

## Case Report

# Diagnostics and multimodal therapy for pulmonary sequestration

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**Abstract:** To investigate the diagnostics and multimodal therapy for pulmonary sequestration. Eleven patients with pulmonary sequestration received treatment between January 2008 and April 2015. Their diagnoses and treatment courses were analyzed. Nine preoperative diagnoses of intralobar pulmonary sequestration were confirmed, while two patients with extralobar pulmonary sequestration were misdiagnosed preoperatively. Enhanced computed tomography (CT) scans showed that the abnormal arteries in seven patients originated from the thoracic aorta, and in two patients, they originated from the abdominal aorta. The diameters of these abnormal arteries ranged from 5 to 15 mm. Seven patients had one abnormal artery and two had many abnormal arteries. The CT scan results were consistent with the intraoperative findings. The procedures performed are as follows: one patient underwent pre-operative embolization therapy, six underwent video-assisted thoracoscopic surgery (VATS), five underwent conventional thoracotomy, nine underwent lobectomy, and two underwent simple resection of sequestered tissues. The operations were successful in all cases without any complications, and postoperative follow-ups revealed that all patients recovered well. No specific symptoms were observed in patients with pulmonary sequestration. Enhanced CT scans are now used as the gold standard for diagnosis of pulmonary sequestration. Surgery is the most effective treatment, and the key to successful operative procedures is the proper handling of abnormal arteries. Conventional thoracotomy and minimally invasive VATS are both effective methods of treatment. Lastly, endovascular embolization can be used in the emergency treatment for massive hemoptysis in pulmonary sequestration patients.

**Keywords:** Pulmonary sequestration, clinical diagnosis, combined therapy

### Introduction

Pulmonary sequestration is a rare congenital lung malformation disease [1]. It can be divided into two subgroups, according to the site of occurrence, namely the intralobar type and extralobar type; intralobar sequestration is more commonly observed. The clinical misdiagnosis rate is relatively high. During January 2008 and April 2015, we performed surgical treatments for 11 cases of pulmonary sequestration. These accounted for 0.16% of lung surgery performed in our department during the same period. We investigated the diagnostics and multimodal treatments of pulmonary sequestration, based on clinical data.

