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Original Article

The Surgical Management Strategies in Congenital Pulmonary Airway Malformations: According to the Location of the Pulmonary Involvement

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ABSTRACT

Aim: Treatment of congenital pulmonary airway malformations (CPAMs) consists of different surgical options. In this study, we aimed to report our surgical management strategy according to the location of pulmonary involvement.

Materials and Methods: We retrospectively analyzed the medical records of patients who underwent surgery for CPAMs between 2005 and 2015. The data including patient demographics, pre-operative clinical features, surgical management strategies and postoperative results were reviewed.

Results: Twenty patients (14 male, 6 female) with a median age of 4 months (1 day-12 years) were operated on. Antenatal diagnosis was positive in 12 patients (60%). The other patients were admitted with a median age of 3.5 years (1 day-12 years). Respiratory infection was seen in four patients (20%). Nine patients underwent early operation due to severe dyspnea and recurrent respiratory infection (45%). Lobectomy was performed on 17 patients with single lobe involvement (85%). Lobectomy for major lesion and segmentectomy for minor lesion was performed on two patients with unilateral multi-lobar involvement. One patient with bilateral multi-lobar involvement required multiple thoracoscopic wedge resections. Two patients who had severe dyspnea before surgery required mechanical ventilation after the operation, and one of them died. Two postoperative complications, empyema and pneumothorax were seen. Mean postoperative follow-up period was 5.5 years.

Conclusion: CPAMs must be excised totally due to the risk of pulmonary infection and malignancy. The resection strategy should be decided according to the number of the affected lobes. Lobectomy should be performed in single lobar involvement. Unilateral multi-lobar involvement requires lobectomy for a major lesion and segmentectomy for a minor one. Thoracoscopic multiple wedge resections should be the option in bilateral multi-lobar CPAMs.

Keywords: Congenital pulmonary airway malformation, congenital cystic adenomatoid malformation, lung cyst, children

Introduction

Congenital cystic adenomatoid malformations (CCAMs) are congenital lung malformations derived from a hamartomatous lesion of the bronchial tree (1). These lesions account for approximately 95% of congenital cystic lung diseases (2). In recent years the name of this clinical entity was changed to congenital pulmonary airway malformations (CPAMs). This new name has gained popularity and is primarily preferred in the current medical literature (3). The course of these lesions could be varied in a broad clinical spectrum as hydrops and fetal death due to rapid growing, or being asymptomatic without clinical problems (4,5). Recurrent pulmonary infections, pnemumotorax and primary lung malignancies are the potential clinical problems which could be seen in the postnatal period (6,7). Pulmonary blastoma, rabdomyosarcoma, bronchogenic carcinoma and various sarcomas were reported in the literature which got derived from CPAMs (8-11). Therefore, surgical resection is usually suggested to avoid these potential devastating clinical problems. Lobectomy is usually preferred in the surgical treatment of CPAMs in order to avoid leaving

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residual disease in the remaining lobe (3). However, the management of CPAMs could be a big challenge in patients with multilobar involvement. Formal lobectomies could result with pneumonectomy in these patients (12). In this study, we aimed to present our surgical management strategies in CPAMs according to the different locations of the pulmonary involvement.

Materials and Methods

The medical records of patients who underwent surgery for CPAMs between 2005 and 2015 were retrospectively analyzed. Data including age, time of clinical admission, respiratory problems, surgical resection strategies according to anatomic locations of the cystic lesions and postoperative complications were reviewed. The time of surgical resection was postnatal 1 to 6 months old in patients with antenatal diagnosis of CPAMs. However, early surgery was required in some of these patients in the early postnatal period due to severe respiratory defficiency. The other patients who were admitted after infancy period underwent elective surgical excision without any delay. The main surgical approach was thoracotomy. Thoracoscopy was preferred in only one patient with bilateral multi-lobar involvement. The surgical strategy was decided according to the anatomic location of the pulmonary involvement;

- Single lobar involvement: Lobectomy.
- Unilateral multi-lobar involvement: Lobectomy to the major lesion, segmentectomy to the minor.
- Bilateral multi-lobar involvement: Thoracoscopic wedge resections in multiple sessions.

All of the parents gave their informed consent prior to their inclusion in the study.

Results

Twenty patients (14 male, 6 female) with a median age of 4 months (range: 1 day-12 years) were operated on. The time of clinical admission was early postnatal period for 12 patients who had antenatal diagnosis (60%). The other eight patients who had not had antenatal diagnosis were admitted with a median age of 3.5 years (range: 1 day-12 years). Main results are summarized in Table I. Pre-operative respiratory infection was seen in four patients (20%). Nine patients underwent early operation due to severe dyspnea and recurrent respiratory infection (45%). All of the patients were examined by computerized tomography after neonatal period as a routine pre-operative work-up. The most frequent location for pulmonary involvement was left lower lobe (40%). The other locations were left upper lobe (20%), right middle lobe (15%), right upper lobe (10%), unilateral multilobar (10%), bilateral multilobar involvement (5%) (Figure 1). Operative strategy was decided according to the location of the pulmonary involvement of the cystic lesions. Lobectomy was performed on 17 patients with single lobar involvement (85%). Lobectomy to major cystic lesion and segmentectomy to minor lesion was performed in two patients with unilateral multi-lobar involvement (10%). One patient with bilateral multi-lobar involvement required multiple thoracoscopic wedge resections (5%) (Figure 2). Two patients who had severe dyspnea before surgery required mechanical ventilation after the operation. Mortality was seen in one of these patients due to severe respiratory insufficiency (5%). Empyema and pneumothorax were two postoperative early complications which were treated succesfully. Recurrence, infection or malignancy was not observed in any of the

