Clinical characteristics and feasibility of thoracoscopic approach for congenital cystic adenomatoid malformation in adults

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Abstract

Objective: Congenital cystic adenomatoid malformation (CCAM) in adults is very rare, and surgical resection is the treatment of choice. We evaluated the clinical characteristics and feasibility and safety of video-assisted thoracic surgery (VATS) in adult patients with CCAM. Methods: Retrospective analysis of nine consecutive adult patients with CCAM treated surgically between January 1995 and April 2006 at a single center. The clinical characteristics and surgical outcome were evaluated retrospectively. Results: The records of five men and four women were examined. The median age at diagnosis was 30.5 years (range 16—44 years). The most frequent symptoms were cough (n = 4) and hemoptysis (n = 2). Three patients had recurrent respiratory infections. Chest computed tomography (CT) was reviewed in all cases. The lesions were on the right side in six patients and on the left in three. All patients had lesions in a single lobe, except one patient who had multi-lobar lesions. The CCAM appeared as thin-walled multi-septated cystic lesions in six patients and as parenchymal consolidations in three patients. Four patients underwent VATS lobectomy and four underwent lobectomy through a conventional open thoracotomy. One patient had open pneumonectomy. The median operating time (160 min in VATS lobectomy and 165 min in open lobectomy), median duration of chest tube drainage (4 days in VATS lobectomy and 6.5 days in open lobectomy), and postoperative complications (one patient in each treatment group) were similar between the VATS and open lobectomy groups. The hospital stay was shorter in the VATS lobectomy group (median 5 days) than in the open lobectomy group (median 7.5 days). No postoperative or procedure-related mortality occurred.

Conclusions: CCAM is rare in adults. Patients usually present with recurrent respiratory infections and thin-walled multi-septated cystic lesions. VATS is a feasible and safe technique to treat adult patients with CCAM.

1. Introduction

Congenital cystic adenomatoid malformation (CCAM) is an uncommon developmental anomaly of the lower respiratory tract, usually diagnosed in neonates and infants [1,2]. Its presentation in adulthood is rare, with fewer than 40 reported cases [3—7]. Adult patients with CCAM usually present with recurrent lower respiratory infection, although recurrent pneumothorax and no respiratory symptoms have also been reported [5,7].

Surgical resection is the treatment of choice, even in asymptomatic patients with CCAM, because patients are prone to pulmonary infections, and if left untreated, these lesions may undergo malignant transformation [5,8,9].

Keywords: Congenital cystic adenomatoid malformation of lung; Adult; Treatment; Surgery; Video-assisted thoracic surgery

Video-assisted thoracic surgery (VATS) is recognized as being as effective as open surgery for a variety of diagnostic and therapeutic conditions, with significantly less morbidity [10—12]. However, in the English literature, only one case report exists of VATS resection in the treatment of CCAM in an adult [13].

This study evaluated the clinical characteristics of adult patients with CCAM and compared the approach using VATS with open thoracotomy in terms of its feasibility, safety, and complications.

2. Patients and methods

2.1. Patients

We retrospectively reviewed the files of all adult patients diagnosed with CCAM and treated by surgical resection at the
Samsung Medical Center (a 1250-bed referral hospital in Seoul, Korea). In the period between January 1995 and April 2006, nine adults underwent surgery for CCAM. All the patients had preoperative chest computed tomography (CT). Four patients were treated using VATS procedures, and five patients underwent open thoracotomy. Of these five patients who underwent open thoracotomy, four had lobectomy and one underwent pneumonectomy. The diagnosis was confirmed in all cases after histopathological examination of the resected lung.

The following data were collected from the medical records and chest radiographs: major symptoms, diagnostic procedures, surgical treatment, and outcome. The operative parameters and findings were evaluated, including operating time, duration of chest tube drainage, duration of hospital stay, and complications.

The Institutional Review Board gave permission to review and publish the patient records retrospectively.

