Original Article

Congenital Malformation of Lung Parenchyma: 15 Years Experience in a Thoracic Surgical Unit

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ABSTRACT

Objective. To review the surgical management of congenital malformations of lung parenchyma in a thoracic surgery unit over a period of 15 years.

Methods. We carried out a retrospective analysis of records of all patients who had surgery for congenital malformations of lung parenchyma between 1995 and 2010.

Results. Forty-five patients underwent surgery for congenital lung lesions out of 3735 thoracotomies performed during the study period. The lesions included 29 lung sequestrations, 12 bronchogenic cysts, 3 congenital lobar emphysema and one congenital cystic adenomatoid malformation. Only 26 (26%) cases were diagnosed preoperatively. Twenty-eight (62.2%) patients underwent lobectomy, 5 (11.1%) patients had pneumonectomy, and 10 (22.2%) patients had removal of cyst while 2 (0.45%) patients had lung resection with repair of the oesophageal connection. No mortality was recorded. One patient had post-operative complication of oesophageal fistula which was successfully managed conservatively. The follow-up was between 8 months to 14 years. All patients were asymptomatic and had no physical limitations during the follow-up.

Conclusions. Surgery is curative and produces good long-term result in patients with congenital malformations of lung parenchyma. It should be offered to patients as a therapeutic option where indicated and feasible.
[Indian J Chest Dis Allied Sci 2012;54:105-109]

Key words: Congenital malformations, Lung parenchyma, Congenital cystic adenomatoid malformation, Bronchogenic cyst, Sequestration, Lobar emphysema.

INTRODUCTION

Congenital lung malformations are uncommon and depending on the size of the lesion and degree of functional impairment, these may lead to considerable morbidity and mortality. Such lesions include congenital cystic adenomatoid malformation (CCAM), bronchopulmonary sequestration, congenital lobar emphysema (CLE) and bronchogenic cysts. These may have a notwithstanding embryological origin as specific features, histologically hybrid lesions may exist among them. This is supported by the reported findings of a complex congenital pulmonary malformation comprising of a congenital pulmonary airway malformation, an intralobar sequestration and two bronchogenic cysts, within the same lobe in a 5-year-old girl.[1] It is widely accepted that congenital lung lesions result from perturbations in lung and airway embryogenesis and the level, degree and timing of obstruction of the embryologic insult correlates with the type of the lesion and the histopathology.[2]

A diagnosis of these conditions may sometimes be made ante-natally by pre-natal sonography but is usually post-natally. In those recognised post-natally, the patient may be asymptomatic and discovered incidentally on thoracic imaging or may present with complications. The most common complication is pneumonia, that may respond poorly to medical treatment. Other complications include the development of malignancies (carcinomas and pleuro-pulmonary blastomas), pneumothorax, haemoptysis or hemothorax.[3]

Although an incidence of around 1 in 5000 to 10000 births have been suggested,[4] only 10% of cases are identified at birth, whereas 14% are diagnosed later by 15 years of age.[5]

Surgery, either open thoracotomy or video-assisted thoracoscopy is the usual modality for management.

[Received: June 14, 2011; accepted after revision: October 5, 2011]

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We present a review of our surgical experience of managing these lesions. The clinical presentation, diagnosis, surgical management and long-term outcome of patients who primarily presented or were referred with congenital malformations of lung parenchyma are discussed.

MATERIAL AND METHODS

During a 15-year period from 1995 to 2010, 3735 thoracotomies were performed. Out of these, 45 patients underwent surgery for congenital lung malformations at the Department of Thoracic Surgery, LRS Institute of Tuberculosis and Respiratory Diseases, New Delhi. Data were collected from case notes, operation register and operation notes. The age, sex, time of presentation, pre-operative symptoms, histological features, radiological diagnosis, surgical procedure, operative and post-operative complications, and follow-up course were documented.

Radiological investigations included plain chest radiograph, computerised tomography, pulmonary function tests, echocardiography, bronchoscopy and angiography as indicated pre-operative. All the cases were operated by one surgical team. We excluded two cases of congenital diaphragmatic hernia with lung hypoplasia and infective/inflammatory lung pathologies mimicking congenital lung hypoplasia seen within this period.

The data were analysed using Statistical Package for the Social Sciences (SPSS version 16.0).

RESULTS

Out of the 45 patients, 28 were males while 17 were females, giving a male to female ratio of 3:2. The age ranged at presentation from one-and-a-half year to 32 years with a mean of 20.3 years.

The commonest lesion was lung sequestration that accounted for 29 (64.4%) cases. This was followed by 12 (26.7%) cases of bronchogenic cysts (Figure 1), three (6.7%) cases of CLE and one (2.2%) case of CCAM. None of the lesions were diagnosed before birth. All the patients presented in childhood or early adulthood with chest symptoms that included cough, expectoration with occasional haemoptysis. Those with congenital lobar emphysema presented with breathlessness, the clinical and radiological picture mimicking pneumothorax.

Pre-operative diagnosis was made in only 12 (26%) cases. Other patients were taken up as cases of inflammatory lung disease with sufficient symptoms to merit surgical intervention.

