Extralobar pulmonary sequestration

Unusual presentation and systemic vascular communication in association with a right-sided diaphragmatic hernia

A 17-day-old infant with a right-sided extralobar pulmonary sequestration associated with an intact diaphragmatic hernia sac presented as an acute respiratory emergency. The diagnosis was obscured by the initial resuscitative treatment of cardiorespiratory arrest necessitating positive-pressure ventilation complicated by bilateral tension pneumothoraces prior to transfer. At operation, unique systemic arterial and venous communications to the sequestration from the dome of the liver through the intact hernia sac were found. A bronchial communication to the right main bronchus was also present. Histologic examination of the sequestration specimen revealed other features common to both intralobar and extralobar sequestration. This case lends support to the hypothesis of a common embryologic basis for both types of sequestration, as well as other congenital pulmonary cystic abnormalities, and the association with diaphragmatic hernia.

Timothy G. Canty, M.D., San Diego, Calif.

Extralobar pulmonary sequestration is an uncommon but well-described congenital anomaly. As distinct from the more common intralobar pulmonary sequestration, both the arterial supply and venous drainage are systemic vessels, frequently multiple, which can arise either above or below the diaphragm. Associated congenital anomalies, especially diaphragmatic hernia, other diaphragmatic defects, and, less frequently, cardiopulmonary abnormalities and foregut communications, are far more common with extralobar sequestration than with the intralobar type.1–8 Although there are other distinguishing features between extralobar and intralobar sequestration, some authors consider both forms to have a common embryologic basis along with other congenital pulmonary cystic lesions and foregut anomalies. A common embryologic defect at the time of thoracic organogenesis is a plausible explanation.1–3, 5, 6, 8–10

This case is an unusual instance of right-sided extralobar pulmonary sequestration in association with a diaphragmatic hernia. The initial clinical presentation was that of an acute respiratory emergency. The systemic vascular communications arose from the dome of the liver through the diaphragmatic hernia sac, and the anatomy and histopathology of the sequestration exhibited features common to both intralobar and extralobar types.

Case report

A 17-day-old infant was transferred following resuscitation from a cardiorespiratory arrest occurring in the waiting room of a nearby clinic. The child, weighing 6 pounds, 14 ounces, was born of a normal term delivery and was discharged from the hospital as a normal neonate at 1 day of age. The first 2 weeks of life were uneventful until 4 days prior to admission, when the infant began to vomit intermittently. On the day of admission vomiting increased with accompanying lethargy and diminished urine output, prompting a visit to a local clinic. While drinking water in the waiting room, the infant suddenly gagged and had a cardiorespiratory arrest. Immediate resuscitation with endotracheal intubation, ventilatory support, and intravenous fluids was instituted and the baby was transported by intensive care van.

Physical examination revealed a well-developed 7 pound cyanotic female infant on a ventilator with a blood pressure of 40 mm Hg and a pulse rate of 150 beats/min. There were deep sternal retractions and diminished breath sounds with diffuse rales bilaterally. Liver and spleen were both palpable below the costal margins. Neurologic examination revealed good spontaneous and appropriate movements, sucking, and grasping. The remainder of the examination showed no abnormalities.

From the Department of Surgery, Division of Pediatric Surgery, University of California, San Diego, Calif.

Received for publication April 28, 1980.

Accepted for publication May 21, 1980.

Address for reprints: Timothy G. Canty, M.D., Head, Division of Pediatric Surgery, University Hospital, 225 Dickinson St., San Diego, Calif. 92103.
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Admission hemogram showed a white blood cell count of 19,000/mm$^3$ and a hematocrit value of 42%. Arterial blood gases were as follows: $P_O_2$ 60 mm Hg, $P_CO_2$ 48 mm Hg, and pH 7.18 on 60% oxygen. The admission chest x-ray film showed bilateral tension pneumothoraces (Fig. 1, A), which were immediately treated by insertion of chest tubes bilaterally. Several midline thoracic vertebral defects were also noted.

The child’s respiratory status dramatically improved over the ensuing few hours, and a roentgenogram after chest tube insertion suggested cystic lesions in the right lower lung field (Fig. 1, B). During the following 24 hours the infant’s condition stabilized considerably and she was eventually weaned from the ventilator and extubated. Follow-up chest x-ray films after extubation suggested the presence of loops of bowel just above the diaphragm in addition to the previously seen cysts in the lower lung field (Fig. 1, C). The child remained in stable condition breathing room air. Four hours after the previous chest film, the right chest tube became dislodged and a repeat chest roentgenogram revealed the right hemithorax to be almost completely filled with loops of intestine (Fig. 1, D).

With a preoperative diagnosis of right-sided diaphragmatic hernia and congenital cystic disease of the right lung, the infant was taken to operation. At right thoracotomy, the lower pleural cavity was obliterated by inflamed and adherent cystic lung tissue, also adherent to a large posterolateral diaphragmatic hernia sac containing the right lobe of the liver and multiple loops of bowel. Further dissection revealed the cystic lower lobe to be an extralobar sequestration deriving its arterial supply and venous drainage via multiple large arteries and veins from the dome of the liver through the hernia sac (Fig. 2). A small branch of the right main bronchus communicated with the sequestered mass. The right upper lobe appeared normal and uninvolved with the inflammatory process, with good expansion on ventilation. The sequestered lobe was excised by individually taking the vascular supply and finally the bronchial connection. The contents of the diaphragmatic hernia were reduced into the abdomen, the excess sac was excised, and the rims of normal diaphragmatic tissue were reapproximated.

The child recovered uneventfully and was discharged from the hospital on the eighth postoperative day. Numerous follow-up examinations through 9 months of age have shown the child to be asymptomatic and gaining weight normally.