2.2. Indications for VATS lobectomy

VATS procedure for benign pulmonary disease was started from January 1998, and VATS lobectomy without rib spreading had been carried out since December 2003 in our institute. In this study, two patients (cases 1 and 2) underwent open thoracotomy before the start of VATS procedure. Three patients (cases 4, 6, and 8) who had wide pleural-based lesion on chest CT and were expected to be associated with pleural adhesion underwent open thoracotomy. Two patients (cases 3 and 5) underwent lobectomy through minithoracotomy with video assistance before December 2003, and two patients (cases 7 and 9) underwent VATS lobectomy without minithoracotomy after December 2003.

2.3. Operative technique

The patients were placed in the lateral decubitus position and single-lung ventilation was achieved in all cases using a double-lumen endotracheal tube.

For thoracoscopic lobectomy, the operating surgeon stood at the patient’s right side facing a monitor independent of the lesion side. We used 30° thoracoscope camera with one working window and two ports for the thoracoscopic lobectomy. Anterolateral minithoracotomy was performed on the fifth intercostal space in two cases undergoing VATS lobectomy (cases 3 and 5). The pulmonary lobar vessels were dissected and divided with ligation and sutures or a stapling device. The bronchus was divided with a stapling device. Once the lobectomy was completed, the resected lung was retrieved through working window using a commercial vinyl bag.

In all patients undergoing open thoracotomy, a standard posterolateral thoracotomy was performed through fifth—sixth intercostals spaces, which varied with the surgical procedure. After thoracotomy, dissection of adhesion in pleura or fissure was performed. The lobar vessels and bronchus were dissected and divided with ligation and suture or a stapling device.

2.4. Postoperative management

Our policy of the drainage management after lung resection surgery was to remove the drainage tube as soon as possible. We usually removed the drainage tube 1 day after stopping air leak. After successful removal of drainage tube, all patients were discharged from hospital after 1 or 2 days, with the exception of one patient who was treated with open pneumonectomy. Because of uncontrolled pain in thoracotomy site, the patient was discharged 8 days after the removal of drainage tube.

3. Results

3.1. Characteristics of patients

The records of five men and four women were examined. The median age at diagnosis was 30.5 years (range 15—44 years). The major symptoms at presentation were a productive cough in four patients and hemoptysis in two patients. Three patients had no symptoms. The demographic characteristics of the patients are shown in Table 1. None had a family history of any congenital malformation. Three patients had undergone treatment for pulmonary tuberculosis. Two patients had a history of lung abscess and one had recurrent pneumonia and pneumothorax. The median duration from the first symptoms to confirmation of diagnosis at operation was 26.5 months (range 21—167 months).

Table 1
Clinical and histological data for nine adult patients with CCAM of the lung

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Age at symptom onset (years)</th>
<th>Clinical history</th>
<th>Lobe</th>
<th>Size (largest cyst, mm)</th>
<th>CCAM type</th>
<th>CT findings</th>
<th>Treatment (surgery type)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>16</td>
<td>Female</td>
<td>15</td>
<td>Pulmonary TB</td>
<td>LUL and LLL</td>
<td>85</td>
<td>1, 2</td>
<td>Multi-septated air-filled cysts</td>
<td>Pneumonectomy (Open)</td>
</tr>
<tr>
<td>2</td>
<td>19</td>
<td>Male</td>
<td>No symptoms</td>
<td>None</td>
<td>RLL</td>
<td>30</td>
<td>2</td>
<td>Consolidation</td>
<td>Lobectomy (Open)</td>
</tr>
<tr>
<td>3</td>
<td>15</td>
<td>Male</td>
<td>13</td>
<td>Lung abscess</td>
<td>RLL</td>
<td>25</td>
<td>1</td>
<td>Multi-septated air-filled cysts</td>
<td>Lobectomy (VATS)</td>
</tr>
<tr>
<td>4</td>
<td>31</td>
<td>Female</td>
<td>29</td>
<td>Pneumothorax recurrent pneumonia</td>
<td>LLL</td>
<td>30</td>
<td>2</td>
<td>Consolidation</td>
<td>Lobectomy (Open)</td>
</tr>
<tr>
<td>5</td>
<td>39</td>
<td>Male</td>
<td>No symptoms</td>
<td>None</td>
<td>RLL</td>
<td>95</td>
<td>2</td>
<td>Multi-septated air-filled cysts</td>
<td>Lobectomy (VATS)</td>
</tr>
<tr>
<td>6</td>
<td>25</td>
<td>Male</td>
<td>23</td>
<td>Pulmonary TB</td>
<td>RLL</td>
<td>50</td>
<td>1</td>
<td>Multi-septated fluid-filled cysts</td>
<td>Lobectomy (Open)</td>
</tr>
<tr>
<td>7</td>
<td>44</td>
<td>Female</td>
<td>No symptoms</td>
<td>None</td>
<td>RLL</td>
<td>40</td>
<td>2</td>
<td>Consolidation</td>
<td>Lobectomy (VATS)</td>
</tr>
<tr>
<td>8</td>
<td>30</td>
<td>Male</td>
<td>28</td>
<td>Lung abscess</td>
<td>RUL</td>
<td>80</td>
<td>2</td>
<td>Multi-septated fluid-filled cysts</td>
<td>Lobectomy (Open)</td>
</tr>
<tr>
<td>9</td>
<td>32</td>
<td>Male</td>
<td>18</td>
<td>Pulmonary TB</td>
<td>LUL</td>
<td>15</td>
<td>1</td>
<td>Multi-septated air-filled cysts</td>
<td>Lobectomy (VATS)</td>
</tr>
</tbody>
</table>

