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Surgical Treatment of Lung Malformations

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Objective. To review the surgical management of Lung Malformations (LM) in a pediatric surgery unit over a period of 16 years.
Patients and Methods. We retrospectively reviewed the medical records of 79 consecutive children who underwent surgery for LM in our clinic between 2006 and 2022.
Results. Seventy-nine patients underwent 83 pulmonary resections during the study period. The mean ages at diagnosis of LM and at the time of surgery were 8.10 ± 4.10 years (range, 1 to 17 years) and 9.70 ± 4.20 years (range, 2 to 18 years), respectively. There were 44 males (55.7%) and 35 females (44.3%). The causes of LM were congenital hypoplasy of bronchi (n = 31), secondary bronchiectasis was additional nonspecific pulmonary infection (n = 42) and foreign body aspiration (n = 6). Chest X-rays, bronchography and bronchoscopy (n = 79), chest HRCT (n = 61), angiopulmonography (n = 23) were used, and pulmonary function tests (n = 48) were performed. The types of resections were segmenectomy (26.5%), lobectomy (35.4%), and lobectomy with segmentectomy (17.8%), bilobectomy (8.9%) and pneumonectomy (11.4%). The postoperative status of the patients as follows: "well" in - 49 patients (60.7%), "improved" in 23 patients (29.5%), "worsened" in 6 patients (7.7%). And unfortunately two patients "died" (2.6%).
Conclusions: A radiologic and morphologic evidence of reversal of airway abnormality has been shown in cases of congenital defects of bronchial wall. The morbidity and mortality rates of LM surgery are within acceptable ranges. Segmentectomy and lobectomy are well tolerated in children without increase in morbidity and mortality.
Keywords: Airway abnormality, bronchiectasis, Lung Malformation, pulmonary resection, Children.

1. Introduction

Lung Malformations (LM) is rather uncommon, and knowledge of this disorder is not very extensive. LM and bronchiectasis are defined as permanent and irreversible abnormal dilatation of bronchi, and is usually considered to be acquired. There is a type of LM and bronchiectasis which is probably congenital in origin, although in most cases this is difficult to prove [6, 10]. Congenital cystic LM is thought to be the result of developmental arrest [16]. This may result in cyst formation, retention of fluid or air, and the risk of subsequent infection [1]. Congenital cystic LM has been found almost exclusively in infants and young children [2, 3] and also has been reported accompanying other congenital anomalies [4], or in patients with deficiency of bronchial cartilages. Surgery was considered the only alternative method for the treatment of LM in the early and mid years of this century. However, the indications for surgery have been limited in recent decades because of expectant results of conservative treatment [12, 14]. Although there are no prospective randomized trials comparing the efficacy of conservative and surgical treatment options, the indications of surgery for LM can be considered in 2 main categories among children. Surgery can be performed for eradication in children suitable for total resection who do not respond to conservative treatment or for controlling the symptoms by removing the worstinvolved part in children with diffuse LM [16, 18]. A retrospective series analysis was conducted to evaluate the results of surgical treatment according to the reasons and procedures in children with LM.

2. Materials and Methods

We reviewed the medical records of all children who underwent surgery for LM between 2006 and 2022 at Pediatric Surgery Department of the TashPMI and NCMC. Medical records of 79 patients were analyzed for age, gender, clinical characteristics, radiologic findings and details of surgery including type of resection, operative morbidity, mortality, and overall outcomes.

Surgical treatment was considered if the symptoms persisted in spite of repeated courses of medical treatment including antibiotics, mucolitics, chest physiotherapy, bronchodilators, or steroids. Candidates were evaluated for the resectability of all diseased lung tissue or the expected benefit after excision of most affected parts of lungs with extensive disease.

Preoperative bronchography or chest high-resolution computed tomography (HRCT) was performed to evaluate the type, severity, and distribution of LM. Pulmonary function tests were done to estimate the lung function. Bronchoscopy was performed in all patients to rule out bronchial obstruction or stenosis caused by foreign body or endobronchial tumor.

Preoperative bronchoscopy was performed to clear bronchi and to prevent spillage of secretions during operation. Surgical acces was obtained via posterolateral thoracotomy in all patients. The fourth or fifth intercostal spaces were used to reach the diseased lung area. The most common surgical procedures were formal segmentectomy and lobectomy, however, in some of the cases, when required lobectomy plus segmentectomy, bilobectomy and pneumonectomy was performed.A chest tube was placed and connected to an underwater seal system by Byulau in all patients. Postoperative management included intensive chest physiotherapy, analgesic, and antibiotics.

Complications were analyzed for intraoperative and postoperative periods. The outcome of surgery was evaluated at last follow-up visit and rated according to the degree of symptoms. Symptoms were categorized as follows: “well” when the patients are free of symptoms, “improved” when patient’s symptoms redused, “worse” if the patient’s symptoms persist.