### Patients and methods

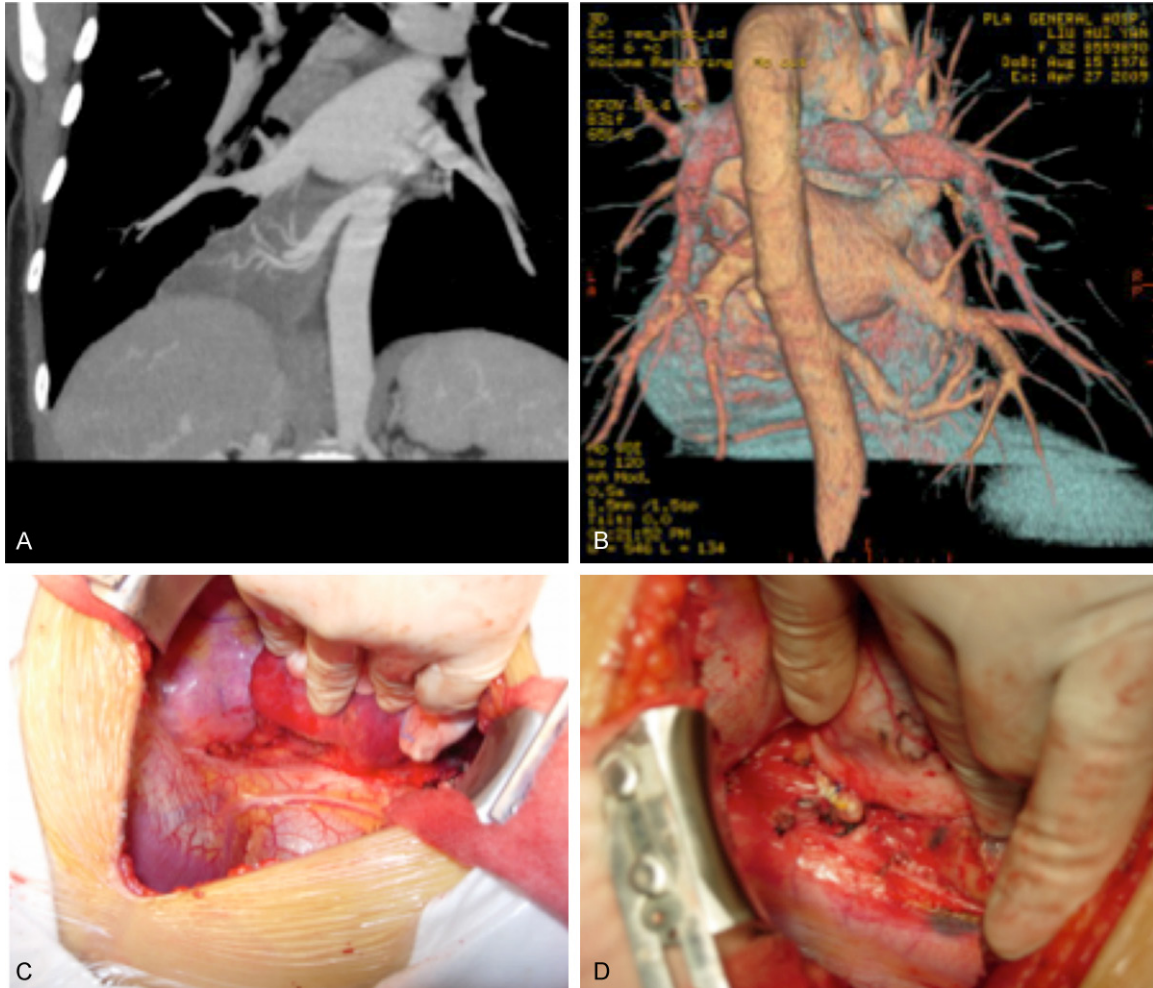
#### *General characteristics*

This study comprised of 11 patients (age range, 21-63 years; 7 men and 4 women), out of which

9 patients had a history of recurrent fevers, cough, and expectoration. Five out of the 9 patients also had intermittent hemoptysis and one had massive hemoptysis. The duration of this disease was between 2 and 35 years. Two patients were asymptomatic.

#### *Diagnostic methods*

Chest radiographies showed tumor-like lesions in three patients, cyst-like changes in two, inflammation-like changes in two, and no abnormal changes in five. Lung CT scans showed that six patients had lesions in the left lower lobe, three had lesions in the lower right lobe, one had lesions in the left cardio-phrenic angle, and one had lesions in the left costophrenic angle (**Figure 4A**). Enhanced CT scans of the chest of 9 patients showed arterial abnormalities. In 7 cases, abnormal arteries originated from the thoracic aorta and in 2 cases abnormal arteries originated from the abdominal aorta (**Figures**



**Figure 1.** Lower right intralobar pulmonary sequestration. A. Enhanced CT scans showing the abnormal artery originating in the thoracic aorta. B. Three-dimensional reconstruction of enhanced CT scans showing the abnormal artery originating from the thoracic aorta. C. Conventional surgery for removal of abnormal arteries. D. Abnormal artery stump after continuous suture.