CCAM, congenital cystic adenomatoid malformation; TB, tuberculosis; LLL, left lower lobe; RLL, right lower lobe; LUL, left upper lobe; RUL, right upper lobe; Open, open thoracotomy; VATS, video-assisted thoracic surgery.
Concerning the location of the lesions, six lesions were on the right and three were on the left. One patient had involvement in multiple lobes. The size of the lesions ranged from 15 to 95 mm (median 40 mm) on the chest radiographs.

On chest CT, six patients had thin-walled multi-septated cystic lesions. Four of these six patients had multi-septated air-filled cystic lesions and two had multi-septated fluid-containing cystic lesions. All of these lesions were interpreted as CCAM by the chest radiologists. Interestingly, in the other three patients, the chest CT showed mainly parenchymal consolidations with no definite multi-septated cystic lesions and was interpreted as bacterial pneumonia or tuberculosis by the chest radiologists. No significant enhancing lesions were observed in any patient after administering intravenous contrast material. Six lesions had a base with the pleural surface, although no patient had a pleural effusion. Representative CT images and histological features are presented in Figs. 1 and 2.

Four patients were treated with thoracoscopic procedures only, and all of them had lobectomies. Conversion of the VATS approach to open thoracotomy was not necessary in these patients. Five patients underwent open thoracotomies. Of these five patients, four had lobectomies and one underwent pneumonectomy, as the lesion affected the left upper and lower lobes.

On pathological examination, three patients were found to be CCAM type 1 and five to be CCAM type 2 according to Stocker’s classification [1]. The patient whose lesion affected two lobes had mixed CCAM with types 1 and 2. In all patients, no associated malignancy was observed in the histological studies of the surgical specimens.

### 3.2. Treatment outcome and morbidity

Table 2 shows the treatment outcome according to surgical procedure in eight patients undergoing VATS or open lobectomy. One patient, who underwent open pneumonectomy, was excluded from this analysis. The operating time and duration of tube drainage were similar in the patients who underwent VATS and open lobectomy. The amount of blood loss was more in the VATS lobectomy group (median 600 ml, range 400—600 ml) than in the open lobectomy group (median 400 ml, range 250—400 ml), although this was not statistically significant ($p = 0.099$). The duration of hospital stay was shorter in the patients who underwent VATS (median 5 days, range 3—13 days) than in the patients treated with open lobectomy (median 7.5 days, range 5—21 days), although this was also statistically nonsignificant ($p = 0.384$).

