Extralobar pulmonary sequestration

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■ A neonate presenting with a density of the right mid-thorax was found, at thoracotomy, to have an extralobar pulmonary sequestration. The sequestration was resected, and the infant made an uneventful recovery. The morphogenesis, diagnosis, and management of this uncommon lesion are discussed.
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PULMONARY SEQUESTRATION is an anomaly in which a segment of lung lacks a normal connection with the tracheobronchial tree and receives its blood supply from the systemic rather than pulmonary circulation. Of the two types of sequestrations, the intralobar type is more common, occurring from three to six times as frequently as the extralobar variety.[1,2] The latter is distinguished by its complete pleural investment and by the venous drainage, which is usually systemic rather than pulmonary.

Extralobar pulmonary sequestrations occur on the left side in up to 90% of cases.[3] These lesions may be asymptomatic in the neonatal period in the absence of associated anomalies. This report describes an extralobar bronchopulmonary sequestration presenting shortly after birth.

CASE REPORT

A white male neonate was born at 38 weeks gestation by normal vaginal delivery after an uncomplicated pregnancy. Following artificial rupture of membranes, the amniotic fluid was found to be meconium stained, and a small amount of meconium was aspirated from the oropharynx during delivery. A single nuchal cord was present. Apgar scores were 6 and 8 at one and five minutes, respectively. After suctioning and mask ventilation, the infant was placed in 40% oxygen. His blood gases were satisfactory. Empiric antibiotic therapy was initiated due to a history of maternal fever prior to delivery. His respiratory status readily improved, and he was rapidly weaned to room air.

A chest radiograph obtained soon after delivery revealed a density projecting over the right posterior midlung field, appearing to arise posteriorly with a well-defined anterior margin (*Figure 1*). After his condition was stabilized, the infant was transferred to The Children's Medical Center in Dayton, Ohio. On admission, physical and laboratory findings were unremarkable. Repeat chest radiographs showed no change from the previously obtained images. Urinary vanillylmandelic-acid determinations were normal.

Computed tomography (CT) was performed, and images showed a soft-tissue density extending from the level of the carina to the 10th thoracic vertebra and from the right lateral chest wall to the mediastinum (*Figure 2*). No calcification was present within the lesion, and there was variable contrast enhancement. Ultrasound confirmed the solid, homogeneous nature of the mass and demonstrated arterial vascularity centrally.

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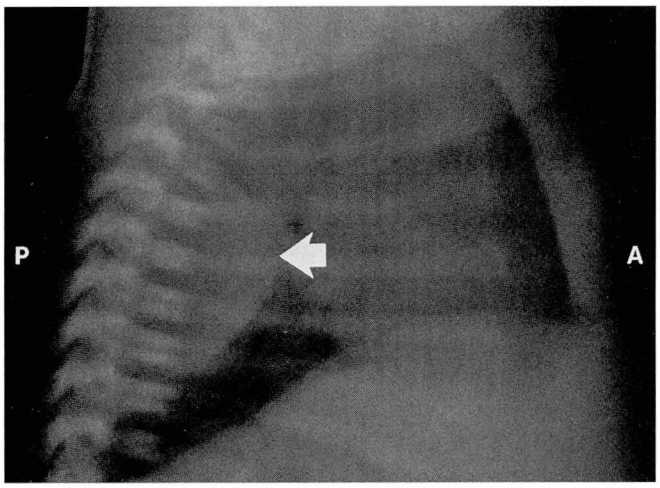

FIGURE 1. Lateral chest radiograph shows density (arrow) effacing the posterior mid-lung field.