3. Results

Seventy-nine patients underwent 83 pulmonary resection operations with the diagnosis of LM during the study period. The mean ages at diagnosis of LM and at the time of surgery were 8.10 ± 4.10 years (range, 1 to 17 years) and 9.70 ± 4.20 years (range, 2 to 18 years), respectively. There were 44 males (55.7%) and 35 females (44.3%). The causes of LM were nonspecific pulmonary infection (n = 42), congenital hypoplasy of bronchi (n = 31), and foreign body aspiration (n = 6). The details of comorbidities are shown in Table 1.

Table 1. The comorbidities with LM

Number of reported associated hereditary and inborn diseases in patients with LM (n = 31)

Comorbidities	No.	%
Oligophrenia	3	3.8
Congenital atrophy of the optic nerve	1	1.2
Chronic diseases of ENT organs	21	26.6
Chestwalldeformity	5	6.3
Congenitalheartdisease	9	11.4
Aplasia or hypoplasia of the kidney	4	5.0
Hernias (diaphragmatic, inguinal, umbilical)	8	10.1
Congenital dislocation of the thigh	1	1.2
Total	52	54.8

The majority of comorbidities were diagnosed in the younger age group. Hidden leaking anomalies were identified with additional examination. It should be noted that in 11 (13.9%) children these diseases are detected with various combinations that significantly affect the type of treatment.

Initial radiologic evaluation was made through chest X-rays in all patients, and the diagnosis of LM was confirmed by bronchography (n = 79) or chest HRCT (n = 61). Bronchography had been used in the early years of the study period. High-resolution CT has been preferred instead of bronchography afterwards. Pulmonary vascular angiography was performed in 23 patients to evaluate the functional and anatomical status of pulmonary circulation. Pulmonary function tests were performed in 48 children and showed pathologic results in 31.

The operative resections consisted of segmenectomy (26.5%), lobectomy (35.4%), and lobectomy with segmentectomy (17.8%), bilobectomy (8.9%) and pneumonectomy (11.4%). The details of surgical resection were given in Table 2.

Table 2. Type of Surgery and Distribution of Resected Lung Parts

SurgicalProcedure			No. (%)
Segmentectomy			21 (26.5)
Lobectomy	Upper	7	28 (35.4)
	Middle	2	
	Lower	19	
Lobectomy+Segmentectomy	Left lower+		14 (17.8)

	IV-V segments		
Bilobectomy	Upper	2	7 (8.9)
	Lower	5	
Pneumonectomy			9 (11.4)
Total			79 (100)

A radiologic and morphologic evidence of reversal of airway abnormality has been shown in cases of post-infectious LM and congenital defects of the bronchial wall. All specimens underwent the pathologic examination that confirmed the diagnosis. Histopathological examination of all specimens showed varying degrees of bronchiectatic changes, including destruction of the lung parenchyma, congenital defects of segmental bronchus and bronchioles.

Four patients with bilateral LM subsequently required second operation of the other lung. Two patients of them had undergone right lower lobectomy, by one childhoods underwent right upper lobe, another left lower lobectomies. One patient with incomplete resection subsequently required second operation for ongoing LM underwent complementary right pneumonectomy.

Postoperative complication was encountered in 6 (7.6%) patients. One patient underwent reoperation for decortication in the second postoperative week. The remaining five children developed post-operative pneumonia, atelectasis and respiratory failure; those were managed by conservative means with bronchoscopy. Long-term results of treatment in terms of 6 months up to 14 years studied in 78 (98.7%) teens. The patients who have undergone surgery were postoperatively rated as follows: well, 48 patients (60.7%); improved 23 patients (29.5%); unchanged or worsened 6 patients (7.7%). Two patients died (2.6%); one of them in the early postoperative period, second was in the closest after the operating period with bilateral LM. The details of the type of surgery are given in Table 3. Most of the patients who were well or clinically improved had completely resectable lesions.Segmentectomy and lobectomy are well tolerated in children without increase in morbidity and mortality. Therefore, resection of damaged part of the lung tissue may be preferred instead of removing much volume lung tissue ($P < 0.05$).

Table 3. Long-term results of LM Surgery According to Type of Surgical Resection

Procedure	Numberofpatients	Results			
		Well	Improved	Worse	Exitus
Segmentectomy	21 (26.5%)	17	4	-	-
Lobectomy	28 (35.4%)	22	6	-	-
Lobectomy +Segmentectomy	14 (17.8)	7	5	1	1
Bilobectomy	7 (8.9%)	2	3	1	1
Pneumonectomy	9 (11.4%)	1	5	3	-
Total	79 (100%)	48(60.7%)	23 (29.5%)	6 (7.6%)	2 (2.6%)

4. Discussion

Demonstration of abnormalities of bronchial architecture by using nebulized bismuth powder and iodized oil allowed further understanding of the pathogenesis of LM. In modern description, LM is characterized by abnormal, irreversible dilatation of the bronchi in association with a variable degree of chronic bronchitis, pneumonitis, or other pathologic

changes. The common cause of LM is bacterial infection usually after pneumonia. Other causes include pertussis, viral infections, and obstruction secondary to foreign body aspiration, tracheobronchial aspiration, congenital lesions and immune deficiency [5,17]. Cystic fibrosis is the most common cause of LM especially in the white population of Europe and North America. Nonspecific pulmonary infection was the most frequent cause of LM in our series. The prevalence of LM decreased significantly over recent decades. In the base of some of LM lays the hypoplasia of the bronchial wall elements, bronchioles and alveolus in whole or limited area of the lung [8, 10]. Antimicrobial therapy and immunization against viral and bacterial agents significantly decreased the incidence of LM. Early recognition of foreign bodies with bronchoscopy also decreased the incidence of postobstructive LM. However, the incidence of LM is still high in developing countries [9]. The initial treatment of LM is conservative and includes reducing airway obstruction and elimination of microorganisms from the lower respiratory tract. Therefore, medical therapy for LM consists of antimicrobial therapy, postural physiotherapy, bronchodilators, corticosteroids and bronchoscopic cleaning of the tracheobronchial tree [15].

