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Case Report

Intralobar pulmonary sequestration supplied by vessel from the inferior vena cava: Literature overview and case report ^{☆,☆☆}

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ABSTRACT

Sequestration is a congenital abnormality that can occur in both children and adults. The clinical presentation often manifests as recurrent pneumonia throughout the lifetime of the patient. Pathologically, sequestration is a disorganized region of lung parenchyma without a normal pulmonary artery and with no interconnecting air passage (ie, it is isolated from the bronchi and pulmonary arteries). Sequestration can be either intralobar or extralobar and is usually supplied with blood from an anomalous vessel originating from the thoracic aorta or abdominal aorta (big circulation/systemic circulation), which is one of the mandatory criteria for diagnosing sequestration. CT angiography or catheter angiography can assist in identifying the anomalous vessel both for diagnosis and surgical resection. We present a rare case of intralobar sequestration with the distinction of being supplied with blood from the abdominal vena cava, as confirmed by CT angiography, surgery, and postoperative pathology. It is hoped that this study will contribute to the literature by introducing a rare case of congenital lung abnormality and pulmonary vascular malformation.

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Introduction

Congenital lung abnormalities can be classified in several ways. The simplest classification usually consists of two

groups: (1) lung abnormalities not associated with vascular abnormalities; and (2) lung abnormalities associated with/or including vascular abnormalities [1]. Sequestration is a congenital abnormality derived from anomalies in the primitive alimentary canal bud and its related structures, which occurs

Abbreviations: MRR, Multiplanar reconstruction; VRT, Volume rendering technique; VAS, Video-assisted surgery.

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during lung, bronchial, and vascular development. As such, sequestration belongs to the group of pulmonary abnormalities associated with vascular abnormalities. Two common types are intralobar sequestration and extralobar sequestration, depending on whether or not it is covered by visceral pleura. Although intralobar and extralobar sequestration share some features, they also differ significantly in some important clinical and radiographic features.

Intralobar sequestration is the more common of the two types. Intralobar sequestration is wrapped in the visceral lobe of one lung, and is more common in the left lung, with two-thirds of cases found in the costophrenic sulci of the left lower lobe posterior basal segment. In 75% of cases, the artery supplying the sequestration usually originates from either the thoracic aorta, the abdominal aorta (or its branches), or the intercostal arteries. Abnormal arteries arising from the systemic artery usually enter the lung via the basal ligaments. Usually, venous return is via the pulmonary vein, although in rare cases it can be via the azygos or hemiazygos system. However, it is extremely rare for sequestration to receive blood supply from the vena cava. The literature has also not seen unclear mention and has not been documented in case reports around the world [1,2].

Intralobar sequestration can occur in both adults and older children. Acute or recurrent pneumonia is a common symptom in these patients and hemoptysis may also occur. Pulmonary shunts, although also a symptom, are usually small and of no clinical significance. However, some cases of congestive heart failure have been reported. Bilateral sequestration can also occur, and esophageal involvement or other congenital anomalies have been observed, although rarely [1].

Intralobar sequestration has many different manifestations in the uncomplicated stage. It may appear as a well-defined and co-attenuated mass, as multiple cysts containing gas or fluid, as a hyperlucent and hypovascular area of the lung, or a combination of the above. Hyperlucency is common in isolated, uncomplicated lungs due to air trapping, and is difficult to detect on a plain-film chest X-ray; however, it is often seen on CT. The presence of mucus or fluid cysts with fluid-air levels may be seen with or without infection. In many cases, sequestration can look a lot like a lung abscess. Bilateral sequestration may occur, but very rarely [1–3].

Case report

A 40-year-old female was hospitalized with abdominal pain on February 13, 2021 (negative rRT-PCR of COVID-19 and one-dose mRNA COVID-19 vaccine). The patient had a history of coughing and fever from a young age and was often diagnosed with bronchitis, which was cured with a course of antibiotics. The patient had also had many chest X-rays, although no abnormalities were detected. Three days before admission, the patient felt pain in the upper abdomen, penetrating to the back, but experienced no nausea, vomiting, cough, or fever. The patient went to Bach Mai hospital for a chest X-ray. The radiologist noted an abnormality on the image, but the patient's family did not want to continue treatment at the hospital. The patient then went to the National Lung Hospital, where they



Fig. 1 – Chest radiograph on admission (high-kV film). Suspected abnormal opacity located close to the spine, medial to the right radial-diaphragmatic angle (arrow).

were subsequently hospitalized due to abnormality on the X-ray.