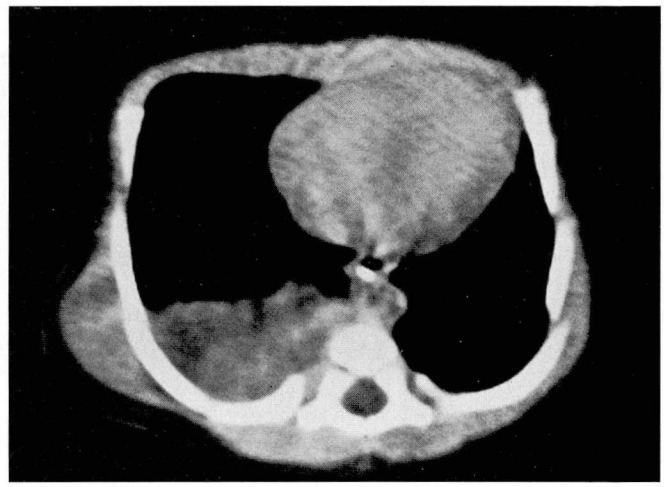

FIGURE 2. CT scan shows a mass in the right posterior hemithorax.

Intraspinal extension was not evident. No displacement or fistulous involvement of the esophagus was seen on an esophogram.

The lesion was thought to be a neuroblastoma or ganglioneuroma, although an atypical pulmonary sequestration was considered. Angiography was deferred. A right thoracotomy was performed and revealed a well-circumscribed mass of the posterior hemithorax. The lesion had the appearance of normal lung tissue and was supplied by a small vascular pedicle arising from the intercostal vessels adjacent to the azygous vein. Resection required only simple ligation of the vascular pedicle. Pathological examination was consistent with an extralobar pulmonary sequestration. The postoperative course was uneventful, and the infant was discharged on the third postoperative day.

DISCUSSION

Although now recognized more frequently, pulmonary sequestration remains an uncommon entity. In a collective review of 540 sequestrations, 133 of the extralobar type were reported.[1] Stocker and Kagan-Hallet[4] found only two extralobar sequestrations in 306,952 patients admitted to Denver Children's Hospital over a 30-year period. The true incidence is likely to be higher than these figures suggest as the sequestration is often asymptomatic.

It has become increasingly apparent that, although traditionally categorized into intralobar or extralobar types, these lesions are part of a spectrum of congenital bronchopulmonary malformations that can, in part, be explained by derangements during embryologic development. This concept was introduced by Sade et al in 1974.[5] Abnormalities of the pulmonary parenchyma, pleura, arterial supply, venous drainage, bronchial connections, and vestigial communications with the gastrointestinal tract may occur singly or in combination.

Insight into the morphogenesis of sequestrations can be gained by evaluating associated anomalies. Pulmonary sequestrations may retain rudimentary or patent fistulous communications with the gastrointestinal tract, usually involving the stomach or esophagus. Gerle et al[6] presented three such cases and suggested the term "congenital bronchopulmonary foregut malformations" to describe these lesions. In a later review of 29 patients with similar anomalies, Heithoff et al,[7] presenting a unifying concept, postulated that a supernumerary lung bud arises posterior to the normal lung bud at five to seven weeks of gestation and migrates along with the growing esophagus. Usually the communication between the sequestered lung and gastrointestinal tract involutes. If this involution is incomplete, a fibrous strand may remain or a patent fistulous tract may persist. According to this concept, the presence of a pleural covering (extralobar sequestration) or lack of it (intralobar sequestration) depends simply on the time of formation of the abnormal lung bud relative to formation of the pleural investments. This theory is supported by the finding of coexisting intralobar and extralobar sequestrations.[8] The evidence for a common etiology for

both types of pulmonary sequestrations is compelling but remains conjectural.

Over half of patients with extralobar sequestrations may have one or more associated anomalies. The most commonly reported of these is diaphragmatic hernia. The incidence varies from 15% to 30%[9] to as high as 58% in one series.[3] The generally accepted explanation for this association involves mechanical interference with closure of the pleuroperitoneal canal by the abnormal position of the forming sequestration.[10] The sequestration may, in rare cases, be subdiaphragmatic or even intradiaphragmatic.

Just as associated diaphragmatic hernias are predominantly left-sided, there is a preponderance of left-sided lesions of both the intralobar and extralobar types. In a large series, Savic et al[1] reported 78.9% of extralobar sequestrations were located on the left. Several authors quote a two-to-one predominance.[9,11] This laterality has been attributed to the greater likelihood of survival of a developing sequestration that receives its vascular supply from the left dorsal aortic arch. Another explanation implicates the later closure of the left hemidiaphragm.[9]

Several other associated anomalies have been reported with some frequency. Bronchogenic cysts coexist in 10% to 15% of children with extralobar sequestrations.[11] Pericardial defects occur in up to 25%.[1,11,12] In some series, sternal deformities have been frequently associated, predominantly the excavatum type.[11–13] Other findings include tracheoesophageal fistulas, gastric duplications, and abnormalities of the heart and great vessels.

