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Minimally invasive surgery for congenital cystic adenomatoid malformations - early experience

So Hyun Nam, Min Jeong Cho[1], Dae Yeon Kim[2]

Department of Surgery, Dong-A University Hospital, Dong-A University College of Medicine, Busan, [1]Division of Surgery, Konkuk University Medical Center, Seoul, [2]Department of Pediatric Surgery, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea

Purpose: The aim of this study is to present our experience with minimally invasive surgery (MIS) for congenital cystic adenomatoid malformations (CCAMs).
Methods: The medical records of infants under 2 years of age who underwent operation for a CCAM from 2009 to 2014 were retrospectively reviewed.
Results: MIS (9 of thoracoscopy and 1 of laparoscopy) was performed for 10 infants (male:female = 7:3) with CCAM. CCAM were discovered prenatally around gestational age of 24.7 weeks. The median gestational age was 38.6 weeks, and the median body weight was 2,817.5 g. None had respiratory distress after birth. The median age at the time of operation was 0.94 years (range: 8 days–1.66 years). Two underwent the operation during the neonatal period; one because of a coexisting large esophageal duplication cyst and the other due to diagnostic uncertainty. While awaiting operation, 5 of CCAM had grown without respiratory symptoms, and 2 infants had experienced pneumonia. The mean operative time was 98 minutes (range: 70–227 minutes), and there were no conversions or perioperative complications. The infants resumed enteral feeding within 2 days and were discharged within 7 days, except for 1 infant who underwent esophageal duplication cyst excision. During the follow-up period, there were no cases of either remnant lesions or respiratory symptoms.
Conclusion: MIS for CCAMs is safe and feasible, with excellent cosmesis and short hospital stays. Increasing experience with various MIS procedures will widen the indications for MIS in lung pathology.
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Key Words: Congenital cystic adenomatoid malformation of lung, Thoracoscopy, Minimally invasive surgical procedures, Bronchopulmonary sequestration

INTRODUCTION

The safety of laparoscopic interventions in pediatrics has been confirmed over the past 10 years [1]. Currently, the spectrum of minimally invasive surgeries extends to thoracoscopic surgery, even in neonates. Over several years, the utility and feasibility of thoracoscopic procedures for lung pathology have extended from lung biopsy to lobectomy [1-6]. We began to perform thoracoscopic repair for congenital diaphragmatic hernias in neonates in 2008. After gaining experience with thoracoscopy, we attempted to manage congenital lung disease, including pulmonary sequestration and congenital cystic adenomatoid malformations (CCAMs). The aim of this study is to present our experience with minimally invasive surgery (MIS) for CCAM.

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Corresponding Author: Dae-Yeon Kim
Department of Pediatric Surgery, Asan Medical Center, University of Ulsan College of Medicine, 88 Olympic-ro 43-gil, Songpa-gu, Seoul 05505, Korea
Tel: +82-2-3010-3961, **Fax:** +82-2-474-9027
E-mail: kimdy@amc.seoul.kr

METHODS

The records for 10 infants who underwent operative treatment for CCAMs between March 2009 and March 2014 were retrospectively reviewed. The median follow-up period was 50.5 months (range, 17.7–77.6 months). We excluded cases of thoracoscopic excision for pulmonary sequestration. The medical records were reviewed for gestational age, sex, body weight, Apgar score, associated anomalies, and associated respiratory symptoms. The operative details that were recorded included the age at operation, the location of the CCAM, operative time, postoperative ventilator days, time to enteral feeding, length of hospital stay, postoperative complications, and pathologic findings.

The operative methods are as follows. For the thoracoscopic procedure, the patient was positioned in the lateral decubitus position with the affected side elevated. The 1st trocar was inserted at the midaxillary line so that the level of the trocar's tip was parallel with the tip of the scapula. The CO_2 was insufflated gradually to reach a pressure of 3 mmHg. Next, a 5-mm 30° telescope was inserted. The 2nd and 3rd working ports were inserted medially and laterally to the 1st trocar under direct visualization to prevent interference between the working instruments. After identifying the lesion in the lower lobe, we excised the CCAM using LigaSure (Covidien, Mansfield, MA, USA) or an ultrasonically activated scalpel. The specimens were removed with a retrieval bag via the initial 5-mm port incision after enlarging the incision. A chest tube was placed via another working port. For the laparoscopic operations, the patient was positioned in the lithotomy position. The surgeon stood between the patient's legs. After creating a pneumoperitoneum, we explored the abdominal cavity and identified the infradiaphragmatic mass. The procedures for excision and retrieval were identical to those in the thoracoscopic operation.