<table>
<thead>
<tr>
<th></th>
<th>VATS lobectomy ($n = 4$)</th>
<th>Open lobectomy ($n = 4$)</th>
<th>$p$-value*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Operating time (min)</td>
<td>160 (130—180)</td>
<td>165 (140—210)</td>
<td>0.661</td>
</tr>
<tr>
<td>Duration of tube drainage (days)</td>
<td>4 (2—10)</td>
<td>6.5 (2—18)</td>
<td>0.663</td>
</tr>
<tr>
<td>Duration of postoperative hospitalization (days)</td>
<td>5 (3—13)</td>
<td>7.5 (5—21)</td>
<td>0.384</td>
</tr>
<tr>
<td>Estimated blood loss (ml)</td>
<td>600 (400—600)</td>
<td>400 (250—400)</td>
<td>0.099</td>
</tr>
<tr>
<td>No. of complications</td>
<td>1 (25%)</td>
<td>1 (20%)</td>
<td>1.000</td>
</tr>
</tbody>
</table>

The data are presented as the number (%) or median (range). VATS, video-assisted thoracic surgery.

* Mann–Whitney U-test or Fisher’s exact test.

![Fig. 1. (a) A 39-year-old man with CCAM. Chest CT shows multi-septated air-filled cystic lesions in the right lower lobe. (b) A 25-year-old woman with CCAM. Chest CT shows multi-septated fluid-filled cystic lesions in the right lower lobe. (c) A 19-year-old man with CCAM. Chest CT shows mainly parenchymal consolidation in the right lower lobe.](image-url)
Two postoperative complications occurred: a pneumothorax after removing the chest tube in one patient who underwent VATS lobectomy and a persistent air leak of the chest tube for 10 days postoperatively in a patient who was treated with open lobectomy. No procedure-related mortality occurred.

4. Discussion

CCAM is a congenital pulmonary development disorder. It is a hamartomatous lesion that consists of cystic and adenomatous overgrowth of terminal bronchioles and occasionally the airspaces [1,2]. It causes respiratory distress in infants. In adults, CCAM most commonly presents as recurrent pulmonary infection [3,5], but may be asymptomatic for years and diagnosed from routine chest X-ray in some cases [4]. Interestingly, one-third of our patients (3/9) presented with an abnormal chest X-ray and no symptoms.

Patz et al. [3] found that most frequent findings on chest CT of CCAM in adults were air-filled cystic lesions (5/7, 71%), and six lesions had a broad pleural base with no pleural effusion. In our study, the most frequent chest CT findings were also air-filled cystic lesions (6/9, 67%), and the lesions with a pleural base had no effusions. Interestingly, chest CT showed parenchymal consolidation without definite multi-septated cystic lesions in one-third of our patients. Two of these three patients had no respiratory symptoms. The radiologists had misdiagnosed these lesions as other diseases, such as organizing pneumonia or tuberculosis.

It is generally accepted that surgical resection is the treatment of choice in CCAM. Despite the lack of symptoms, patients with CCAM are prone to recurrent pulmonary infection, and the lesion can undergo malignant transformation [5,8,9]. For these reasons, CCAM is resected electively and pulmonary lobectomy via a posterolateral thoracotomy incision is the standard surgical procedure.

In recent years, VATS has become an increasingly important tool in thoracic surgery. The complications and morbidity associated with standard posterolateral thoracotomy are avoided with the thoracoscopic approach. Many studies in the adult population comparing thoracoscopy and thoracotomy highlight the benefits of VATS in terms of morbidity, pain control, hospital stay, and cosmetic result [10–12]. Recently, Cano et al. [14] reported the feasibility and safety of VATS in six infants and small children with CCAM. However, the feasibility of VATS in adult patients with CCAM has been reported in one patient only [13].

In this case series of nine adult patients with CCAM, we successfully performed VATS lobectomy in four with CCAM, and these patients had shorter hospital stays. In addition, the results were similar in terms of operating time, duration of chest tube drainage, and postoperative complications.

In conclusion, CCAM is a rare congenital malformation of the lower respiratory tract that may be diagnosed during adulthood. Patients usually present with recurrent respiratory infections and thin-walled multi-septated cystic lesions on chest CT. The VATS procedure is a safe and feasible technique in adult patients with CCAM.

Acknowledgement

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References