The incidence of clinical manifestations of extralobar pulmonary sequestrations varies widely and may reflect associated abnormalities and the population studied. Diagnosis has been made involving a fetus at 24 weeks with hydrops fetalis.[14] The association between maternal hydramnios and extralobar sequestrations has also been reported.[4] Infants with sequestrations of sufficient size may present with respiratory distress[15]; associated pulmonary hypoplasia or diaphragmatic hernia may also be evident. However, as in the case presented here, symptoms may occur in the absence of associated anomalies. In a large pathologic series of extralobar sequestrations, 12 of 15 patients presented at birth with either absent spontaneous respirations or neonatal respiratory distress.[4] All of the infants died within 27 hours of delivery, most often due to associated massive hydrothorax, diaphragmatic hernia, or hyaline membrane disease.

Of those patients manifesting no symptoms in the neonatal period, some will present with feeding difficulties or various cardiovascular and respiratory symptoms during infancy and childhood. Leijala and Louhimo[12] determined a mean age at diagnosis of 3.6 years in 16 patients. These children usually presented with dyspnea, intermittent cyanosis, cough, and asthmatic symptoms. Unlike its intralobar counterpart, patients with extralobar sequestrations only rarely develop recurrent pulmonary infections in the absence of gastrointestinal communications.[2] Congestive heart failure can occur as a result of arteriovenous shunting through the sequestration.

The remaining group of patients may remain asymptomatic for many years. The lesion may be discovered at surgery or on postmortem examination. Not infrequently, an extralobar pulmonary sequestration is first demonstrated on routine chest radiographs or in the course of evaluation for respiratory symptoms. Typically, it appears as a triangular or pyramidal density adjacent to the costophrenic sulcus and mediastinum.[2]

Ultrasound studies have detected extralobar sequestrations in both intrathoracic[14] and extrathoracic locations in utero.[16] The ultrasonic appearance of the sequestration is nonspecific, showing either a cystic or homogeneous density. The diagnosis, however, is suggested by the demonstration of a vascular supply originating from the aorta or other systemic vessel.[17] Findings on CT scans are similarly nonspecific, showing only an intrathoracic or extrathoracic mass.[18] Recently, magnetic resonance imaging has been employed to diagnose pulmonary sequestration preoperatively.[19]

Arteriography, using either conventional or digital subtraction techniques, has generally been considered the most definitive study for the diagnosis of extralobar sequestrations. The demonstration of abnormal feeding vessels, while suggestive, is not necessarily pathognomonic of a sequestration. Both neoplastic and inflammatory lesions of the lung have been misdiagnosed as pulmonary sequestrations based on the arteriographic appearance of a systemic vascular supply.

Since the diagnosis of pulmonary sequestration cannot be made with certainty without obtaining tissue for histologic diagnosis, many authors recommend excision of the lesion at the time of presentation.[3,9,12] Collin et al[20] cite the rare cases of malignant degeneration of congenital cystic lesions of the lung as further justification for operation. Resection of extralobar sequestrations can be accomplished with minimal morbidity. Buntain et al[13] reported no significant complications and an average hospital stay of 10 days for nine children who underwent surgical resection. The infant presented in this report was discharged from the hospital three days after surgery.

Technically, resection of the sequestration presents little challenge as the lesion is separate from the normal lung and usually requires only ligation of the vascular supply. These vessels are often elastic rather than muscular and not infrequently arise from below the diaphragm, traversing the inferior pulmonary ligament. Awareness of the abnormal blood supply is, therefore, essential to prevent inadvertent and potentially fatal transection of an anomalous vessel.

An alternative approach, advocated by some, is to observe these lesions and intervene surgically if complications develop.[2,21] This approach assumes a high degree of diagnostic certainty based on clinical and radiographic findings.

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