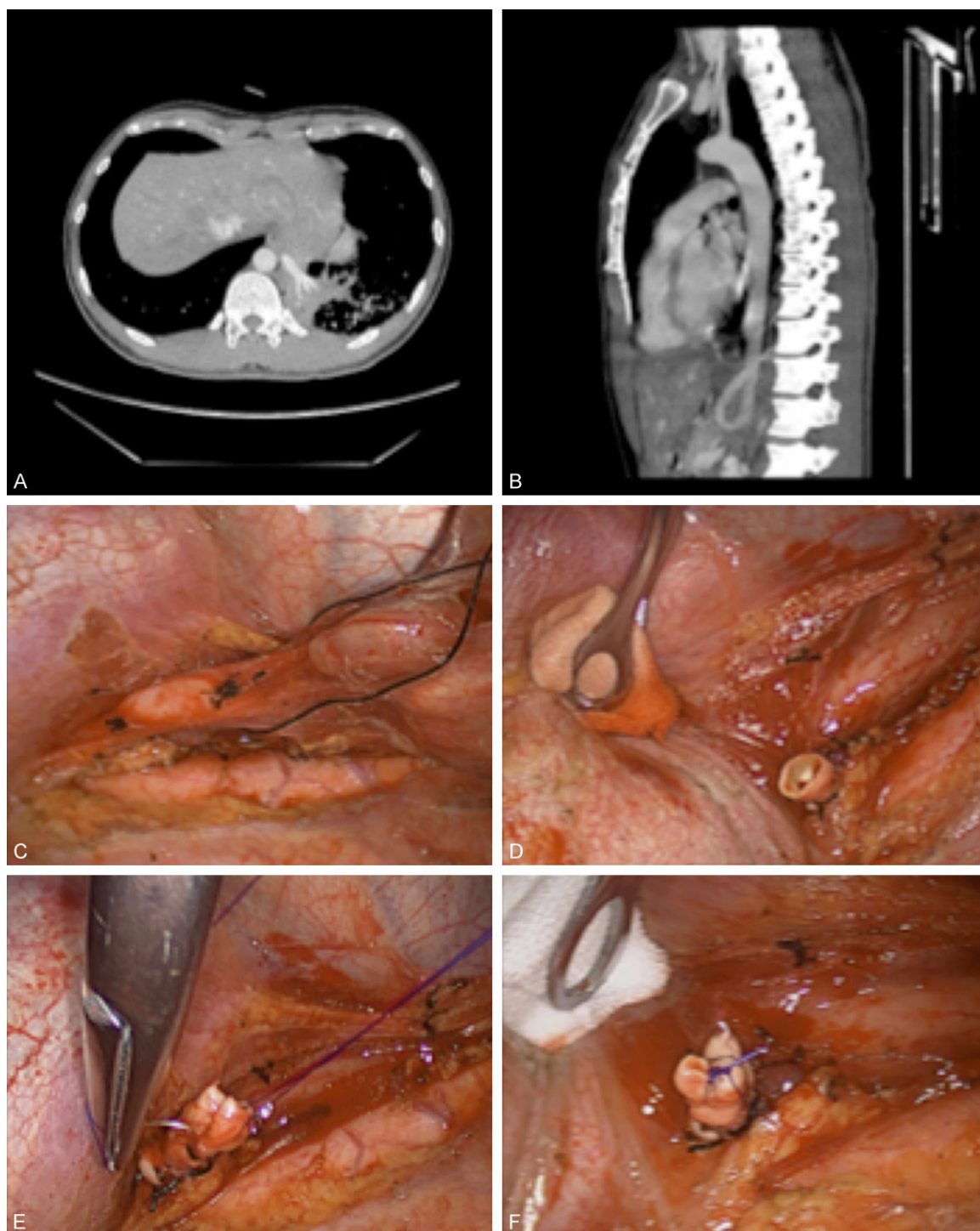
**1A, 1B, 2A, and 2B).** In two cases of left lower lung pulmonary sequestration, two abnormal arteries were found, both adjacent to the thoracic aorta (**Figure 3A** and **3B**). In one case, enhanced MRI examination suggested a tumor on the left side of the diaphragm (**Figure 4B**). Preoperative diagnoses suggested nine cases of pulmonary sequestration, one case of posterior mediastinal tumor, and one case of diaphragmatic tumor.

#### *Surgical treatment*

All procedures were performed under general anesthesia with a double-lumen endotracheal tube and single-lung ventilation. The choice of surgical incision was determined as follows: (1)

VATS surgery: the observation port lay at the posterior axillary line in the 8<sup>th</sup> intercostal space with a length of 1 cm, and the port into the chest was located in the 4<sup>th</sup> intercostal space between the anterior and posterior axillary lines with a 4-6-cm incision. (2) Conventional thoracotomy: a posterolateral incision, 10-15 cm long, was made in the 5<sup>th</sup> intercostal space.

