CONGENITAL LUNG MALFORMATIONS, UPDATE AND TREATMENT

STEVEN ROTHENBERG MD.(1)

1. Department of Pediatrics. The Rocky Mountain Hospital For Children. Hospital for Children, Denver, Colorado, USA.
steverberg@aol.com

SUMMARY

There is a broad spectrum of bronchopulmonary malformations which present in early infancy and childhood. These include Bronchogenic cysts, Bronchopulmonary sequestrations, congenital cystic adenomatoid malformation (CCAM), and congenital lobar emphysema. These lesions maybe detected by prenatal diagnosis, present as acute respiratory distress in the newborn period, or may remain undiagnosed and asymptomatic until late in life.

Pulmonary sequestration (PS) is a rare congenital malformation of the lower respiratory tract. It consists of a non-functioning mass of lung tissue that lacks normal communication with the tracheobronchial tree and receives its' arterial blood supply from the systemic circulation. CCAMs are extremely rare with the reported incidence being between 1/25,000 and 1/35,000 . The pathogenesis is uncertain but appears to result from a abnormality of the branching morphogenesis of the lung and represents a maturation defect. Congenital Lobar Emphysema (CLE) is a rare anomaly of lung development which often presents in the neonatal period with hyperinflation of one or more pulmonary lobes.

For the last decade we have preferred to use minimally invasive techniques to perform lobectomies in all of these infants with congenital cystic lesions. The benefits of avoiding a formal thoracotomy and the morbidity associated with it greatly out way the disadvantages of the increased technical difficulty and operative time.

Key words: Pulmonary malformations, bronchogenic cysts, sequestrations, congenital cystic adenomatoid malformation, lobar emphysema, thoracoscopy, minimally invasive surgery

INTRODUCCION

There is a broad spectrum of bronchopulmonary malformations which present in early infancy and childhood. These include Bronchogenic cysts, Bronchopulmonary sequestrations, congenital cystic adenomatoid malformation, and congenital lobar emphysema. These lesions maybe detected by prenatal diagnosis, present as acute respiratory distress in the newborn period, or may remain undiagnosed and asymptomatic until late in life. Treatment may vary somewhat depending on the time of diagnosis, the presentation, but in most cases complete resection is the desired therapy. New minimally invasive techniques now allow this to be done with much less pain and morbidity and long term consequence for the infant or child. Each of these main categories will be discussed.

Pulmonary sequestration (PS) is a rare congenital malformation of the lower respiratory tract. It consists of a non-functioning mass of lung tissue that lacks normal communication with the tracheobronchial tree and receives its' arterial blood supply from the systemic circulation (1). The majority of sequestrations fall into 2 categories, intralobar (ILS) and extralobar (ELS). ILS is defined as a lesion which lies within the normal lobe and lacks its' own visceral pleura. ELS is a mass located outside the normal lung and has its' own visceral pleura. There is a third, rarer

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variant, which is a bronchopulmonary foregut malformation (BPFM) in which the abnormal lung is attached to the gastrointestinal tract (2).

The exact embryologic basis for the development of PS of the lower respiratory tract is unclear. The lesion likely occurs early in embryologic development prior to the separation of the aortic and pulmonary circulations (3). One explanation is that there is an abnormality lung bud formation (4, 5, 6). This might result in not only PS but also Congenital Cyst Adenomatoid Malformation (CCAM), bronchogenic cyst, foregut duplication, or even congenital lobar emphysema. Another explanation is that a portion of the developing lung bud is mechanically separated from the rest of the lung by compression from cardiovascular structures, traction by aberrant systemic vessels, or inadequate pulmonary blood flow.

Whatever the etiology, it is clear that there is some spectrum which exists between PS, CCAM, and other bronchopulmonary malformations, and all need to be considered in the diagnosis when one of these lesions is encountered.