RESULTS

Ten infants (male:female = 7:3) underwent MIS for CCAM (9 thoracoscopies and 1 laparoscopy). All of the CCAM cases were discovered prenatally at a gestational age of approximately 24.7 weeks. Associated symptoms such as polyhydramnios and fetal deceleration were not discovered. The median gestational age at birth was 38.6 weeks (range, 32^{+2} to 40^{+6} weeks) and the median body weight was 2,817.5 g (range, 2,150–3,700 g). None of the infants had respiratory distress after birth. The patients had median Apgar scores of 8 and 9 at 1 and 5 minutes, respectively. One infant had a cardiac anomaly (subvalvular aortic stenosis) and an esophageal duplication cyst. The median age at operation was 0.94 years (range, 8 days to 1.66 years). Two infants underwent the operation in the neonatal period. One infant had a coexisting large esophageal duplication cyst, and a second infant required a differential diagnosis for a retroperitoneal mass. While awaiting operation, 5 of the 8 infants' lesions grew without respiratory symptoms, and 2 infant had an episode of pneumonia. All of the CCAMs were single lesions and were located in the right lower lobe in 6 cases and in the left lower lobe in 4 cases. Nine operations were performed with thoracoscopy. One of the 10 infants had a laparoscopic operation to remove an infradiaphragmatic mass that was actually determined to originate in the left lower lobe. The mean operative time was 98 minutes (range, 70–227 minutes),

Table 1. The demographic findings of patients

Patient	Gestational age (wk)	Sex	Associated anomaly	Birth weight (g)	AS1	AS5	Symptoms	Mass size
1	38^{+6}	M	No	3,350	8	9	No	15 mm → 45 mm
2	39^{+2}	F	No	3,085	8	9	No	No change
3	37^{+6}	F	No	2,865	3	8	No	13 mm → 36 mm
4	40^{+6}	M	No	3,375	8	9	No	17 mm → 37 mm
5	38^{+4}	M	Subvalvular aortic stenosis & esophageal duplication cyst	2,710	9	9	No	No change
6	32^{+2}	M	No	2,150	8	9	1 Episode of pneumonia	No change
7	40^{+1}	M	No	2,385	8	9	No	15 mm → 55 mm
8	35^{+3}	M	No	2,410	9	10	1 Episode of pneumonia	No change
9	38^{+3}	F	No	2,770	9	9	No	No change
10	38	M	No	3,700	9	10	No	No change
Median	38.6	M:F=7:3	-	2,817.5	8	9	-	-

AS1, Apgar score 1 minute; AS5, Apgar score 5 minutes.

Table 2. The operative method and results

Patient	Method	Op. age (day)	Location	Pathology	Feeding artery	Draining vein	Post op ventilator (day)	Op. time (min)	Time to feed (day)	Chest tube (day)	Hospital days
1	T	210	RLL	CCAM	-	-	2	216	2	2	6
2	L	8	LLL	CCAM	-	-	2	227	2	0	7
3	T	370	RLL	CCAM & PS	Descending aorta	Intrapulmonary vein	1	100	1	3	5
4	T	609	LLL	CCAM	-	-	0	96	2	6	7
5	T	10	RLL	CCAM & esophageal duplication cyst	-	-	0	88	7	4	16
6	T	459	RLL	CCAM	-	-	0	83	1	4	5
7	T	401	RLL	CCAM & PS	Descending aorta	Inferior pulmonary vein	0	177	1	5	6
8	T	442	RLL	CCAM	-	-	0	120	1	4	5
9	T	317	LLL	CCAM & PS	Descending aorta	Azygos vein	0	70	0	4	6
10	T	134	LLL	CCAM	-	-	0	77	0	4	6
Median	-	0.94 yr	-	-	-	-	0	98	1	4	6

Op., operation; T, thoracoscopic excision; RLL, right lower lobe; CCAM, congenital cystic adenomatoid malformation; L, laparoscopic excision; LLL, left lower lobe; PS, pulmonary sequestration.

and there were no conversions or perioperative complications. The ventilator was used in 3 patients postoperatively, but the patients were weaned from the ventilator within 2 days. The chest tube was removed at a median of 4 days (range, 2–6 days after the operation). The infants received enteral feeding within 2 days and were discharged within 7 days, except for 1 infant who underwent the excision of an esophageal duplication cyst. Pathologically, 7 were type 1 CCAMs and 3 were type 2 CCAMs. Three out of the 10 CCAMs were hybrid lesions with intralobar pulmonary sequestration. Feeding artery was descending aorta for all of pulmonary sequestration, and draining vein were intrapulmonary vein and azygos vein. There was no mucin containing cell in the lesion. There were no remnant lesions or respiratory symptoms during the follow-up period (Tables 1, 2).

DISCUSSION

CCAM and pulmonary sequestration are the most common intrathoracic masses observed in prenatal ultrasonography [1]. CCAM is believed to result from an arrest in lung development [7]. The incidence of CCAM is between 1 in 10,000 and 1 in 35,000 in Australia and Canada [8,9]. Most of CCAM do not associate with severe respiratory distress after birth, but larger lesions can cause respiratory insufficiency making mediastinal shift and hydrops [10]. The symptoms of CCAM that do not present during infancy include repeated chest infections, bronchiectasis, lung abscesses, hemoptysis, pneumothorax, air embolisms, hemothorax, pyopneumothorax, steroid-resistant asthma, or rarely, malignant transformation [10-12].

