VANISHING LUNG SYNDROME AND LUNG VOLUME REDUCTION SURGERY - A CASE REPORT

Cheng-Hsiung Chen¹, Chin-Shui Yeh¹, Cheng-Huag Tsai¹, Bin-Chuan Ji¹, Fu-Yuan Siao², Jing-Lan Liu³, Woei-Horng Chai¹

Abstract

Development of multiple lung bullae [variously termed primary bullous disease of the lung, giant bullous emphysema (GBE), or vanishing lung syndrome (VLS)] is rare, and has been described in young male smokers. We presented a 41-year-old man with vanishing lung syndrome diagnosed via chest computed tomography (CT). He had a 22-year history of smoking 40 cigarettes a day, and had suffered from exertional dyspnea and productive cough with whitish sputum for 2 years. These symptoms had increased from one week ago. He was advised to stop smoking and received lung volume reduction surgery (LVRS). Post-operatively, significant improvements in dyspnea, spirometry values, lung volume measurement and chest radiography were observed. Three months postoperatively the patient was in good health and back at work. This case demonstrated that giant bullae can be successfully managed with surgical resection, and that their size can be determined by different techniques, including chest CT and lung volume measurements.

Key words: Bullous emphysema, Vanishing lung syndrome, Pneumothorax

Introduction

Bullous emphysema is an expansion of the alveolar space with a diameter over 1 cm and a wall thickness less than 1 mm.¹ In 1937, Burke described a case of vanishing lung in a 35-year-old man who experienced progressive dyspnea, impending respiratory failure, and radiographic and pathologic findings of giant bullae, which occupied two-thirds of both hemithoraces.² Since Burke’s original description, scattered cases have been reported. Bullae are most commonly seen in the upper portion of the lung. The underlying disease process is characterized by abnormal enlargement of the air space distal to the terminal bronchioles, accompanied by destruction of their wall. These anatomic changes result in decreased elastic recoil, increased airway resistance and decreased maximal expiratory rate.³ Lung volume reduction surgery or bullectomy can improve these pulmonary function changes.

Lung volume reduction surgery (LVRS) was developed as a means of surgical treatment for severe pulmonary emphysema. Preliminary results
of follow-up studies (up to five years) have already been published, indicating improved pulmonary function and quality of life after surgical treatment. However, the alarming results from the National Emphysema Treatment Trial (NETT) Research Group indicated a considerable risk for death in patients with homogenous emphysema and low forced expiratory volume in one second (FEV1) undergoing LVRS. Among the 511 non-high-risk patient in the NETT who underwent LVRS, the rate of operative mortality was 6 percent, major pulmonary morbidity was 30 percent, and the major cardiovascular morbidity was 20 percent. An air leak is a common adverse consequence of LVRS. Approximately 90 percent of patients will develop an air leak within 30 days of LVRS.

Giant bullae are generally resected in the setting of thoracoscopic surgery. Resection using an automated stapling device is popular. We report a case of bilateral upper lung giant bullous emphysema (GBE), also known as vanishing lung syndrome (VLS) with obvious symptoms; chest radiography and lung function showed improvement after thoracoscopic lung volume reduction surgery (LVRS).

**Case report**

The patient was a 41-year-old previously healthy man who complained of exertional dyspnea and cough with whitish sputum for the last 2 years. These symptoms progressed from one week ago. He had a 22-year history of smoking 40 cigarettes a day. On physical examination, there was evidence of hyperinflation with hyperresonance to percussion and an absence of breathing sounds from the mid-to-upper zones of the bilateral lung field. Chest radiography and chest computed tomography (CT) revealed giant bullae in both upper lung fields, occupying more than half of the hemithorax (Figure 1a and Figure 2a), and bilateral centrilobular bullous emphysema (Figure 2b). On spirometry, a severe obstructive and restrictive pattern was observed (Table 1). The His α1 antitrypsin level was normal.

He was advised to stop smoking and received lung volume reduction surgery via thoracoscopy. We removed bilateral multiple giant bullae via thoracoscopy and then pleurodesis was done. Histologically, it showed multicystic lesion lined by flat to cuboid cells along the pleural surface (Figure 3). Post-operatively, there was radiological improvement and diaphragmatic configuration (Figure 1b). Significant improvements in spirometry values, lung volume measurement and arterial oxygen tension were also documented (Table 1). Three months postoperatively, the patient was in good health and back at work.

