Extralobar pulmonary sequestration: The importance of intraoperative vigilance

Erin G. Brown*, Clifford Marr, Diana L. Farmer

University of California, Davis, Department of Surgery, 2315 Stockton Blvd., OP512, Sacramento, CA 95817, USA

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Pulmonary sequestration is a rare congenital malformation in which a segment of nonfunctional lung lacks a bronchial connection and possesses an aberrant blood supply. Most often, blood supply is from the infradiaphragmatic aorta. Typically, preoperative evaluation via computed tomography (CT) with contrast provides adequate information regarding vascular supply; however, the unique presence of three separate arterial branches was not identified in this case.

1. Case report

A five-month-old baby boy was referred to our clinic for evaluation of a prenatally diagnosed left lower lobe pulmonary sequestration. This was diagnosed via prenatal ultrasound and serially monitored during the course of pregnancy. Postnatal CT scans demonstrated a 3.2 × 2.7 × 3.6 cm mass of opacified lung parenchyma consistent with extralobar sequestration (ELS) versus possible congenital pulmonary airway malformation. A large, single systemic arterial branch to the lesion was identified (Fig. 1). The venous drainage was felt to be into the hemiazygos vein.

The decision was made to proceed with thoracoscopic resection of the lesion. Intraoperatively, the patient was indeed found to have an extralobar sequestration with a well-demarcated parenchymal connection to the normal left lower lobe. However, instead of finding one systemic feeding vessel, three systemic arteries directly from the aorta to the lung were identified (Fig. 2). One vein was identified. We were able to remove the sequestration thoracoscopically with preservation of the lower lobe. Postoperatively, the patient did well. He was discharged home on postoperative day number four after successful chest tube removal.

2. Discussion

Pulmonary sequestration is a rare malformation with an estimated incidence of 0.15–6.4% [1]. Defined as the presence of nonfunctional lung tissue that is lacking a connection to the bronchus and possessing an aberrant arterial blood supply, over 40 hypotheses have been suggested to explain the origin of the defect with the most favored etiology being the development of an accessory lung bud between weeks 4 and 8 of gestation [2–5]. Pulmonary sequestrations can be either intralobar or extralobar. Intralobar sequestration (ILS) is abnormal lung tissue surrounded by normal lung pleura, while extralobar sequestration has its own separate pleura.

Extralobar sequestration is less common than its intralobar counterpart and comprises approximately 25% of all sequestrations. They are predominantly featured in males (4:1), left-sided (65%), and are often associated with other congenital malformations (65%) [3]. The most common associated anomaly is a diaphragmatic hernia (16%), but they may also be associated with other lung malformations such as congenital cystic adenomatoid malformation (CCAM) and bronchogenic cysts, pectus excavatum, pericardial defects, and enteric duplication cysts among others. Anomalous arterial variants are also extremely common. Most often, arterial

* Corresponding author. Tel.: +1 916 734 2724; fax: +1 916 734 5633. E-mail address: erin.brown@ucdm.ucdavis.edu (E.G. Brown).

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blood supply is derived from the thoracic aorta (46%), but it has also been described from the abdominal aorta, pulmonary artery, intercostal, left gastric, or even subclavian arteries [4,6]. Furthermore, the presence of dual arterial supply has been reported in 5% of cases and rare reports of three or more vessels have been recounted. In the same review, 3.7% of ELS demonstrated a connection with the lower lobe.

ELS most often presents in neonates prior to 6 months of age [3]. However, numerous reports of ELS in adults have been reported and should be considered in the differential diagnosis at any age. With more widespread use of prenatal ultrasound, increasing numbers of ELS are being diagnosed prenatally and can be seen on ultrasound as early as 16 weeks gestation. Typical presentation includes cough, respiratory distress, feeding difficulties, or congestive heart failure, although many are asymptomatic.

For diagnosis of ELS, CT with contrast is the preferred imaging modality [7]. Traditionally, formal arteriography was recommended, yet with significant improvements in CT technology, this mode of invasive imaging is rarely indicated. Interestingly, in this case, the preoperative CT scan failed to identify three separate arterial branches supplying the ELS. Given the high rates of anomalous arterial vessels, preoperative imaging is important, but constant vigilance during surgery for aberrant vessels is crucial. Although several cases of severe hemorrhage and/or death have been described during resection of pulmonary sequestrations, none have been reported in recent years [4,6].

While recurrent infection is a common complication of intralobar sequestration, this is not the case with ELS. This leads some to advocate conservative management in asymptomatic patients as a viable option. However, often the diagnosis of ELS is not 100% certain based on imaging, and there is a high rate of synchronous malformations such as CCAM (which possess a possibility of malignant transformation). Therefore, most surgeons would recommend resection [3,4,8]. Traditional gold standard therapy was resection of ELS via mini-thoracotomy. As minimally invasive surgery techniques continue to improve, this has become an increasingly accepted alternative [3,7–9]. Several studies have demonstrated the safety of thorascopic surgery in infants within the first few months of life without increased morbidity or mortality [8–10]. While ILS often requires lobectomy, ELS can usually be resected without removing adjacent normal lung parenchyma. One new alternative to surgery is coil embolization. This minimally invasive procedure has been safely performed for pulmonary sequestration and has also been shown to successfully result in complete or near-complete regression of the lesion within a few months of the procedure [11–13].

3. Conclusion

Extralobar sequestration is a rare congenital malformation. Although ELS is a benign disease, preoperative imaging is unable to differentiate this condition with 100% certainty from other diagnoses among the differential with the potential for malignant transformation. Although coil embolization presents a possible alternative to surgical intervention, long-term follow up studies are not yet available. Considering these factors, we continue to favor surgical resection. Given the high percentages of anatomic variants, preoperative evaluation via contrast-enhanced CT remains the gold standard for investigation of the anatomy prior to proceeding with surgical resection. However, this case demonstrates failure to identify the unique presence of three separate arterial branches on preoperative CT and stresses the importance of maintaining strict intraoperative vigilance as the consequences of missed anomalies is potentially fatal.

Consent

Written informed consent was obtained from the patient’s guardian for publication of this case report and accompanying
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