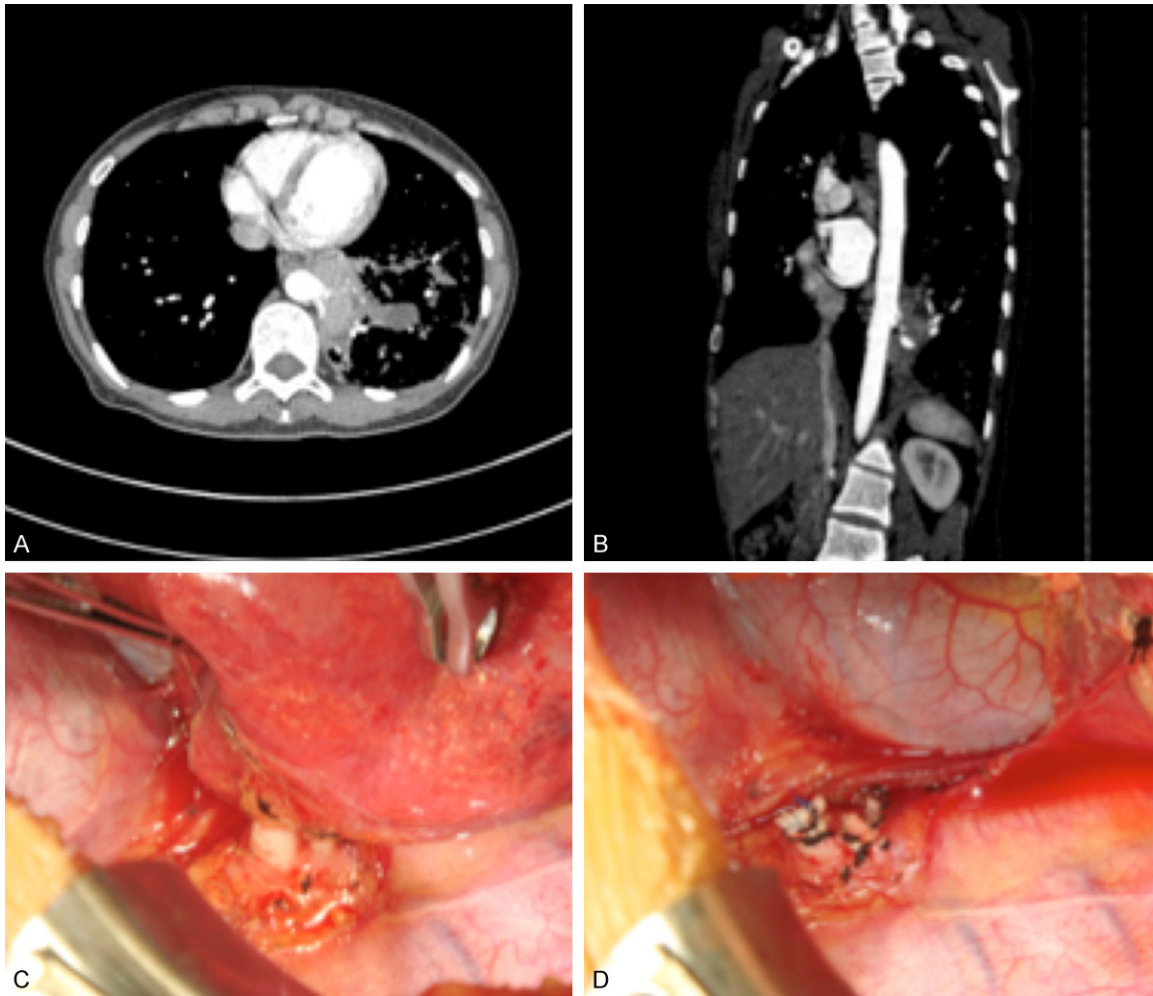
For the cases with intralobar pulmonary sequestration, the abnormal arteries were located using thin-section CT scans and then carefully separated from the surrounding tissues during the surgery (**Figures 1C, 2C, and 3C**). The abnormal arteries were ligated twice with No. 7 heavy silk threads at the proximal end and then the ends were sutured continuously



