# Intralobar pulmonary sequestration associated with congenital broncho-esophageal fistula

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**Pulmonary sequestration and congenital broncho-esophageal fistula are both rare malformations; nonetheless, in the presence of recurrent pulmonary infection, the possibility that one or the other may be involved should not be disregarded. To our knowledge, the concurrence of intralobar pulmonary sequestration and congenital broncho-esophageal fistula is very rare. Herein, we report a case of intralobar pulmonary sequestration combined with congenital broncho-esophageal fistula, both of which were successfully treated with surgery.**

***Key words: pulmonary sequestration, congenital broncho-esophageal fistula***

Pulmonary sequestration is a rare congenital anomaly characterized by nonfunctional lung tissue separated from the normal tracheobronchial tree and fed by an aberrant systemic artery, usually from the aorta. Pulmonary sequestration accounts for 0.15% to 6.45% of all pulmonary malformations. There are two types of pulmonary sequestration; an extralobar sequestration is separate from the normal lung and has its own visceral pleura, while an intralobar sequestration is situated within the normal lung parenchyma[1,2].

Congenital broncho-esophageal fistula is another rare congenital anomaly, which affects the respiratory and upper gastrointestinal tracts. The rarity of these cases means that their embryological origin continues to be a source of controversy[3].

The present instance is the second case report in the English-language literature of an intralobar pulmonary sequestration combined with a congenital broncho-esophageal fistula[4]. We herein report the case of a 5-month-old boy with an intralobar pulmonary sequestration and a congenital broncho-esophageal fistula concomitantly.

## Case Report

A 5-month-old boy was admitted to the Department of Pediatric Chest Disease with a history of recurrent pulmonary infection. There were no pathological findings on his physical examination. The laboratory data were within the normal limits. The posteroanterior chest roentgenogram (Fig. 1) showed subtle lucency in the medial part of the right lower zone. Subsequent computed tomography (CT) revealed the aberrant systemic artery arising from the descending aorta and supplying the right lower lung lobe. It also revealed increased lucency in the medial part of the right lower lobe, which was supplied by this aberrant artery (Fig. 2). On the basis of these findings, the patient was diagnosed with intralobar pulmonary sequestration, and the decision was made to proceed with the resection of right lower lobe via thoracotomy. On the right thoracotomy, the inferior pulmonary ligament was identified and the aberrant arterial supply stemming from the aorta toward the sequestration was noted. Passing under the lower pulmonary vein, it ascended with mild kinking and entered the right lower lobe. It was consistent with an intralobar pulmonary sequestration. We divided the inferior pulmonary ligament and then carefully dissected the aberrant artery. The artery was ligated with 2/0 silk suture from proximal side twice, from distal side once, and then