Histologic examination of the sequestered lung tissue...
showed large numbers of cystic spaces lined by respiratory epithelium, some with cartilage in their walls, and poorly formed alveolae. The bronchial stump ended in several small tertiary bronchioles with no true alveolar connection.

**Comment.** The acute presentation of this patient with vomiting, aspiration, and cardiorespiratory arrest was most likely secondary to the partial intestinal obstruction frequently seen with otherwise asymptomatic diaphragmatic hernia. The vigorous resuscitation was complicated by bilateral tension pneumothoraces. The additional requirement for positive-pressure ventilatory support during the early postresuscitation period masked the presence of the diaphragmatic hernia by keeping the contents reduced into the abdomen. It was only when the youngster was weaned from the ventilator and the chest tube became inadvertently dislodged that the diaphragmatic hernia became evident.

**Discussion**

Extralobar pulmonary sequestration is a relatively rare congenital anomaly. Intralobar sequestration is four to five times more common, and both types of sequestration are four to six times more common on the left side. Extralobar sequestration is associated with diaphragmatic hernia in 30% to 50% of cases. Of interest, the diaphragmatic hernia associated with sequestration is more likely to have an intact sac than is an isolated diaphragmatic hernia. Associated anomalies of the vertebrae, chest wall, and foregut are more frequently seen with extralobar sequestration.

Several theories of origin of both types of sequestration have been proposed. Some advocate a separate embryologic basis for each type, and others favor a common explanation for both, as well as for other cystic pulmonary lesions and foregut anomalies. There are several recent comprehensive reviews of this material which are beyond the scope of this paper.

The systemic arterial supply and venous drainage of the extralobar sequestration is supradiaphragmatic in 80% to 90% of cases. Infradiaphragmatic vascular communications, however, have been thoroughly described and are more common when the sequestration is associated with diaphragmatic hernia. Infradiaphragmatic arterial supply from the abdominal aorta and the splenic, celiac, and gastric arteries, occurring alone or in combination, has been documented. Arterial supply and drainage directly from the dome of the liver through an adherent intact diaphragmatic hernia sac, as seen in this case, have not been previously described.

Bronchial communication is common with intralobar sequestration but rare with the extralobar type. These communications are abnormal morphologically and histopathologically and rarely communicate with true alveolar spaces. Bronchus-like communications to the gastrointestinal tract, usually the esophagus, are seen more commonly with extralobar sequestration.

Since extralobar pulmonary sequestration is associated with a variety of altered anatomic relationships, clinical manifestations are also varied. Typically, the extralobar sequestration without a communication to the tracheobronchial tree or gastrointestinal tract goes unrecognized for years or may be noted as an unexplained supradiaphragmatic mass on a routine chest x-ray film. When bronchial communication or gastrointestinal tract communication is present, recurrent pulmonary infections or gastrointestinal symptoms are common presenting complaints. Congestive heart failure from a left-to-right shunt has also been documented as a mode of presentation.

Extralobar sequestrations rarely cause difficulty in
the newborn period. Acute respiratory distress secondary to the mass effect of the sequestration crowding the mediastinum, sequestration of the entire lung, or as the result of massive hemorrhage into a sequestration has been described. In those neonates in whom the sequestration is associated with a diaphragmatic hernia, it is usually discovered as an incidental finding at the time of operative repair of the hernia. Acute respiratory distress secondary to the inadvertent injury of the anomalous vessels.

The diagnosis of extralobar pulmonary sequestration is frequently suggested by a routine chest x-ray film showing the presence of a supradiaphragmatic intrathoracic mass. Bronchoscopy, bronchography, and tomography all have been advocated and utilized but probably are not necessary. Aortography and selective arteriography will give the definitive diagnosis by demonstrating the abnormal vascular communications. Whether this somewhat hazardous procedure is indicated in all cases is questionable, but it may provide helpful information to the surgeon in regard to the origin of the abnormal vessels, especially those that communicate below the diaphragm. Some authors advocate routine angiography in all cases to avoid an intraoperative catastrophe secondary to the inadvertent injury of the anomalous vessels.

The pulmonary ventilation-perfusion scan offers a relatively noninvasive means of demonstrating an area in the lung field with no pulmonary vascular supply or bronchial communication. This procedure can be used as a screening technique to better select patients for arteriography.

Treatment of symptomatic sequestrations, or those found incidentally at the time of operation for other congenital anomalies, is simple excision. Expectant treatment of asymptomatic extralobar sequestrations is advocated by some authors, although most favor resection. Infection, rupture, or hemorrhage into the degenerative cystic lung tissue may well convert an asymptomatic state to a life-threatening situation.

This case is unusual in several respects, both clinically and anatomically. The chief presentation of vomiting, aspiration, and respiratory arrest requiring vigorous resuscitation masked the diaphragmatic hernia. This is a noteworthy side effect of both tension pneumothorax and prolonged positive-pressure ventilation. The true spectrum of disease did not become evident until the time of definitive operation. Bronchial communication, microcystic and macrocystic lung lesions, and the unique vascular communications in this case are characteristics of both extralobar and intralobar sequestration. These histopathological features support the idea of a common embryologic basis for the two types of sequestration as well as other cystic lung lesions. The interference of proper diaphragmatic formation as the result of extralobar sequestration with infradiaphragmatic vascular supply is also suggested by this case. Treatment of this youngster was relatively straightforward once the diagnosis became evident. All that was required was simple excision of the sequestrated right lower lobe and repair of the diaphragmatic hernia. The child is presently thriving at 9 months of age.

REFERENCES