Surgical Management of Congenital Pulmonary Malformations After the First Decade of Life

Alice Wang, MD, Thomas A. D'Amico, MD, and Mark F. Berry, MD

Department of Surgery, Division of Thoracic Surgery, Duke University Medical Center, Durham, North Carolina

Background. **Most congenital pulmonary malformations are discovered early in life, but some are diagnosed in adulthood. We evaluated patients treated surgically after the first decade of life.**

Methods. **All patients who underwent surgical treatment for a congenital pulmonary malformation diagnosed after 10 years of age at a single institution from 1997 to 2012 were evaluated for presenting symptoms, surgical management, perioperative outcomes, and symptom resolution.**

Results. **Twenty-two patients met the inclusion criteria. The most common malformations were pulmonary sequestration (n = 12, 55%), congenital cystic adenomatoid malformation (n = 2, 9%), and bronchial agenesis (n = 2, 9%). The median age at diagnosis was 36 years (range, 10–66 years). The most common presenting symptoms were dyspnea (n = 6, 27%) and hemoptysis (n = 4; 18%); 4 (18%) asymptomatic patients received diagnoses. The median duration of symptoms before operation was 14 months. An emergency room visit or hospitalization occurred in 11 patients (50%) before their referral for surgical evaluation. The surgical approach was thoracotomy for 7 patients (32%) and thoracoscopy for 15 patients (68%). All vascular anomalies requiring a pneumonectomy (n = 3, 14%) were done by a thoracotomy, and 83% (10/12) of pulmonary sequestrations were treated thoracoscopically. The median hospital stay was 3 days. There were no perioperative deaths, and minor morbidity occurred in 4 patients (18%). Complete resolution of symptoms after operation occurred in 94% (16/17) of patients, with a median follow-up time of 3 weeks.**

Conclusions. **Early surgical management of congenital pulmonary malformations found after the first decade of life is recommended to control symptoms and avoid hospitalizations. Most adult pulmonary sequestrations can be treated with minimally invasive techniques.**

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Congenital pulmonary malformation is a rare condition and includes bronchopulmonary sequestration, congenital cystic adenomatoid malformation (CCAM), and bronchial and vascular abnormalities. CCAM is the most frequent congenital malformation of the lung [1], and sequestrations account for 0.15% to 6.4% of congenital malformations, with intralobar sequestrations accounting for three-fourths of all cases [2]. In many large series of patients with diagnoses of congenital pulmonary malformations, symptoms occurred within the first decade of life [2–4], and an increasing number of patients are receiving prenatal diagnoses [5]. Many patients, however, do not receive accurate diagnoses until the age of 10 years or even as late as adulthood [6].

The optimal management of congenital pulmonary malformations discovered later in life is not entirely clear. Only a few case series on the surgical management of pulmonary sequestration in children have been published [7–9], and several case studies have reported on adult presentation of pulmonary sequestration [10–12], but few reports on consecutive surgical management of late-presenting congenital pulmonary malformation have appeared. In addition, the utility of a minimally invasive approach for resection with thoracoscopy has been extensively reported for malignant conditions but has been less well documented for benign disease [13–17]. This study was, therefore, undertaken to examine the presentation and treatment of patients who underwent resection of congenital pulmonary malformations after the age of 10 years.

Patients and Methods

After local institutional review board approval was granted, including waiver of the need for patient consent, the Duke University Medical Center data bank was queried to identify patients 10 years of age or older who had undergone resection of congenital pulmonary abnormalities between 1997 and 2012. A retrospective review of these patients documented demographics, quality and duration of symptoms, age at diagnosis, hospitalizations or emergency room visits because of symptoms, surgical management, and postoperative course. The preresection course was evaluated for patterns of signs or symptoms that might have raised the suspicion of a congenital abnormality and allowed earlier diagnosis. In addition, the details of surgical management and patient outcomes were assessed to better characterize the role of

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Address correspondence to Dr Berry, Box 3652, Duke University Medical Center, Durham, NC 27710; e-mail: berry037@mc.duke.edu.

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minimally invasive resection techniques for these conditions. All pathology reports were reviewed in detail to confirm pathologic diagnosis and exclude malignancy. Any postoperative event prolonging or otherwise altering the postoperative course was recorded. Operative morbidity was defined as the occurrence of any of these events. Operative deaths were defined as deaths that occurred within 30 days after operation or those that occurred later but during the same hospitalization.