The patient was thin with pale skin, and no edema was noted. Pulmonary ventilation was normal, with no rales and no fever, and the patient had an SpO₂ of 95% (breathing air). A test for tuberculosis came back negative, and blood count, blood chemistry, and CRP levels were all within normal limits. Additionally, the blood coagulation and respiratory function tests were within normal limits, and an abdominal ultrasound and echocardiogram revealed no abnormalities. However, a bronchoscopy revealed inflammation of the bronchial mucosa.

The patient had a chest X-ray on admission using the high-kV technique, and the results are shown in [Figure 1](#).

The patient had chest CT scans using a 64-slice machine before intravenous contrast injection, immediately after injection, and 20 seconds after injection (later). Xenetix contrast agent was used (350 × 100 ml) and was injected at 4 ml/sec. The parameters used were 139 kV and 114 mA, with a slice thickness of 3 mm. The WW/WL was 1200/–800 (lung window) and 350/50 (mediastinal window). The image was then reconstructed in 0.75 mm slices using multiplanar reconstruction (MPR) and Volume rendering technique (VRT) rendering.

The chest CT scan showed an abnormal mass with soft tissue density in the basal segment of the right lung, located close to the lung edge and the spine. In particular, there was a prominent pulmonary vein-like structure that drained from the mass into an azygos vein in front of the spine (see [Fig. 2](#)).

On the angio CT, the MPR image shows a branch vessel originating from the abdominal vena cava just above the liver veins, passing through the diaphragm with the inferior vena cava, ascending outward through the tissue mass and branching into the normal lung parenchyma (see [Fig. 3](#)).

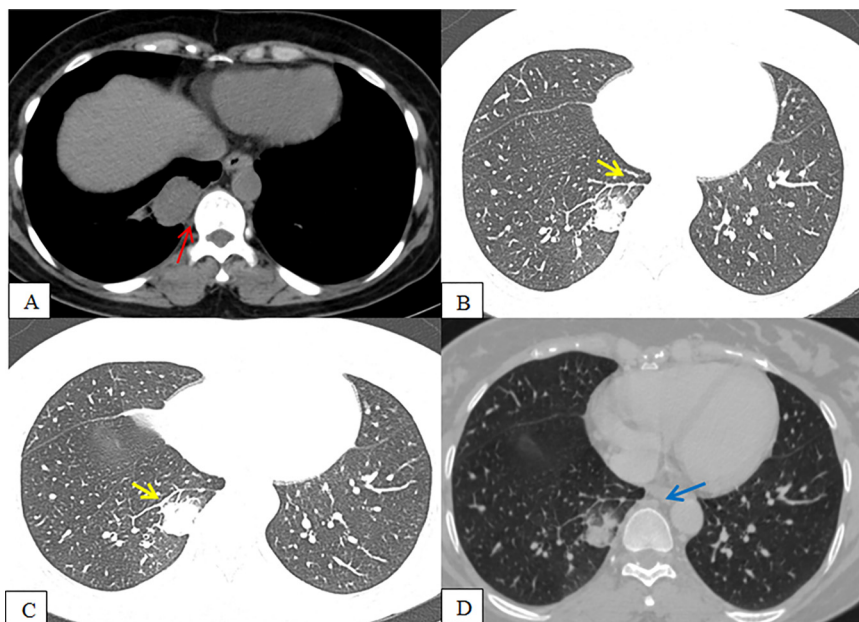


Fig. 2 – CT scan of the lung window and mediastinum before intravenous contrast injection. A: A soft tissue mass is seen, with a size of 36 × 26 mm (measured on lung window), a clear borderline, located close to but still separate from the spine (separated by a fat layer denoted by the red arrow). B, C, D: Anteriorly, there is a characteristic image of the pulmonary vein branching into the soft tissue space in front of the spine and D: connecting to an azygos vein (blue arrow).

The late-phase chest CT (20 seconds after injection) shows that the mass has an irregular structure with many mucilaginous cysts (foci of low density). The origin of the abnormal vessel branch, from the inferior vena cava, is more clearly seen on the coronal image (see Fig. 4).

On 3D rendering (VRT) of the thin slices reconstructed after contrast injection, the abnormal branch arising from the abdominal vena cava is more clearly seen (see Fig. 5).