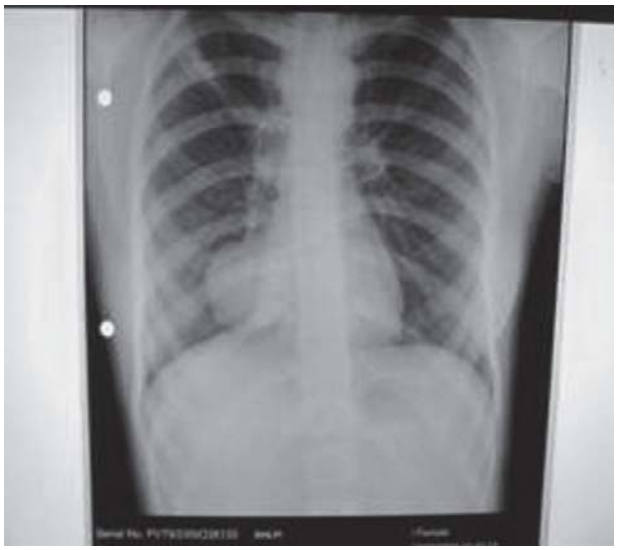

Figure 1. Chest radiograph (postero-anterior view) showing a bronchogenic cyst.

Twenty-eight (62.2%) patients underwent lobectomy, 5 (11.1%) patients had pneumonectomy, 10 (22.2%) patients had removal of cysts while 2 (0.45%) patients had lung resection (Figure 2) with repair of the associated oesophageal connection. The surgical time ranged from one hour to 2.5 hours with a mean of 105 minutes. Intra-operative blood loss ranged from 40 mL to 700 mL with a mean of 200 mL. Intensive care stay ranged from one day to four days with a mean of two days, while the length of admission ranged from 15 days to 45 days.

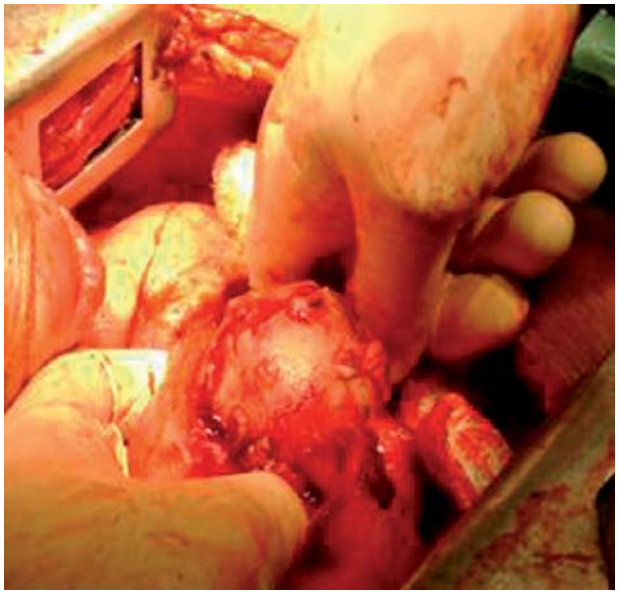

Figure 2. Photograph showing resection of a bronchogenic cyst.

There was no mortality and negligible post-operative morbidity except for an oesophageal fistula that developed in one patient. This was in a child of 8

years age who had sequestration in the right lower lobe that was communicating with the oesophagus. The fistula was treated with long term chest tube drainage, and feeding by jejunostomy. It healed over a period of one and a half months.

All the patients were followed up for a period ranging from 8 months to 14 years. There have been no further complaints except for occasional chest pain at the site of thoracotomy.

DISCUSSION

In the developing countries, the thoracic surgeon is most commonly challenged with infective lung pathologies, such as complications of pulmonary tuberculosis.[6] Concomitant recognition and successful management of rare congenital lung pathologies over a period of 15 years has been reported only rarely. Hence, we have presented our experience. Similar studies on congenital lung parenchymal malformations have also reported a male predominance[7,8] as observed in this study.

In our series, the commonest pathology observed was pulmonary sequestration followed by bronchogenic cysts. Other lesions included CCAM and CLE. Our findings were similar to those of Evrard *et al*.[7] However, some authors have reported CCAM as the predominant pathology in their series[8,9], while others have found bronchogenic cysts as the commonest congenital lung malformation in their study.[10] Pulmonary sequestration is widely regarded as being the commonest and represents 0.15% to 6% of all pulmonary malformations.[11] CCAM are extremely rare with the reported incidence being between 1/25000 and 1/35000,[12] while CLE is a relatively rare developmental anomaly that occurs in 1/20000 to 1/30000.[13]

The surgeon requires a high index of suspicion in making a pre-operative diagnosis of these conditions, especially in the setting of a developing country where infectious conditions of lungs are common.[14] Congenital parenchymal lung malformations are now not only increasingly being diagnosed by routine usage of ante-natal ultrasound, but adequate intervention is being employed with reasonable success. An ante-natal ultrasound of CCAM finding ranges from an incidental finding of a cystic appearing lesion to massive pulmonary involvement with the development of hydrops foetalis. Hydrops will result from a mass effect resulting in mediastinal shift causing vena caval obstruction or cardiac compression with decreased venous return.[15]