**Figure 2.** Lower left intralobar pulmonary sequestration. A. Enhanced CT scans showing the abnormal artery in the lower left lobe. B. Digital reconstruction of enhanced CT scans showing the abnormal artery originating from the abdominal aorta. C. Thoracoscopic surgery for removal of abnormal arteries. D. Abnormal arterial stump after two ligations. E. Thoracoscopy to suture the ends of the abnormal artery. F. Abnormal artery stump after continuous suture.

with a 4-0 absorbable suture (**Figures 1D, 2D-F, and 3D**). Afterwards, the arteries and veins of the lobe with pulmonary sequestration were

isolated and treated with vascular closure devices. Lastly, the bronchi were isolated and treated with bronchial closure devices.



**Figure 3.** Lower left intralobar pulmonary sequestration. A. Enhanced CT scans showing two abnormal arteries in the lower left lung. B. Digital reconstruction of enhanced CT scans showing two abnormal arteries originating from the thoracic aorta. C. Conventional surgery for removal of two abnormal arteries. D. Two abnormal arterial stumps after double ligations and continuous sutures.

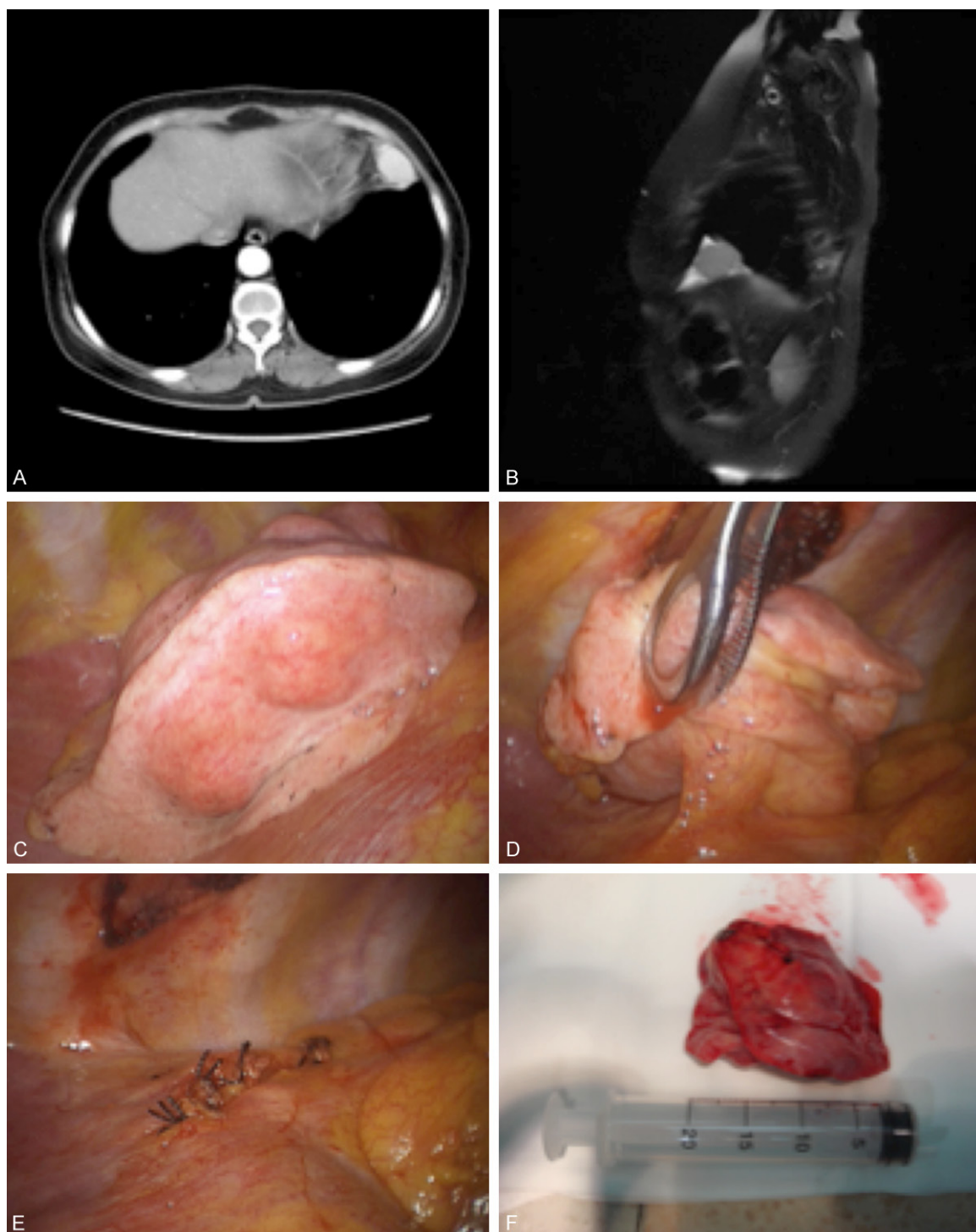
The extralobar sequestration lung tissue had a fetal-like appearance (**Figure 4C** and **4D**). The feeder arteries were abnormally small and were not detected using enhanced CT scans. During the surgical process of isolating the tissues, No. 4 silk threads were used for ligation (**Figure 4E**). The tissues could also be removed with a linear cutter.