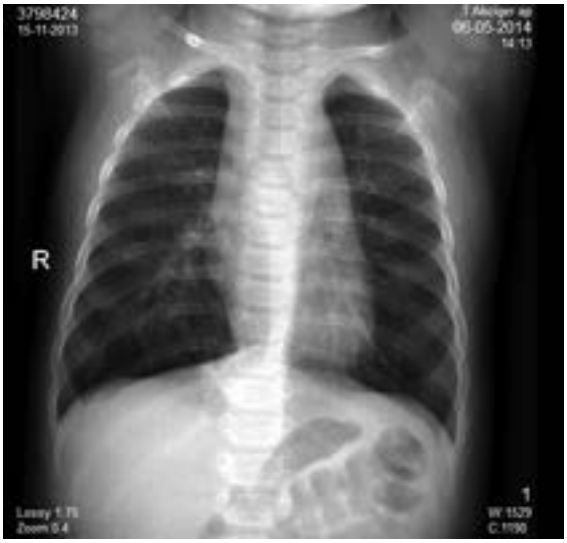


**Fig. 1.** Posteroanterior chest roentgenogram showing subtle lucency in the medial part of the right lower zone.

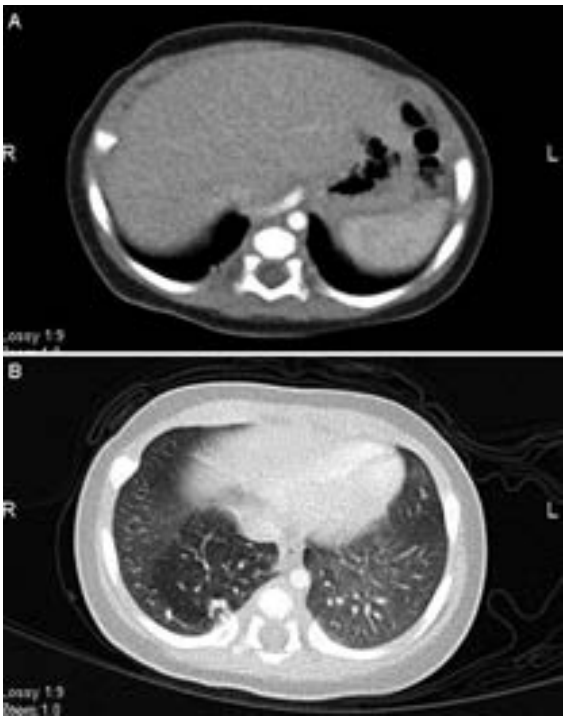


**Fig. 2.** Computed tomography showing the aberrant systemic artery arising from the descending aorta and supplying the right lower lung lobe, and increased lucency in the medial part of the right lower lobe, which is supplied by the aberrant artery.

divided. At this point, we observed a tubular component between the distal esophagus and right lower lobe, and ligated with 2/0 silk suture from the proximal side. After that, we divided this tubular component and confirmed that it was a broncho-esophageal fistula. Following ligation, we sutured the proximal side with 5/0 prolene as a second line to avoid leakage from the esophagus. Subsequently, we performed the right lower lobectomy using standard technique. The postoperative course of the patient was uneventful. On postoperative day five, we performed esophagography to determine whether there was any esophageal leakage. Having confirmed that there was no leakage, oral feeding was commenced the same day. The patient was discharged on postoperative day seven. Macroscopic

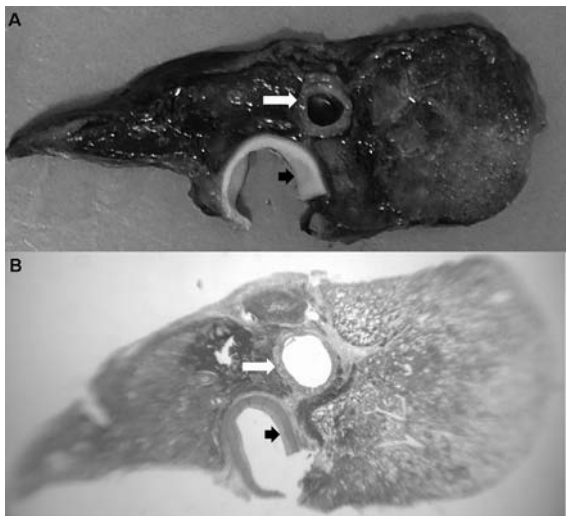


**Fig. 3.** Whole-mount section beneath the surgical margin of the specimen (A) and its corresponding histopathologic view (B). The artery (black arrow) and the bronchus (white arrow) are seen.

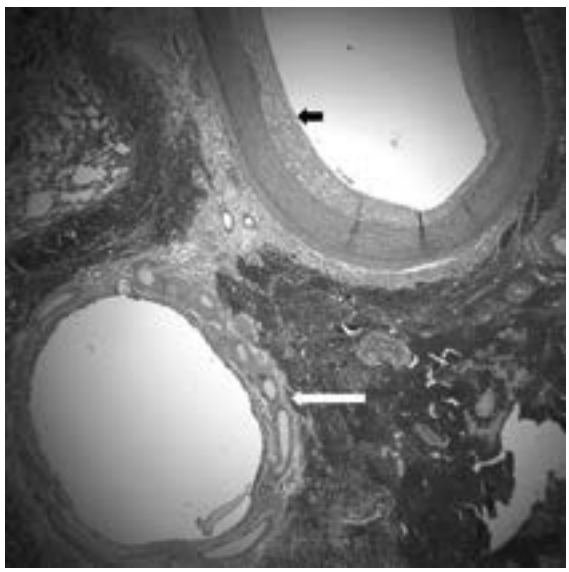

**Fig. 4.** Histological view of the ectatic artery (black arrow) and the bronchus (white arrow) in the sequestrated lung tissue.

examination of the surgical specimen confirmed the presence of the anomalous blood supply and a bronchus extending from the lung tissue (Fig. 3). Microscopic examination revealed intralobar pulmonary sequestration characterized by nonaerated pulmonary parenchyma with dilated bronchioles and ectatic branches of the penetrating artery (Fig. 4).

## Discussion

Pulmonary sequestration is a rare congenital anomaly. It constitutes 0.15% to 6.4% of congenital pulmonary anomalies and is divided into two types, based upon their pleural investment: intralobar or extralobar. Intralobar sequestration (ILS) is more common (75% of cases) and shares the same visceral pleural cavity as the native lung, whereas extralobar sequestration (ELS) (25% of cases) has its own visceral pleural investment outside the normal lung. ILS often develops a fistulous connection to the airway as the result of infection[1,2]. Pulmonary sequestration is defined as an area of lung parenchyma not connected with the tracheobronchial system and having its own aberrant systemic arterial supply, usually from the aorta, as in our case. Venous drainage is usually toward the pulmonary veins for ILS and toward the systemic venous system for ELS[2,5,6].