Because the abnormal mass adheres closely to the spine and is somewhat continuous with the synaptic foramen, it is necessary to exclude this mass as a possible nerve tumor (Schwannoma or Neurofibroma) using Gadolinium-enhanced magnetic resonance imaging (see Fig. 6).

After collating all the imaging and clinical data, the patient fit the criteria to be diagnosed with interlobar sequestration. The only difference from other cases in the literature was that this sequestration was supplied from a branch of a vessel that originated from the venous system (the abdominal vena cava). The logic of this blood supply is to remain in the direction of the vena cava circulation (from bottom to top).

The case was consulted on by a multi-specialist team, and video-assisted surgery was proposed to remove the sequestration mass. The surgical process is shown in Figure 7. The postoperative report (February 25, 2021) clearly shows the ascending vessel branch from below the diaphragm. The injury to the right lower lobe adheres to the diaphragm and the triangular ligament. Branch resection with one cardiac Stapler, then resection of the sequestration mass with three other cardiac Staplers. The apex of the lung was tested with water before being closed. A right pleural drainage tube was inserted before the trocar was withdrawn. The surgery was successful, and the patient's drainage tube was removed 4 days later.

Macroscopic surgical specimens were analyzed under a microscope for signs of pathology. The macroscopic results are shown in Figure 7, while the microscopic results are shown in Figures 8 and 9. It was interesting to note that there was no venous valve in the lumen of the abnormal vessel that supplied blood to the sequestration mass.

Biopsy of the lung parenchyma showing thick alveolar walls, foaming macrophages, multiple lymphocytes infiltrating the interstitial tissue, fibrosis, chronic inflammation, and an abnormal artery entering the nest. The lung wall is thick and fibrous, with cystic organization infiltrating chronic inflammation. Conclusion: Intralobar sequestration.

Biopsy specimen showing the organization of the artery wall consisting of an inner, middle, and outer layer. The inner layer is covered by flattened squamous epithelial cells, the middle layer consists of a ring-shaped elastic layer, and the outer layer is composed of fibroblasts. Conclusion: Histopathological image is consistent with the organization of the artery wall.

The patient was discharged, in good health, 10 days after the operation, and had a follow-up examination 1 month later. The chest X-ray image 40 days after surgery is shown in Figure 10 and a chest CT image 10 months after surgery is shown in Figure 11.

Discussion

Sequestration is a congenital anomaly derived from an abnormality in the primordial alimentary canal bud, with its structural abnormalities occurring during lung, bronchial, and vas-