## Results

### *Surgery results*

The operation was conducted on the right side of the chest in three patients and on the left in eight patients. VATS was used in six cases, while conventional thoracotomy was performed in five. During the operations, nine patients

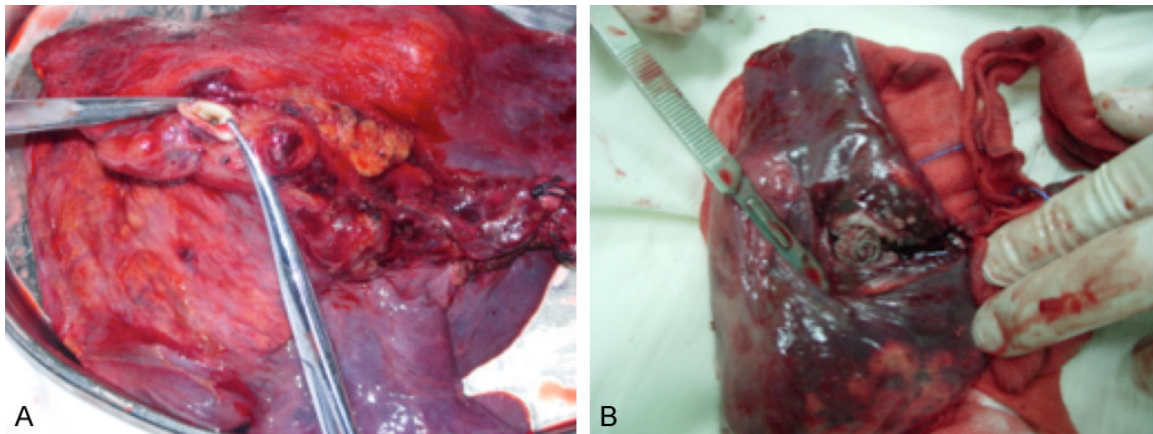
with intralobar pulmonary sequestration were found to have apparent abnormal feeder arteries, which originated from the thoracic aorta in seven of the nine patients or from the abdominal aorta in two of the patients. Seven patients had one abnormal artery, while two patients had two abnormal arteries. The diameter of the blood vessels was 5-15 mm. The abnormal arteries found during the operation were consistent with the findings of the preoperative enhanced CT scans. Lobectomy at the lesion sites were performed in all of the nine patients with intralobar sequestration; out of these patients, the left lower lobe was resected in six patients, while the right lower lobe was resected in three. Preoperative enhanced CT scans of the two extralobar pulmonary sequestration



