of intraoperative hemorrhage [8,9].

Pulmonary sequestration is rarely associated with lung cancer. The treatment of lung cancer associated with pulmonary sequestration is surgery after a thorough work-up and proper staging of the patient. In the case of an incidental finding, a postoperative workup needs to be undertaken to stage the tumor, and where necessary, additional treatment (chemotherapy, radiotherapy, or both) should be carried out [10, 11]. CPAMs are a group of cystic and non-cystic lung lesions resulting from early airway maldevelopment. CPAMs can communicate with the airways and are relatively frequently infected (30%). Blood supply is from the pulmonary artery with drainage via pulmonary veins. Hybrid lesions with histological and imaging features of both a CPAM and bronchopulmonary sequestration will have, by definition, a systemic arterial supply [6]. CPAM is a lesion characterized by the presence of anomalous bronchiolar or acinar structures, variable in size, either cystic or not cystic. CPAMs can communicate with the airways and are relatively frequently infected (30%). Blood supply is from the pulmonary artery with drainage via pulmonary veins. Hybrid lesions with histological and imaging features of both a CPAM and bronchopulmonary sequestration will have, by definition, a systemic arterial supply [6, 12]. In our 3rd case, we describe a 2-days old premature female newborn (7 months gestation age), with a very rare association between CPAM and IPS. Post mortem examination was not performed because we did not get consent from the patient's parent. This patient died because of her complex condition; severe respiratory distress, pneumonia, neonatal jaundice, sepsis, accompanied by other abnormalities, ASD and PDA.

The limitation of the study, we do not include photos related to anatomical pathology because our study is focused on explaining from the side of the radiologist.

#### 4. Conclusion

Pulmonary sequestration is a relatively rare entity comprising a small portion of all congenital pulmonary malformations. MDCT is the technique of choice for evaluating pulmonary sequestrations, MDCT depicts the feeding artery and the venous drainage, also allows simultaneous evaluation of the lung parenchyma. There is a very rare association between CPAM and IPS. Hybrid lesions with histological and imaging features of both a CPAM and pulmonary sequestration will have, by definition, a systemic arterial supply. Pulmonary sequestration is rarely associated with lung cancer and its treatment is still controversial. Radiology plays an important role in diagnostics which can then assist clinicians or surgeons in determining the next course of therapy, whether surgery, interventional radiology (embolization), or a combination.

#### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Guarantor

Anita Widyoningroem.

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#### Author contribution

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#### Registration of research studies

Name of the registry: -.

Unique Identifying number or registration ID: -.

Hyperlink to your specific registration (must be publicly accessible and will be checked): -.

#### Declaration of competing interest

The authors declare no conflict of interest.

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#### Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2022.103268>.

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