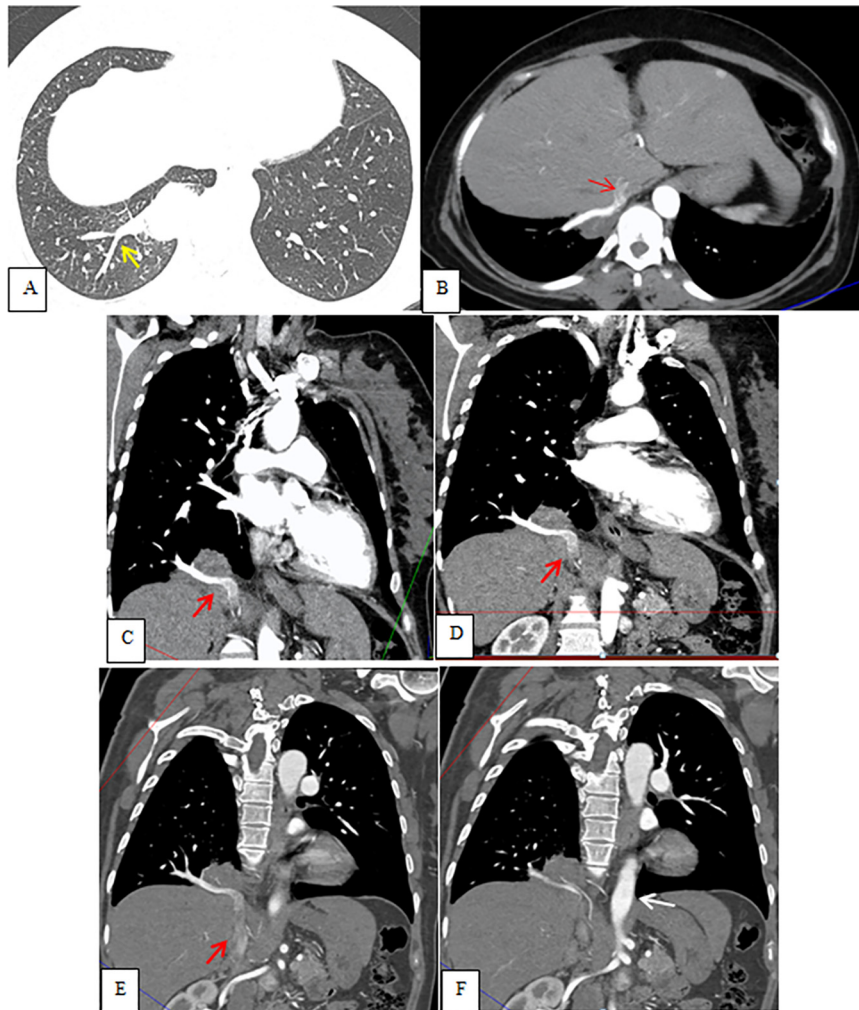


Fig. 3 – Images of chest CT scan in lung and mediastinal windows after contrast administration (early phase). Axial and MPR images. **A:** The lung window shows an abnormal vessel emerging from the mass, branching in the normal lung tissue (yellow arrow). **B:** The vascular branch is separated from the abdominal vena cava (just above the point of origin of the hepatic vein); **C, D, E:** Branches via the diaphragm, ascends outwards, penetrates the mass, and branches in the parenchymal region to the right border of the mass (red arrow). **F:** The abdominal aorta (which is not associated with any other abnormal vessel) passes through the diaphragm and branches off the intercostal vessels (white arrow).

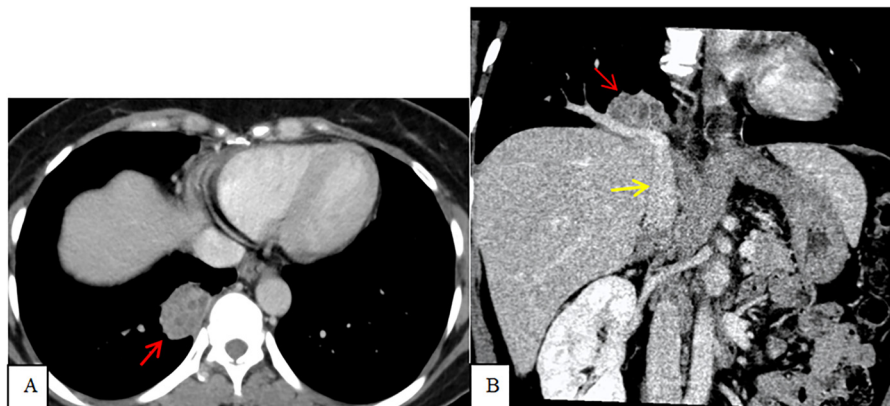


Fig. 4 – Late-phase chest CT scan after intravenous contrast. **A, B:** Axial and coronal mediastinal windows show a mass with many cyst-like hypodense foci inside (red arrow). **B:** Abnormal branching to sequestration arising from the abdominal vena cava (yellow arrow).

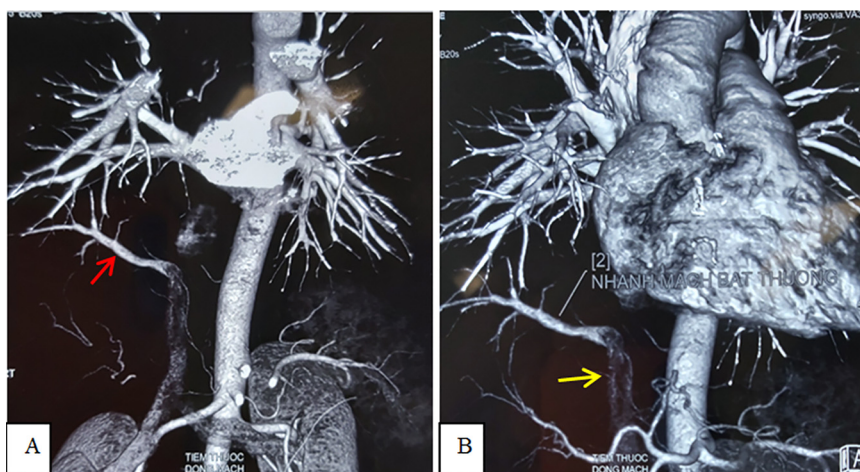


Fig. 5 – VRT anterior chest image view. A: Abnormal branch of blood supply to sequestration mass (red arrow). B: Abdominal vena cava (yellow arrow).