The majority of sequestrations occur in the lower lobes, but they can occur anywhere in the chest as well as below the diaphragm and in the retroperitoneal space (7). Intra-lobar sequestration is far more common then ELS. Approximately 13% of ELS arelocated below the diaphragm. Approximately 60 % of ILSs involve the left lower lobe, the majority in the posterior basal segment (8). BPFM is more common on the right side. The incidence of a communication with the foregut is much higher with ELS then IS. The vascular supply for both ILS and ELS generally arises from the lower thoracic or upper abdominal aorta. The majority of cases have a single arterial feeder but up to a third may have multiple vessels. The venous drainage is usually normal to the left atrium but abnormal drainage to the right atrium, vena cava, and Azygus have been documented. Both ELS and ILS have been documented in the same patient.

Congenital Cystic Adenomatoid Malformation (CCAM) is a rare developmental anomaly of the lower respiratory tract. The term encompasses a spectrum of conditions, the origins of which remain debatable. Affected patients may be completely asymptomatic or present with severe respiratory distress in the newborn period. Others become symptomatic later in life with acute respiratory distress, acute infection, or other manifestations. Many cases which previously would have gone on undetected until complications arose later in life are now detected by routine prenatal ultrasound. Thus the pediatric surgeons' role has changed from simply dealing with a patient with acute respiratory issues to often providing prenatal consultation for the expectant parents.

CCAMs are extremely rare with the reported incidence being between 1/25,000 and 1/35,000 (9). The pathogenesis is uncertain but appears to result from a abnormality of the branching morphogenesis of the lung and represents a maturation defect. The different types of CCAMS are thought to originate from different levels of the tracheobronchial tree and at different stages of lung development. CCAM is distinguished from other lesions and normal lung by five main criteria. These include polypoid projections of the mucosa, an increase in smooth muscle and elastic tissue within the cyst walls, an absence of cartilage in the cystic parenchyma, the presence of mucous secreting cells, and the absence of inflammation. While the CCAM portion of the lung does not participate in normal gas exchange there are connections to the tracheobronchial tree which can lead to air-trapping, and respiratory distress in the newborn period.

The exact mechanism resulting in CCAM is unknown but is thought to include an imbalance between cell proliferation and apoptosis. (increased cell proliferation and decreased apoptosis as compared to gestational controls). CCAMs are hamartomatous lesions that are comprised of both cystic and adenomatous overgrowth of the terminal bronchioles. If large cysts develop in utero it can compress and compromise the growth of normal surrounding tissue.

CCAMs can affected all lobes and are equally distributed between the right and left side. They can affect more then one lobe and be bilateral but this extremely rare (10, 11). CCAMs are connected to the tracheobronchial tree although the connecting bronchi are generally not normal. The blood supply comes from the normal pulmonary vasculature as opposed to sequestration which receives its' arterial supply for the systemic arterial system.

The classification system currently comprises 5 types based on the size of the cyst and the cellular characteristics (4). The initial classification included Type 1, which include large cysts comprised of primarily bronchial cell type characteristics; Type 2, intermediate cysts with bronchiolar type cells; and Type 3, small cysts with bronchiolar/alveolar duct cells. Types 0 and 4 were added later and were based on the site of origin of the malformation.

The presentation of CCAM is quite variable and can extend from the early prenatal period to late in adult life. The spectrum runs from an incidental finding on a routine chest xray, in a completely asymptomatic patient, to severe respiratory distress in the newborn period.

More and more of these lesions are now picked up in the prenatal period on routine screening ultrasound, allowing for prenatal consultation and planning (12). The findings on ultrasound ranges from an incidental finding of a cystic appearing lesion to massive pulmonary involvement with the development of hydrops Hydrops can develop in up to 40% of cases and regression of the lesion is seen in up to 60 % during the course of gestation. The need for fetal intervention is rare and limited to those cases with severe hydrops (13) with a predicted mortality of near 100%. Because of the increased prenatal detection rate it is now recognized that there is complete spontaneous resolution during gestation in up to 20% of lesions.