The main rationale for surgical treatment of LM is to increase the quality of children's lives and protect them from complications such as empyema, hemoptysis, and lung abscess. The successful results can be achieved only by careful and proper selection of candidates for surgery. Instead of following rigid criteria for patient selection, each child should be evaluated separately for surgery [7, 11].

The indications for surgery are not fully established in children [17]. However, there seems to be agreement that the appropriate antibiotic therapy and chest physiotherapy should be tried for a period. In adults, indications for surgery have been reported to be resistance to appropriate antibiotic therapy in addition to an extensive postural drainage and chest physiotherapy program for a reasonable period of at least 2 years. In children, in addition to the above-mentioned criteria, growth retardation, failure to thrive, inability to follow the educational program because of frequent infections, and socioeconomic status of the family should also be considered when deciding on surgical treatment [10].

Since all diseased parts of the lung can be resected, the decision for surgery is relatively easy in children with localized LM [13]. The presence of accompanying complications such as growth retardation, abscess, and hemoptysis help surgeons in their decision for operation. However, when the LM diffusely affects multiple lobes or segments and sometimes both lungs, the operative decision is quite difficult. Especially in the presence of comorbidities such as deformation of thorax and congenital heard diseases, total resection of all affected tissue usually is impossible. All candidates of surgery should be free of an ongoing pulmonary infection at the time of operation. Broad-spectrum antibiotics should be given in the preoperative period. Bronchoscopy should be performed in all candidates to exclude bronchial obstruction caused by a foreign body. Bronchoscopy should be also done just before the operation to clear airways. In our series, a rigid or flexible bronchoscopy, or both, was performed for the removal of secretions and the detection of a foreign body or an endobronchial lesion [15].

The localization of disease should be evaluated radiologically. Bronchography had been used extensively for both diagnostic and follow-up purposes in LM. Although it was very sensitive

and informative, it has been almost totally abandoned today in children. High-resolution chest CT has replaced the bronchography. Angiography has been recommended to find out whether the diseased area is perfused and detected of pulmonary vessels hypoplasy. It has been suggested that only nonperfusing lung and pulmonary vessels hypoplasy areas should be resected [9, 18]. Pulmonary function tests may not be required in children with localized LM. However, it should be performed in cases of extensive involvement or in patients undergoing reoperations.

The aim at operation is to excise all diseased lung areas whenever possible and to preserve as much healthy lung parenchyma as possible. It is known that even only 2 or 3 preserved segments can fill the hemithorax, if operation had performed till 8 age old. All resections should be performed within anatomic limits. Lobectomy was the most frequently performed operation in our series. Left lower lobe was the frequently affected area as seen usually in LM. Pneumonectomy was the second most frequent operation. Although pneumonectomy has been accepted to result more complications and mortality than that of other limited resections, complications has occurred after removing much volume lung tissue, such as lobectomy plus segmentectomy, bilobectomy and pneumonectomy in long-term period, and it has been well tolerated in children in the current series [17].

The success of surgery for LM is directly related to the extension of the disease, underlying cause of LM, and the completeness of the resection. Most of the children benefited from LM surgery with regression of symptoms. Bilateral LM exposed a high risk of mortality with all deaths occurring in these children. We could not find the exact cause of deaths in this patient, because no autopsy was allowed. Patients with bilaterally LM should undergo a complete cardiologic evaluation for possible accompanying intrinsic abnormalities before operation [16]. Total excision of the diseased lung areas was accomplished in most children resulting in an apparent improvement in symptoms. However, symptoms have been persistent in children who had incomplete resection [10]. Whether in the same or other site, the presence of residual diseased lung area has the potential to produce symptoms.

5. Conclusions

The decision for surgical intervention should be made in cooperation with the chest diseases unit. Anatomic localization of the disease should be mapped clearly by radiologic investigations. In addition, radiologic and morphologic evidence of reversal of airway abnormality has been shown in cases of congenital defects of bronchial wall. The morbidity and mortality rates of LM surgery are within acceptable ranges. Most of the children benefit from the surgery, especially when total excision of the all diseased lung areas can be accomplished. Segmentectomy and lobectomy are well tolerated in children without increase in morbidity and mortality. Therefore, resection of damaged part of the lung tissue may be preferred instead of removing much volume lung tissue, but pneumonectomy may be preferred instead of leaving residual disease in the absence of contralateral lung involvement.

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