All our 45 patients presented to us in childhood or early adulthood with complaints of cough, breathlessness and mild haemoptysis. Patients with CLE are likely to present early with rapidly progressive respiratory distress as the expanding lesion gradually compresses the healthy lung parenchymal tissue. Intra-lobar pulmonary sequestration usually presents after infancy with recurrent attacks of chest infection while extra-lobar pulmonary sequestration is usually asymptomatic but may also be associated with respiratory distress because of airway compression. Though we did not evaluate the duration of symptoms, other studies have shown similar symptoms at presentation with a mean duration of 15.3 months.[10] The lesions that present in adults are often prone to misdiagnosis and mimic other intra-thoracic pathologies but these can be reliably diagnosed with computerised tomography and magnetic resonance imaging.[16] These may have the potential for malignant transformations, mainly pleuro-pulmonary blastoma in infants and young children and bronchiole-alveolar carcinoma in older children and adults.[16, 17]

In those that were diagnosed pre-natally, Raychaudhuri *et al*[18] reported successful use of amnio-reduction and thoracocentesis in a small minority with rapidly growing lesions that can lead to hydrops foetalis. Adzick[19] performed foetal lobectomy in lesions predominantly solid or multicystic and ex-utero intrapartum treatment (EXIT) has been performed for selected fetuses pre-natally diagnosed with complicated lung malformations.[20] Bethamethasone has also been shown to be beneficial causing resolution of hydrops.[21-23]

In our Centre, we practice open surgical removal of these malformations. Most of our patients underwent lobectomy through a postero-lateral thoracotomy. Others had pneumonectomies, removal of cysts and lung resections with repair of the oesophageal connection. Lobectomy is the treatment of choice for CCAM, CLE and intra-lobar sequestration[24], while a bronchogenic cyst can generally be managed by simple cyst resection (Figure 2), segmentectomy or lobectomy. Open lung resection has been shown to be the gold standard in managing congenital lung malformations as it enables parenchymal sparing surgery, is versatile, has few complications and produces very good long-term results.[8] Elective resection of all CCAM, bronchogenic cysts, and intra-pulmonary sequestrations is advocated because of the risk of complications, such as infection, haemorrhage, pneumothorax, sudden respiratory compromise, and malignant transformations.[25] Even in asymptomatic cases, infection has been implicated. Pellizo *et al*[26] reviewed 24 cases of pre-natally diagnosed CCAM and found that 79% of asymptomatic infants operated electively at three months of age had evidence of infection.

Though, thoracoscopic resection has been found to be a safe and feasible alternative to open resection of congenital lung lesions, none of our patients was found suitable for this procedure as they were usually misdiagnosed pre-operatively to have

inflammatory pulmonary pathologies requiring surgical intervention. Diamond *et al*[27] compared the outcomes in children undergoing thoracoscopic *versus* open resection of congenital lung lesions. They found that peri-operative outcomes including operative time, length of stay, duration and volume of chest tube drainage, and dose and duration of intravenous opioids were similar between the procedures. However, children undergoing thoracoscopic procedures were less likely to have received adjunctive regional anesthesia.[27] Decreased post-operative pain, a shorter hospital stay, and a better cosmetic result are definite advantages of video-assisted thoracoscopy.[28] CLE have been found to be more challenging thoracoscopically. Rahman and Lakhoo[29] recommended that these lesions should be pre-selected for open surgery.

The timing of surgery in asymptomatic patients is not well defined, with recommendations ranging from one month to two years of age.[25] Adzick[17] recommend elective resection at one month of age or later. They chose this age because anesthetic risk in babies decreases after four weeks of age and an experienced pediatric surgeon can safely perform a lobectomy in infants with minimal morbidity. Early resection also maximises compensatory lung growth. Mc Bride *et al*[30] followed up 15 patients who had lobectomies between one week and three years of age for 8 to 30 years and found that vital capacity, residual volume and total lung volume were within normal range in most of the patients.

The length of hospital stay for our patients was between 14 days to 45 days. Costa Júnior *et al*[10] reported an average hospital stay of 14.5 days (3-70).

Though we recorded no mortality from open pulmonary resection and had post-operative morbidity from oesophageal fistula, other authors[7,10] have reported mortality resulting from pulmonary hypoplasia and pulmonary hypertension and post-operative complications including pneumothorax, pleural effusion, prolonged air leak, portal vein thrombosis, haemorrhage requiring re-intervention, pleural empyema, atelectasis and sepsis.

CONCLUSIONS

Congenital malformations of lung parenchyma are rare entities that may be detected early pre-natally or may present in the newborn as acute respiratory distress or may remain asymptomatic until later in adulthood. In developing countries, a high index of suspicion is required to diagnose congenital malformation of lung parenchyma as most cases masquerade as inflammatory lung pathologies. Results of open surgery are gratifying; hence it is an effective way of managing these lesions.

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