As in our case, most intralobar sequestrations are located in the lower lobes. Upper-lobe intralobar sequestrations are very rare, and middle-lobe intralobar sequestrations are even rarer[6]. Savic et al.[7] have reviewed a large series of sequestrations. Out of 391 intralobar sequestrations, 164 were located in the right lower lobe and 227 in the left lower lobe. Contrary to the majority of cases in Savic's review, our case had intralobar pulmonary sequestration in the right lower lobe.

The clinical presentation of pulmonary sequestrations can range from cough, hemoptysis, recurrent pneumonia and congestive heart failure to potential association with other pulmonary and skeletal anomalies, such as congenital diaphragmatic hernia and scoliosis. Symptoms arise in the first decade of life in 37% of patients, but over half become symptomatic after 20 years of age. Although chest radiographs can identify findings indicative of bronchopulmonary sequestration in most cases, CT angiography is the preferred imaging modality for identifying the anomaly and its aberrant arteries[1,2,5]. The case presented here had a history of recurrent pulmonary infection, and thus chest roentgenography and subsequent computed tomography were performed. CT of the chest revealed an aberrant arterial supply and parenchymal changes of the right lower lobe.

Congenital broncho-esophageal fistula (BEF) is another rare malformation. Congenital fistula between the bronchus/trachea and esophagus results from failed trachea-esophageal separation in the early stage of embryonic development; it is generally associated with esophageal atresia and usually presents in infancy. The rarity of this condition, with just over 100 cases reported, and its insidious clinical course sometimes make the diagnosis difficult[3,8,9]. Hanna et al.[10] reported a case of congenital BEF with total sequestration of the right lung. Pedroso et al.[11] reported a combination of extra-lobar pulmonary sequestration with congenital BEF. Gnanamuthu et al.[4] reported the first case report of an intralobar pulmonary sequestration combined with congenital BEF. Our case is the second report in the English-language literature of an intralobar pulmonary sequestration combined with congenital BEF.

If not suspected and left untreated, congenital BEF may lead to fatal complications despite the benign nature of this anomaly. Congenital BEF may cause symptoms in childhood, or may not appear until adult life. It is typically associated with repeated and persistent infections, which can lead to development of bronchiectasis[8,9]. Although the present case had a history of recurrent pulmonary infection, congenital BEF was diagnosed incidentally during surgical resection of pulmonary sequestration. Since our case had two etiologic factors for recurrent pulmonary infection, it is not possible to say which one was responsible.

Despite the benign nature of pulmonary sequestration, its potential complications are serious and may include recurrent pulmonary infection, hemoptysis, congestive heart failure and tumorigenesis. Therefore, the main modality of treatment has always been surgical resection, even for asymptomatic patients with pulmonary sequestrations. The conventional surgical approach for the resection of pulmonary sequestrations is via

a posterolateral thoracotomy. The number and position of aberrant arteries supplying the resection should be carefully noted, as inadvertent injury could cause massive hemorrhage[1,12,13]. Intralobar sequestrations usually require lobectomy, as was performed in our case. Video-assisted thoracic surgery (VATS) offers a less invasive alternative to conventional thoracotomy for sequestration resection. However, factors like dense adhesions can obviously make VATS more difficult to perform, and in these cases thoracotomy may also be required. Moreover, the VATS procedure should be encouraged for use by experienced surgeons only because of the potential risk of life-threatening vascular injury[12,13]. Despite the successful results of surgical treatment in pulmonary sequestrations, novel interventional methods such as transcatheter coil embolization have been identified in recent years[14,15,16]. The authors reported that transcatheter coil embolization was a safe and effective treatment option for pulmonary sequestrations.

As in the case of pulmonary sequestration, patients with congenital BEF should be treated surgically as soon as the diagnosis is established. Delay in treatment may cause serious pulmonary complications and even death[9]. In our case, we chose to perform a lower lobectomy via right thoracotomy, as we did not have sufficient experience or equipment for pediatric thoracoscopic lobar resection. In addition to surgical resection of the sequestrated lower lobe, we were able to treat the incidentally diagnosed congenital BEF at the same time.

In conclusion, even though pulmonary sequestration and congenital BEF are rare malformations, their possible involvement should not be disregarded in the presence of recurrent pulmonary infection. Herein we have reported an extremely rare case of intralobar pulmonary sequestration combined with congenital BEF, both of which were successfully treated with surgery.

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