The differential diagnosis of CCAM includes other cystic diseases of the lung including bronchopulmonary sequestration (BPS), bronchogenic cyst,

and congenital lobar emphysema. The primary differentiation between CCAM and BPS are based on 2 anatomic points. BPS has no connection to the tracheobronchial tree and are supplied by an anomalous systemic artery. CCAMs are not. However the difference between the 2 lesions is not as discreet as once though and it is more likely that the 2 are variants of the same abnormal development pathway

Prenatal diagnosis is usually made by ultrasonography and generally classified into two categories; microcystic lesions with cyst <5mm which appear echogenic and solid and macrocystic lesions of one or more cysts >5mm (14). MRI is also being used more frequently to exam the fetus and can help differentiate CCAM from other thoracic lesions including congenital diaphragmatic hernia, foregut duplications, and others.

In the neonatal period the diagnosis is usually suspected based on clinical presentation and the initial chest x-ray. A CT scan is usually definitive, although the exact diagnosis may not be made until surgical exploration is performed . Diagnosis later in life is usually dependant on late symptoms or in some cases an incidental finding on a routine CXR. CT scan is still the gold standard.

Congenital Lobar Emphysema (CLE) is a rare anomaly of lung development which often presents in the neonatal period with hyperinflation of one or more pulmonary lobes. Other terms for this entity include congenital lobar over-inflation and infantile lobar emphysema. The patient may also present with a radio-opaque mass on cxr because of delayed clearance of lung fluid of the affected lobe. The differential diagnosis must include the other types of congenital cystic lung disease as already mentioned.

CLE is a relatively rare developmental anomaly that occurs in 1/20,000 to 1/30,000 (15). The upper lobes tend to be the most frequently involved with the left side more common (40 t0 50%) then the right (20%). The middle lobe is occasionally involved (25 to 30%) and lower lobe disease is extremely rare (2 to 5%) (16). The male to female ratio is 3 to 1.

The etiology of CLE is multifactorial and in fact is likely a common endpoint for a number of different disease pathways. Progressive hyperinflation is the end result of a number of variations of disruption of bronchopulmonary development. These disturbances may cause a change in the number of airways or alveoli and alveolar size (17).

The most frequently documented cause of CLE is obstruction of the developing airway which occurs in approximately 25% of cases. Airway can either be intrinsic or extrinsic, although intrinsic is the most common. This results in a ball-valve type obstruction which results in air trapping. This results in histologic changes of alveolar distension without structural anomaly.

Treatment in the symptomatic or asymptomatic patient is surgical resection. In most case a complete lobectomy verses a wedge or segmentectomy of the involved lobe is indicated. This is favored because of the difficulty, on a macroscopic level, of determining what portion of the lung is involved and which is not. The increased difficulty and morbidity associated with a partial resection does not warrant the limited benefits of preserving a portion of possibly diseased lung. There are cases where more then one lobe appears to be involved or there maybe bilateral disease. There are also cases where there are no clear anatomic planes between the upper and lower lobe lobes and segments of both are grossly involved. In these cases segmentectomies or some other sort of lung preserving surgery becomes necessary.

The symptomatic neonate needs to be operated on immediately. In extreme cases an emergency thoracotomy with decompression of the chest cavity can be a life saving intervention and an emergency lobectomy is performed. However in most neonatal cases, especially with the increased incidence of prenatal diagnosis, the baby is asymptomatic or has mild to moderate symptoms and a semi-elective resection can be performed.

The timing of surgery remains somewhat controversial but there is little evidence to suggest that delayed resection benefits the child in any significant way. In fact delayed surgery may increase the risk of infection or respiratory compromise. Also early resection maximizes the compensatory lung growth of the remaining lobe (s).

Many centers have recommended resection at between 1 to 6 months of age to allow for some growth and to decrease the risk of the anesthetic. We have favored early intervention to minimize the risk of later complications and because we feel current surgical techniques and anesthetic care eliminate the risks of early resection.

The standard therapy has been a formal lobectomy through a postero-lateral thoracotomy incision. In most case this can be done using a muscle sparing approach, decreasing the morbidity associated with a formal thoracotomy, including scoliosis, shoulder girdle weakness, and chest wall deformity, all of which have been well documented in infants.