Table I. The data of patient demographics and clinical features of overall group

Number of patients	20 patients (14 M, 6 F)
Median age of operation	4 months (1 day-12 years)
Antenatal diagnosis	60% (12 patients)
Median admission age of patients without antenatal diagnosis	3.5 years (1 day-12 years)
Clinical presentations	
Asymptomatic (n=8) Dyspnea (n=6) Respiratory infections (n=4)	40% 30% 20%
Early operation due to severe dyspnea and recurrent respiratory infection	45% (9 patients)
Operative strategy	
Single lobar involvement (n=17)	Lobectomy
Unilateral multi-lobar involvement (n=2)	Major lesion: Lobectomy Minor lesion: Segmentectomy
Bilateral multi-lobar involvement (n=1)	Thoracoscopic wedge resections in multiple sessions

F: Female, M: Male

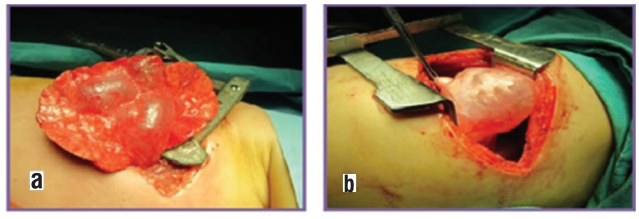

Figure 1. a) The lesion was identified in the right middle lobe and exteriorized through the thoracotomy incision. b) A huge cystic lesion was detected in the left upper lobe

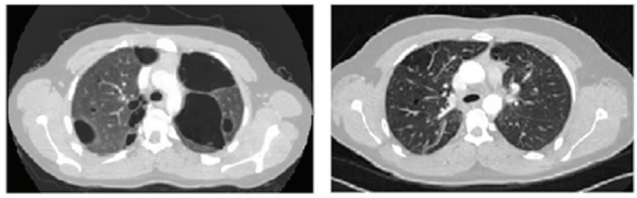

Figure 2. Pre-operative and postoperative computed tomography scans of a patient with bilateral multi-lobar pulmonary involvement of cystic lesions
CT: Computed tomography

patients during follow-up. Mean postoperative follow-up period was 5.5 years.

Discussion

The management strategies in CPAMs are usually based on potential detrimental clinical problems as recurrent pulmonary infections and rarely malignancy. A case was also reported from our clinic in recent years with primary pulmonary rhabdomyosarcoma arising within cystic adenomatoid malformation (11). Lobectomy is the most commonly preferred surgical technique to resect all of the cystic lesions to avoid residual disease. In recent years, non-operative management with clinical follow-up has begun to be advocated in some of the centers due to the frequently benign asymptomatic course of these lesions (13,14). However, the follow-up strategy is not clear. The interval periods between the radiological imaging studies and the type of the imaging modalities as computerized tomograghy or chest X-ray in the follow-up are not clearly defined in the literature, nor is the management strategy in patients with multi-lobar pulmonary involvement (12). Surgical excision is usually suggested to avoid potential clinical problems. Kapralik et al. (15) reported a systematic review and meta-analysis to discuss surgical versus conservative management of CPAMs in children. They suggested elective resection of asymptomatic lesions to avoid development of symptoms. Optimal age for surgery was not clearly defined in the reported studies. Naito et al. (16) reported that earlier lobectomy did not effect the pulmonary functions in the long-term. Sullivan et al. (17) discussed the optimal age for elective surgical resection for asymptomatic CPAMs with meta-analysis. They concluded that the current scientific evidence is not enough to suggest a conclusive recommendation for an optimal age for the timing of elective resection. We performed surgery in an earlier stage of infancy, namely 1-6 months old to reduce the risk of later complications like respiratory infections and pneumothorax (18). Formal lobectomy is the most commonly accepted surgical option for CPAMs in the current medical era (3) although in recent years some authors have suggested lung-sparing techniques such as segmentectomy by thoracoscopy for these lesions (19,20). However, these strategies comprise an increased risk for an incomplete surgical excision. The risk of bringing residual disease was reported as 15% after segmental resection (21). This approach should be rationale for salvaging from radical surgical resections such as pneumonectomy for multi-lobar or bilateral disease (22). In our study, we performed lobectomy for the major lesion and segmentectomy for the minor lesion in the left lobe to avoid pneumonectomy. In another patient with bilateral multilobar disease, we preferred multiple sessions of thoracoscopic wedge resections. We have not seen any recurrence or malignancy in the postoperative follow-up period. This surgical strategy which is based on the location of pulmonary involvement provides a safe and effective treatment option in bilateral and multilobar disease. These patients should be followed by regular radiological imaging studies. We preferred chest X-rays with 3-month-intervals in the first postoperative year. A computerized tomography should be obtained at the end of the first year of postoperative period.

Study Limitations

Due to the retrospective nature of this study, there may have some limitations. Larger number of the patients should be more useful to evaluate the results of the operative strategies.

Conclusion

In conclusion, we suggested lobectomy for single lobar pulmonary involvement, and lobectomy for major lesion lesion; and segmentectomy for the minor lesion in unilateral multi-lobar involvement; and multiple thoracoscopic wedge resections for bilateral multi-lobar pulmonary involvement.

Ethics

Ethics Committee Approval: Retrospective study.

Informed Consent: All of the parents gave their informed consent prior to their inclusion in the study.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: E.D., Z.D., B.T., C.Ö., A.E., Concept: E.D., Design: E.D., Data Collection or Processing: B.T., Analysis or Interpretation: E.D., C.Ö., A.E., Literature Search: E.D., Z.D., Writing: E.D.

Conflict of Interest: None of the authors had conflict of interest.

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