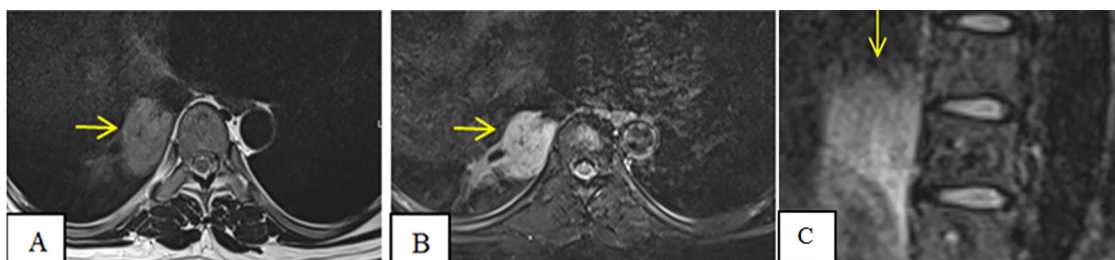


Fig. 6 – Magnetic resonance imaging (MRI) of the thoracic spine with intravenous Gadolinium injection. A: T2 Stir; B: T2 Stir after injection (Axial) and C: T2 Stir after injection (Coronal) both show a dissected right lower lobe parenchymal mass, unrelated to the spine (arrow).

cular development. Pathologically, sequestration is an area of disorganized lung parenchyma, with no normal pulmonary artery, and without an interconnected airway (isolation of the bronchi and pulmonary arteries). It can manifest as mass consolidation, or as a cystic or polycystic lesion containing gas or fluid [1–5]. In our case, the isolated lung mass was solid but contained multiple mucinous cysts.

The sequestration usually receives its blood supply from the branches of the thoracic or abdominal aorta – the exact origin will need to be confirmed by angiography before surgery and severe bleeding can occur if these branches are accidentally cut during surgery [1,4,5]. Our case report is a relatively rare instance of the vessel supplying the sequestration mass dissecting from the abdominal vena cava. Another difference was that although the branch was separated from the abdominal vena cava, both macroscopic dissection and microscopic pathology confirmed that this was an arterial structure, rather than a vein, something that is not mentioned in the literature to the best of our knowledge. When analyzing this case, we questioned whether this was an abnormal case of anomalous pulmonary venous return. However, this possibility was ruled out by the surgeons and pathologists involved in this case.

There are two distinct types of sequestration: intralobar and extralobar [1,4,6,7]. Although they share some features, they also differ significantly in some important clinical and

radiographic features [1,2,4]. Intralobar sequestration is the more common of the two sequestration types. In this type, isolated lung tissue was located in the visceral lobe of one lung, was more common on the left side, and approximately two-thirds of cases were found in the paraphrenic region of the left lower lobe (posterior basal segment). Due to the peculiarity of appearing in the lower lobe, close to the diaphragm, sequestration is often easily missed on conventional chest X-rays, particularly if the lesion is small (and especially if it involves the posterior lung, close to the spine). Our patient had been x-rayed many times previously, but the abnormality was located in a difficult-to-observe position and had always been missed. However, this abnormality will always be missed if a low-kV chest X-ray is taken, as the sacs along the diaphragm are in a “blind area.” A high-kV chest X-ray, by contrast, will allow the spine and parenchyma of the sacral-diaphragmatic sacs to be clearly visualized.

In 75% of intralobar sequestration cases, the artery supplying the sequestration usually originates from the thoracic aorta, the branches of the abdominal aorta, or the intercostal arteries. The systemic arteries normally enter the lungs via the inferior pulmonary ligaments. Usually, venous return is via the pulmonary vein, although the vein does occasionally drain into an azygos or semi-azygos vein [1–3,8]. In our case, the sequestration mass was located in the lower lobe of the