Results

During the study period, 22 patients who underwent surgical resection of a congenital pulmonary abnormality after the age of 10 years were identified. The demographics and characteristics of the patient population are given in Table 1. The most common malformation in this study was pulmonary sequestration (n = 14, 64%), followed by CCAM (n = 2, 9%) and bronchial agenesis (n = 2, 9%).

Eighteen patients (82%) had symptoms that initiated a workup that ultimately led to the diagnosis of a congenital pulmonary abnormality. The presenting symptoms that led to the diagnosis are shown in Table 2. The median duration of symptoms before operation was 14 months (range, 1 month to 27 years). These symptoms required a total of six emergency room visits and six inpatient hospitalizations before surgical management in 11 patients. Four (18%) of the patients were incidentally found to have a congenital abnormality during workup of the following conditions: pulmonary nodules found on routine chest roentgenography (n = 2), nephrolithiasis (n = 1), and esophageal cancer (n = 1). The median age at diagnosis was 36 years (range, 10 to 66 years). The median age at the time of operation was 42 years (range, 13 to 71 years). Sixteen patients underwent surgical treatment within 1 year of diagnosis.

The details of the surgical management of the specific congenital pulmonary malformations are given in Table 3. Of the 22 surgical procedures, 21 were performed electively. One nonelective operation was performed in a patient with congenital right upper lobe emphysema who was transferred from another hospital with a large pneumothorax and a moderate air leak that did not resolve with a chest tube. A thoracoscopic right upper lobectomy was performed within 24 hours of admission.

Table 1. Patient Demographics

Characteristic	Median (range)
Male/female	11/11
Race	
White	21
African American	1
Age at diagnosis (years)	36 (10–66)
Age at operation (years)	42 (13–71)
BMI (kg/m^2, n = 16)	24.6 (18.1–40.7)

BMI = body mass index.

Table 2. Presenting Symptoms in 22 Patients

Symptom	No. of Patients (%)	Median Symptom Duration, Months (Range)
Shortness of breath	6 (22%)	9 (2.5–180)
Hemoptysis	4 (15%)	18 (1–36)
Cough	3 (11%)	9 (6–24)
Pneumonia	3 (11%)	96 (24–240)
Pleuritic chest pain	2 (7%)	164.5 (5–324)
Lethargy	2 (7%)	5 (4–6)
Abscess	2 (7%)	9 (2–16)
Pneumothorax	1 (4%)	0.1
Asymptomatic	4 (15%)	22 (3.5–36)

Some patients had multiple symptoms.

Seven surgical procedures (32%) were completed with resection through a thoracotomy (Table 3A). All vascular malformations requiring pneumonectomies were performed by thoracotomy. Of these open procedures, a left pneumonectomy for absence of the left pulmonary artery was performed at the time of an Ivor Lewis esophagogastrectomy for esophageal cancer, and a right pneumonectomy for aberrant right pulmonary artery origin was performed at the time of tricuspid valve repair with atrial septal defect repair. A minimally invasive approach with thoracoscopy was performed in 15 (68%) patients (Table 3B). Ten of 14 patients (83%) with pulmonary sequestrations had resections performed with thoracoscopy.

The median duration for isolated pulmonary operation was 2.13 hours (range, 1.13 to 3.10 hours) with median estimated blood loss of 50 mL (range, 5 to 750 mL). No intraoperative surgical complications were noted, and none of the thoracoscopic procedures required conversion to thoracotomy. In 1 patient with an intralobar pulmonary sequestration, a planned thoracoscopic segmentectomy was converted to a lobectomy because the residual lobar tissue was devitalized. The arterial supply for the 12 pulmonary sequestrations arose from the following: six from the thoracic aorta, one from the celiac artery, two from the abdominal aorta, and three were unspecified. A representative computed tomography image of pulmonary sequestration of the left lower lobe with a large aortic feeding branch is shown in Figure 1. This was successfully managed with a thoracoscopic resection. One of the major difficulties in surgical resection is identification of aberrant anatomy. Figure 2 demonstrates thoracoscopic identification of the feeding artery after careful dissection of the pulmonary ligament. The arterial branch was found in the pulmonary ligament in 8 of these 12 patients, and the locations in the rest were unspecified. All pathology reports were reviewed to confirm diagnosis and absence of malignancy.

There were no perioperative deaths. The median postoperative chest tube duration was 2 days (range, 1 to 15 days). The median postoperative hospital stay was 3 days (range, 2 to 15 days). A total of five postoperative events occurred in 4 patients: 2 with episodes of fever,