For the last decade we have preferred to use minimally invasive techniques to perform lobectomies in all of these infants with congenital cystic lesions. The benefits of avoiding a formal thoracotomy and the morbidity associated with it greatly out way the disadvantages of the increased technical difficulty and operative time. In fact with experience the operative times have equaled or are faster then with a standard thoracotomy.

The technique of a lower lobectomy is detailed here for demonstration purposes.

The surgeon and assistant are at the patient's front with the monitor at the patients back. The chest is initially insufflated through a veres needle placed in the mid-axillary line at the 5th or 6 interspace. A low flow, low pressure of CO_2 is used to help complete collapse of the lung. A flow of one liter per minute and pressure of 4 to 6mm Hg is

maintained throughout the case. The first port (a 5mm) is placed at this site to determine the position of the major fissure and evaluate the lung parenchyma. A 4mm or 5mm 30 degree lens is used. This allows the surgeon to look directly down on the fissure and his instruments. In general this will be the camera port. Position of the fissure should dictate the placement of the other ports as the most difficult dissection occurs in this plane. The working ports (3mm or 5mm) are then placed in the anterior axillary line between the 5th and 8th or 9th interspace. Upon entering the chest the anatomy is often difficult to identify because of the large space occupying cysts. Therefore to create space and improve visualization the cysts are involuted using the Ligasure device. The cysts are grasped and sealed until enough compression is achieved to allow for identification of all anatomic structures. The compressed lobe is also much easier to manipulate with laparoscopic instruments.

The first step is mobilization of the inferior pulmonary ligament. Care should be taken to look for a systemic artery coming off the aorta incase this is a misdiagnosed sequestration or one of the hybrid lesions. The inferior pulmonary vein is dissected out but not ligated at this point. Ligation prior to division of the pulmonary artery can lead to congestion in the lower lobe, which can create space issues especially in the smaller child and infant. The fissure is then approached going anterior to posterior. In cases of an incomplete fissure the Ligasure or Endo-GIA can be used to complete the fissure. Gradually the pulmonary artery to the lower lobe is isolated. Often it is necessary to dissect into the parenchyma of the lower lobe to gain extra exposure and length. If possible the artery can be ligated at its' main trunk to the lower lobe as it passes thru the fissure. However it is often easier to dissect out the vessel after the first or second bifurcation. This also provides a longer segment of artery to work with. The bronchus to the lower lobe lies directly behind the artery and can often be palpated before it is seen.

Once the artery is divided the complete dissection of the vein is facilitated because of improved exposure. The vein can also be taken in assorted ways depending on the size of the vessel. Prior to taking the vein it is helpful to divide the pleura along the posterior border of the lobe to complete it's mobilization. The inferior pulmonary vein is then divided and the bronchus to the lower lobe isolated. The bronchus is divided with the EndoGIA in larger children or cut sharply and closed with 3-0 PDS suture in smaller patients (In infants and patient's under 5 kg it is possible to seal the bronchus with Endo-clips. The specimen is then brought out through the lower anterior axillary line trocar site, which is slightly enlarged if necessary, either whole or piecemeal with a ring forceps. If more then one lobe is involved on the same side then a lung sparing procedure should be performed. This is best illustrated by a case in which on CT scan the CCAM appeared to involve just the left lower lobe.

We have now performed over 200 thoracoscopic lobectomies for congenital lung lesions.

Recent study evaluated 144 patients underwent video assisted thoracoscopic lobe resection, ages ranged from 2 days to 18 years, average stay in 2.8 days. Operative time ranged from 35 minutes to 220 minutes (average 125 minutes). Only 3 intraoperative complications required conversion to an open thoracotomy (18, 19).
Thoracoscopic lung resection is a safe and efficacious procedure, it avoids the morbidity of a thoracotomy and is associated with less postoperative pain and hospital stay. We highly recommend this approach as well as early intervention as the surgery becomes much more difficult if a significant pulmonary infection occurs before resection.

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El autor declara no tener conflictos de interés, en relación a este artículo.