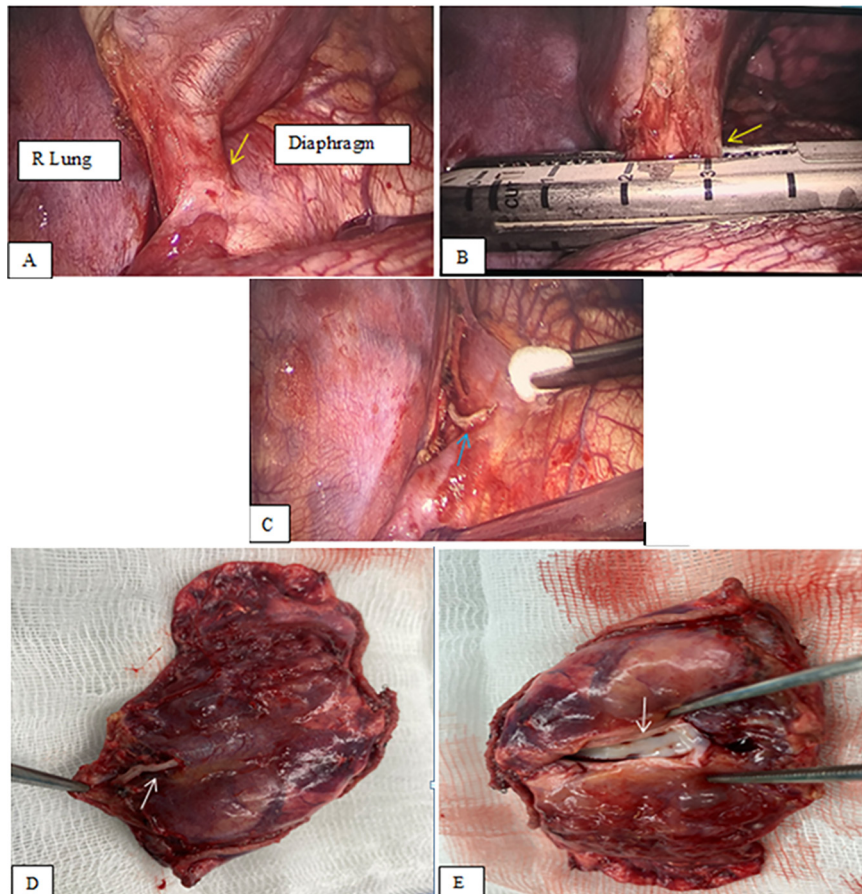


Fig. 7 – Observational images during laparoscopic surgery and gross specimens taken after surgery. A: Abnormal vascular branch ascends from the superior surface of the right diaphragm (yellow arrow). B: Vasectomy with cardiac staple. C: Posterior vascular truncation process cut above the diaphragm (blue arrow). D, E: Postoperative isolated lung mass and abnormal branch lumen which was smooth and showed no venous valve (white arrow). Conclusion: The macroscopic image is consistent with the sequestration structure and the blood supply artery.

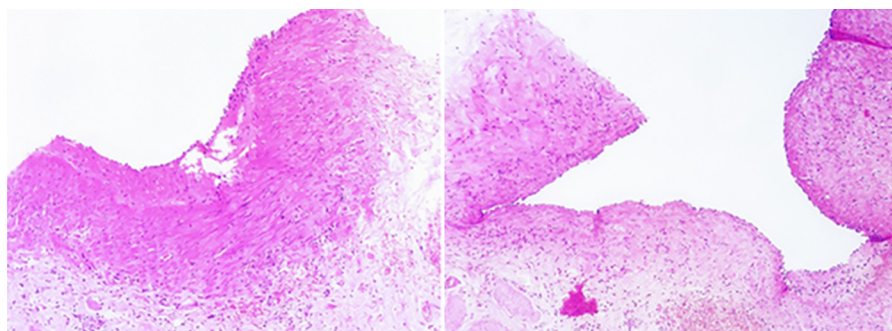


Fig. 8 – Microscopic anatomy of the sequestration specimen after surgery.

right lung and had a venous system draining from the sequestration mass into an azygos vein. Although rare, this has previously been reported in the literature.

Sequestration can occur in both adults and older children. Acute or recurrent pneumonia is common in these patients [1,2,4,5,9] and hemoptysis may also occur. Dynamic pulmonary venous shunts due to sequestration veins are usually

small and of no clinical significance. However, some cases of congestive heart failure have been reported. Bilateral sequestration can occur, and esophageal involvement or other congenital anomalies have also been observed, although these are rare [10].

Our patient is middle-aged, despite the long-term course of the disease, but they have never suffered from hemoptysis.