The need for the surgical treatment of CCAM is well established, even though CCAM cases are asymptomatic. The overall complication rate beyond the neonatal period was estimated to be 3.2%, occurring at a median age of 7 months [12,13]. Studies from Canada reported that most asymptomatic patients become symptomatic (80%) if their lesions are not removed [14]. Infection is the most common presenting symptom [15]. Additionally, CCAMs have the potential for malignant transformation. Transformation is rare, but a review by Laberge et al. [15] reported 5 cases of pleuropneumoblastoma, 7 cases of rhabdomyosarcoma, and 8 cases of bronchioalveolar carcinoma in the literature [10,16].

When we do the operation, and how to operate upon these lesions must be determined. However, the timing of the operation is controversial. Calvert et al. [7] strongly recommend that the operation should be performed at approximately 3 to 6 months of age. These authors experienced a higher incidence of inflammation and infection with surgery performed after 6 months of age, making the surgery itself technically more difficult. Stanton et al. [12] determined that the average time of onset of symptoms in prenatally detected cases was at approximately 7 months of age, or 10 months of age when

considering all infants. Therefore, operative treatment should be pursued before 10 months of age. In our experience with a small number of patients, approximately 12 months of age is acceptable to the surgeon when considering technical feasibility and is also the acceptable age to the parents from the standpoint of their willingness to have their children undergo the operation. There was only an episode of infection in 2 patients while waiting for the operation, and we had no difficulties due to adhesions in this patient. However, the extent of the CCAMs increased in more than half of the cases during the observational period. Two neonates underwent the operation early, one because of diagnostic uncertainty and one because of a coexisting esophageal duplication cyst. Thoracoscopic operations are safe and feasible even in neonates. Many surgeons recommend elective surgery rather than an emergency operation. An elective operation has definitively good surgical outcomes [1,7,10,12]. Moreover, complications after emergency surgery were 2.8 times more likely to occur than after elective surgery [12].

With respect to the extent of the surgery, the standard procedure for CCAMs is lobectomy [16-20]. There is concern regarding potential infectious complications with a hidden residual lesion as well as malignant transformation during the follow-up period. Additionally, the parenchyma-saving resection can be complicated by a prolonged air leak in the early postoperative period [21]. However, the treatment for multifocal CCAM is somewhat complicated. In addition, CCAMs are generally diagnosed in neonates and infants who have small lung volumes. For this reason, the optimal extent of resection in patients with CCAMs has been controversial. Muller et al. [16] observed that the extent of CCAM on CT is very different than the extent on a pathologic specimen. Moreover, the sensitivity of CT is 33%, the specificity is 100%, and the negative predictive value is 60%. A previous study recommends an atypical pulmonary resection, reserving a parenchymal-saving operation only for multifocal CCAM lesions.

However, in theory, if the parenchyma-saving resection is safe, preserving the normal pulmonary parenchyma would be ideal in neonates and infants because they have growth potential [21]. Kim et al. [21] compared 2 groups of subjects who underwent either lobectomy or parenchyma-saving resection for CCAM. There were no significant differences in the incidence of early postoperative complications or late morbidity between the 2 groups. A complicated air leak was more common in the group with parenchyma-saving resection, but the difference between the 2 groups was not statistically significant. In our series, we performed a segmentectomy using a LigaSure for the CCAM in all of the patients to preserve the lung parenchyma. Fortunately, there was no residual disease on the CT and no associated symptoms during the follow-up period. We cannot advocate segmentectomy as a standard procedure based only on our small series with a short follow-up period. However, we found that the complication rate was less than that reported in the literature. Getting the experiences including more cases could justify the recommendation for a parenchyma-saving resection.

Conventional thoracotomy is the accepted treatment for CCAM. However, MIS has changed the trends in surgery, even for lung pathology. For the pediatric surgeon, the extension from laparoscopy to thoracoscopy is not difficult. There are limited studies comparing thoracotomy and thoracoscopy. However, many surgeons report good clinical outcomes beyond feasibility and safety [1,4-6]. The rate of postoperative complications after thoracoscopy is comparable with that of complications after thoracotomy. Thoracoscopic procedures contribute to shorter hospital stays with less pain and better cosmesis than thoracotomy. Although the operating time for thoracoscopy tends to be longer than that for thoracotomy, the former procedure may be worthwhile because of the small scar, which may be particularly important for small children. Moreover, the musculoskeletal deformities present after thoracotomy can be prevented.

This review describes the benefits of thoracoscopic operations for CCAMs. Despite the small numbers of patients in this study and the starting period of thoracoscopic operation, there were no postoperative morbidities or residual lesions. The use of laparoscopy for infradiaphragmatic lesions is also helpful for diagnosing and treating cases of unusual radiologic findings. We cannot justify segmentectomy in CCAM, but we are confident that the thoracoscopic operation provides a good clinical outcome with minimal trauma in small children. As the use of thoracoscopic procedures increases, the understanding in cases with complicated pathology will also grow.

In conclusion, MIS for CCAMs is safe and feasible with excellent cosmesis and short hospital days. Gaining experience in various MIS procedures will expand its use for treating lung pathologies.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

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