**Figure 4.** Left chest extralobar pulmonary sequestration. A. Enhanced CT scans suggesting left anterior mediastinal tumors. B. Enhanced MRI diagnosis of left diaphragmatic tumors. C. VATS confirmed extralobar pulmonary sequestration. D. Extralobar sequestration tissue attached to the diaphragm. E. Ligation of the stumps of small feeder arteries. F. Extralobar sequestration tissues with fetal-like morphology.

cases showed no clear abnormality in the arteries; however, during the separation of the adhesive tissues, suspicious feeder arteries were

noted, and all were simply resected from the affected lung tissues. There were no complications during the operations in all cases, no



**Figure 5.** Lower left intralobar pulmonary sequestration following endovascular embolization. A. Congestion and consolidation of sequestered tissue and thickening and hardening of the abnormal arterial walls. B. The specimen was dissected to reveal the bracket.

intraoperative and postoperative bleeding, and no mortality.

## Pathology results

Among patients with intralobar pulmonary sequestration, lung consolidation was observed in eight cases. In one case where endovascular embolization was performed, there was abnormal hardening and thickening of the arterial walls (**Figure 5A** and **5B**). For two cases with extralobar pulmonary sequestration, we found fetal-like lung tissues (**Figure 4F**).

## Follow-up results

Follow-ups were conducted for 1-7 years, and all the patients showed good recovery and resumed normal life and work styles.

## Discussion

Pulmonary sequestration is a rare congenital lung malformation disease first reported by Pryce in 1946 [1]. It mainly occurs in the lower lobes of the lungs, with the lower left lobe being the most common site. There is no clear boundary between intralobar sequestered tissue and its surrounding normal lung tissue, while extralobar sequestered tissue is isolated from normal lung tissue. The abnormal feeder arteries of pulmonary sequestration usually originate from the thoracic aorta, and in most cases, only one of them is present. The clinical symptoms are nonspecific, and early-stage clinical misdiagnosis rates are higher than 95% [1, 2]. In this study, all nine patients with intralobar

pulmonary sequestration had been treated in the internal medicine department as chronic pneumonia patients with hemostatic therapy or anti-infection therapy.

No specific characteristics are observed on the chest radiography examination for pulmonary sequestration. As a result, it is often misdiagnosed as bronchiectasis or pneumonia. In some patients, a CT scan may show indirect signs of abnormal arteries. If the affected area of the lung and thoracic aorta are connected by rat-tailed tissue, the patient is advised to undergo enhanced CT scanning. The abnormal feeder arteries are visualized clearly on enhanced CT scans, and the digital reconstruction technique provides clear information on the origin, number, and diameter of the abnormal feeder arteries. Currently, clinically enhanced CT scanning has replaced angiography and has become the gold standard for the diagnosis of pulmonary sequestration [2-5]. An enhanced MRI can be adopted as supplementary to a CT examination. Since abnormal arteries originate from the thoracic and abdominal aorta, for patients with suspected pulmonary sequestration, the upper abdomen should be examined. Alternatively, the abnormal arteries in extralobar pulmonary sequestration are often much smaller and cannot be detected using enhanced CT scans. As a result, preoperative misdiagnoses are common.

In this study, the preoperative enhanced CT scanning results of nine patients with intralobar pulmonary sequestration were consistent

with surgical findings, and hence, the preoperative diagnostic accuracy was 100%. Nevertheless, the two patients with extralobar pulmonary sequestration were misdiagnosed with thoracic tumors on both the preoperative enhanced CT scans and by the enhanced MRI examination.

For patients with pulmonary sequestration, the recurrent secondary infection or hemoptysis makes it difficult to eliminate the symptoms with simple conservative treatment, such as anti-bacterial agents. Most researchers believe that once pulmonary sequestration is diagnosed, either surgical treatment or embolization should be performed as soon as possible [2].