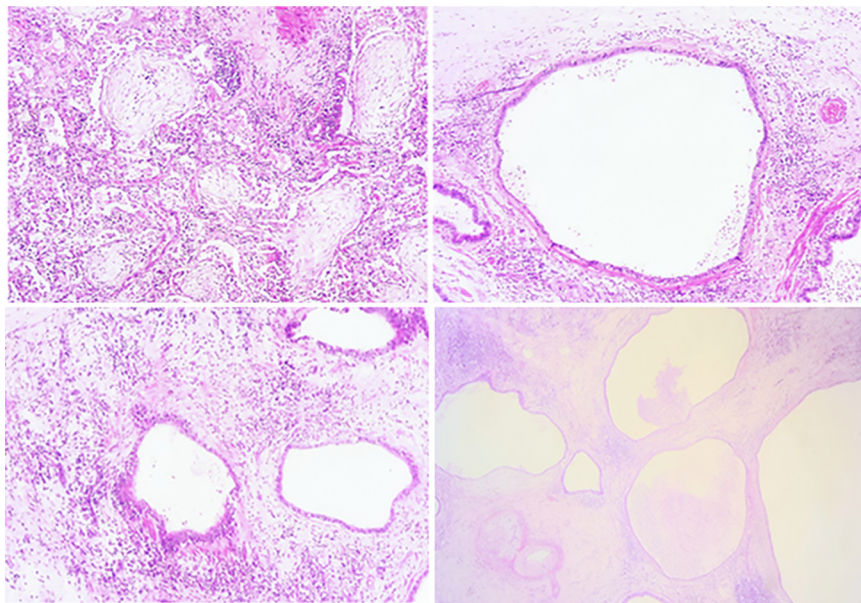


Fig. 9 – Microscopic anatomy of the patient's blood supply to the sequestration mass after surgery.

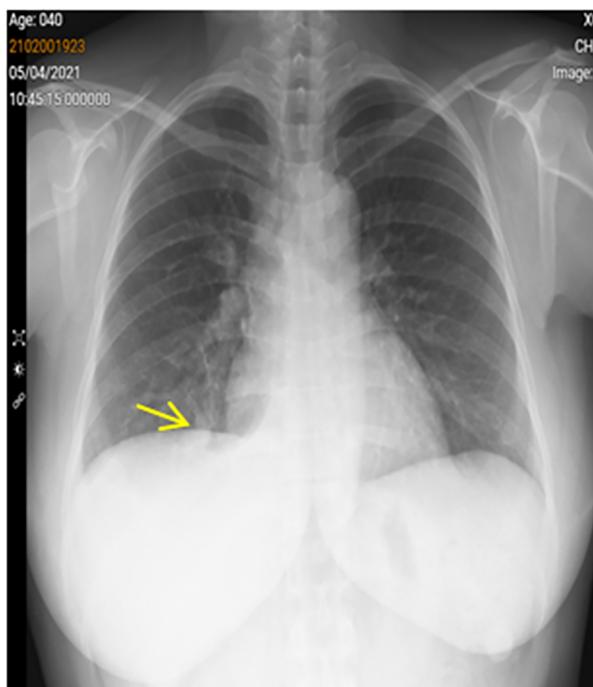


Fig. 10 – Chest X-ray 40 days after surgery. A blurred band is visible at the base of the right lung (arrow).

In our opinion, this may be because the sequestration mass was supplied with blood from the vena cava branch, meaning that the pressure would be much lower than arterial pressure, and hemoptysis would not occur.

Intralobar sequestration has many different manifestations in the absence of other complications. It may present as a homogeneous, well-defined mass lesion, one or more mul-

ticystic air- or fluid-filled lesions, a hyperlucent and hypovascular area in the lung, or a combination of the above. Hyperlucency is common in isolated, uncomplicated lungs because of air trapping. It is often difficult to see on plain-film chest X-rays but can often be seen with CT. The presence of mucus or fluid-filled cysts with fluid-air levels may be seen both with and without infection. In many cases, sequestration can look a lot like a lung abscess. A mentioned earlier, bilateral sequestration can occur, although rarely. They are usually supplied with blood by a branch of the artery that originates from the systemic artery [11]. In our patient's chest, CT film with contrast injection, a rather typical image of a sequestration mass appeared late: Solid mass, with many foci of reduced density in the form of mucus.

On a chest CT, a normal bronchus or pulmonary artery can be seen above the lesion but not in the sequestration mass. On contrast-enhanced spiral CT, the systemic artery supplying the sequestration is sometimes seen. If it cannot be seen, then angiography may be used to confirm diagnosis. Venal drainage can also be seen when contrast is injected [1,4,5,7].