---

Fig. 1. Chest radiography taken (1a) and (1b) before and after lung volume reduction therapy.
Table 1. Pulmonary function and arterial gas data before and one month after LVRS

<table>
<thead>
<tr>
<th>Lung function</th>
<th>Predicted</th>
<th>Pr-eoperative (%)</th>
<th>One month post-operative (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>FEV1 (L)</td>
<td>3.67</td>
<td>0.82</td>
<td>22.2</td>
</tr>
<tr>
<td>FVC (L)</td>
<td>4.44</td>
<td>1.49</td>
<td>33.6</td>
</tr>
<tr>
<td>MMEF 75/25 (L/S)</td>
<td>4.25</td>
<td>0.25</td>
<td>5.9</td>
</tr>
<tr>
<td>VC (L)</td>
<td>4.63</td>
<td>1.71</td>
<td>36.9</td>
</tr>
<tr>
<td>TLC (L)</td>
<td>6.58</td>
<td>7.4</td>
<td>112.5</td>
</tr>
<tr>
<td>RV (L)</td>
<td>1.91</td>
<td>5.69</td>
<td>297.8</td>
</tr>
<tr>
<td>PaO2 mmHg#</td>
<td></td>
<td></td>
<td>70</td>
</tr>
<tr>
<td>PaCO2 mmHg#</td>
<td></td>
<td></td>
<td>59</td>
</tr>
</tbody>
</table>

#FiO2: 0.21

Fig. 2a,2b. Chest computed tomography showed bilateral giant bullae occupying more than half of the hemithorax and diffused small centrilobular lucency.

Fig. 3. Histologically, it showed multicystic lesion lined by flat to cuboid cells (arrow) along the pleural surface (Hematoxylin and eosin stain 40X, inset: 400X).
Discussion

Development of multiple lung bullae [variously termed primary bullous disease of the lung, GBE, or VLS] is rare, and has been described in young male smokers. VLS is a progressive disease characterized by dyspnea on exertion, hemoptysis, and infrequently, spontaneous pneumothorax. Tobacco smoking has been implicated in the etiology of large giant bullae. Low BMI in combination with smoking may play an important role in the development of pleural blebs in healthy young adults. Bullae occupying one third to one half of one or both hemithoraces may produce a dramatic chest radiograph, but such patients may have little abnormality of lung function. More often, patients with giant bullae have airflow obstruction ranging from minimal to severe. A comprehensive 1996 review of surgery for GBE concluded that three features portend success: bulla size exceeding one third of one hemithorax, marked compression of adjacent lung tissue (demonstrated by CT scan), and preoperative FEV1 less than 50% of predicted. Our patient met all three criteria. Surgical therapy has been used for bullous emphysema since the mid-20th century. The best surgical candidates are often those with giant pulmonary bullae rather than those with diffuse disease.

Hypoxemia and hypercapnia appear to be among the measurements most frequently improved by surgery. Increases in FEV1, FEV1/FVC, and diffusion of carbon monoxide (DLCO) were generally modest, and vital capacity (VC), total lung capacity (TLC), residual volume (RV) and trapped gas generally decreased. Bullectomy leads to significant improvement in dyspnea, gas exchange, pulmonary function, and exercise capacity, with the best results being obtained in the more significant VLS cases. On average, improvements persisted for approximately 3-4 years, but began to decline thereafter. Together, these physiological improvements resulted in an improvement in dyspnea with exercise and an improvement in quality of life. But the National Emphysema Treatment Trial (NETT) Research Group reported that overall lung volume reduction surgery increased the improvement in exercise capacity, but did not confer a survival advantage over medical therapy.

This case demonstrates that giant bullae can be successfully managed with surgical resection, and their size can be determined by different techniques, including lung volume measurements and chest CT. Obvious improvements in dyspnea and pulmonary function values were observed after surgery.

References


消失的肺症候群和肺部減體手術 - 病例報告

陳正雄¹，葉金水¹，蔡政宏¹，紀炳銓¹，
蕭富源²，劉淨蘭³，蔡偉宏¹

摘要

多發性巨型大泡性肺氣腫又稱消失的肺症候群，臨床上很罕見，它通常發生在年輕的抽菸患者。我們報告一個四十一歲男性經由胸腔電腦斷層診斷為 Vanishing 肺症候群，他每天抽四十支香菸持續二十二年，主訴活動性喘持續二年，最近一週症狀加劇，患者戒菸並接受肺部減體手術，手術後，他的臨床症狀，胸腔X光，肺功能，都明顯改善，手術後三個月，患者恢復健康並回歸工作崗位，這個病例報告證實巨型大泡性肺氣腫可以成功經由手術處理，且它的大小可經由胸腔電腦斷層和肺容積檢查作判斷。

關鍵詞：大泡性肺氣腫，消失的肺症候群，氣胸

聯絡人：蔡偉宏醫師
500 彰化市南校街 135 號，彰化基督教醫院彰化基督教醫院胸腔內科¹
電話：04-711-6917；傳真：04-722-8289；E-mail：77112@cch.org.tw
彰化基督教醫院急診醫學部²
彰化基督教醫院病理科³