Surgical resection is the most effective method of treatment and includes traditional surgery as well as the minimally invasive VATS. The minimal invasive VATS is a safe procedure with several advantages such as smaller surgical openings, quicker recovery, and cosmetically appealing incisions. Currently, VATS lower lobectomy is already widely used in clinical settings to treat cases with lung cancer [6].

In cases of intralobar pulmonary sequestration with chronic recurrent infections that have spread to the surrounding normal tissue, it is often difficult to completely remove the lung lesions locally. Therefore, the surgical procedures usually include abnormal artery transection and lobectomy. Because abnormal arteries tend to have more adhesive tissue surrounding them, surgeons must examine the preoperative thin-section CT scans carefully to precisely locate these arteries. During surgery, blunt dissection should be performed when isolating corresponding sections. Electrocautery and dissection should not be performed blindly so as to avoid damages to abnormal arteries. In cases where the abnormal feeder artery originates from the abdominal aorta, when the abnormal artery is damaged during surgery, it may retract under the diaphragm and causes fatal bleeding that cannot be ligated. One should isolate more than 1 cm of abnormal arteries. Because the abnormal arteries lack muscle lining and possess lower elasticity than regular vessels, we consider that the best practices are to ligate their proximal ends twice with No. 7 heavy silk threads, transect them as close as possible to the sequestered lung tis-

sue, and afterwards, use the 4-0 absorbable suture continuously on the ends. The area should be examined carefully to ensure that there is no blood oozing around the stumps. This treatment is more reliable and is a safer method of handling abnormal arteries with a diameter greater than 10 mm, during both conventional surgery and VATS.

In this study, all patients were treated with this procedure, which did not result in any hemorrhage and was easy to operate with a thoracoscope. Extralobar sequestered tissues mostly have a fetal-like morphology [1]. Although CT scans do not show abnormal arteries, when performing dissection, one should avoid applying cautery blindly to prevent small feeder arteries from massive bleeding during or after operations. Incisions may be ligated with sutures or linear cutters. Since the sequestered tissue is not connected to normal lung tissue, simple resection of pulmonary sequestration tissues is sufficient. In this study, the tissues of the two cases of extralobar pulmonary sequestration were attached to the diaphragm, and both possessed small feeder arteries. In one of these two cases, we applied sutures, while in the other case, a linear cutter was used.

To prevent possible hemorrhage, when treating abnormal arteries with linear cutters, some surgeons first use a vascular closure device with no cutting function to ligate the proximal end of the abnormal artery and then use a linear cutter at the distal end [2, 7] 1. However, in order to perform sutures twice, at least 20 mm of an abnormal artery needs to be isolated, which is at times clinically not possible. Other authors reported that they used a linear cutter for dissection only once on abnormal arteries with a diameter greater than 10 mm and no hemorrhage occurred 1 [8]. In this study, the abnormal artery of one patient was ligated with a vascular closure device with no hemorrhaging. Since these are all reports of single cases, we will need to accumulate more data to determine whether the single use of a vascular closure device is suitable for thicker abnormal arteries.

Endovascular embolization is safe and minimally invasive and can be used for pediatric patients with mild pulmonary infections or patients with only hemoptysis that are unable

to tolerate surgical resection. For most adult patients with pulmonary sequestration, symptoms such as coughing, expectoration, fever, or even hemoptysis could recur, if only endovascular embolization is administered. This is due to chronic infections of lung lesions. In addition, endovascular embolization may result in the rupturing of abnormal arteries [2]. In this study, one patient with lower left pulmonary sequestration coughed up 200 mL of blood and received stent-graft embolization treatment. After four days, there was recurrence of massive hemoptysis, and the patient underwent emergency thoracotomy. The abnormal artery was ligated, and the lower left lobe was resected. Eventually, the patient completely recovered and was discharged. Therefore, we believe that for pulmonary sequestration in adult patients, endovascular embolization may be used to stabilize acute massive hemoptysis; however, surgical treatment should be administered immediately when the patient is fit to receive it.

### Disclosure of conflict of interest

None.

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