Surgical resection of an isolated lung mass is essential for definitive treatment [4,9]. In the past, before laparoscopic surgery was developed, surgeons often had to perform open surgery. The lobectomy technique is often preferred over selective resection of sequestration masses because the lungs are elastic, air-filled organs, and apices are difficult to suture as a result [4,5,9]. Today, laparoscopies, lobectomies, and selective pneumonectomies are widely applied, and the use of staples for vascular resection and apical suturing has become quick and safe. With just one push of the slider, two side stitches can be added and a cut between the two lines can be made. In order to minimize bleeding during surgery, the intervention of the branch vessel that supplies blood to the sequestration is usually performed before surgery. New plugging materials are also constantly being developed. Using glue pumps

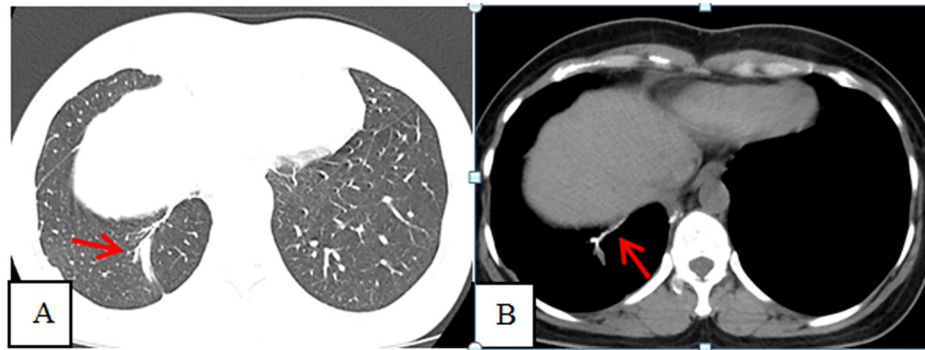


Fig. 11 – Unenhanced chest CT 10 months after surgery. A strip of the staple suture is still visible in the right lower lobe.

for small vessels and coils for large vessels, which temporarily or permanently occlude the blood supply branch, combined with anti-inflammatory treatment before surgery, has minimized iatrogenic bleeding and infection complications [4,10]. Our patient is still in good health 12 months after surgery. On the chest CT, the apical seams of the lung were clearly visible along with the stapled blood vessels, demonstrating absolute parenchymal stability.

Extralobar sequestration is an abnormality where the isolated lung tissue is surrounded by a separate membrane. It is less common than intralobar sequestration and 90% of cases occur in the base of the left lung, adjacent to the left diaphragm. The arterial supply is usually from the abdominal aorta and venous return is mostly via the major venous systems (inferior vena cava, azygos vein, semi-azygos vein, and portal vein). Unlike intralobar sequestration, extralobar sequestration is usually found in young children. It was discovered incidentally and presented as a mass lesion. Unlike with intralobar sequestration, tissue infection is very rare. It is often associated with other congenital anomalies, particularly diaphragmatic abnormalities and ipsilateral pulmonary hypoplasia. Because it is completely surrounded by its own pleura, extralobar sequestration rarely becomes infected.

On X-ray and CT, extralobar sequestration appears as a well-defined mass that does not contain air (unlike intralobar sequestration). It usually presents as a homogeneous lesion but may also present with peripheral air-filled cysts. The supplying artery can be seen on angio CT. If absent, angiography may be necessary for diagnosis [1].

Conclusion

In summary, we report a rare sequestration case regarding a group of congenital lung abnormalities and pulmonary vascular malformations. The difference, in this case, is that the blood supply to the sequestration was not from the systemic artery. The clinical symptoms are similar to those of common intralobar sequestration, but chest X-rays consistently missed abnormality due to the mass being small and located in the right lung X segment. A multi-slice chest CT with intravenous contrast (angio CT) combined with imaging reconstruction techniques (MPR, VRT) will help us accurately diag-

nose this disease in the future. Surgical resection with video-assisted surgery (VAS) has allowed the patient to lead a normal life again.

Authors' contributions

Cung-Van C and Nguyen MD contributed equally to this article as co-first authors. All authors read and approved final version of this manuscript.

Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

Ethics approval

Not applicable.

Patient consent

Written informed consent was obtained from the patient for the publication of patient information in this article.

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