

## An Early Case of Pulmonary Lymphangiomyomatosis Diagnosed by Video-assisted Thoracoscopic Surgery

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**Pulmonary lymphangiomyomatosis (LAM) is a rare and progressive disease of young women that usually causes death from respiratory failure. Here we report an early case that was diagnosed by thoracoscopy. A 28-year-old woman presented to a local clinic with chest pain and her chest X-ray film showed left pneumothorax. After placement of a chest tube, the lung re-expanded fully. Following the recurrence of left pneumothorax, she was referred to our hospital and underwent video-assisted thoracoscopic surgery (VATS). However, left pneumothorax recurred again one month later. She underwent a second thoracoscopic operation, during which a bulla was recognized in the lingular segment of the left lung and was resected. On histological examination of the surgical specimen findings consistent with LAM were obtained. Taking the preoperative imaging findings and the thoracoscopic findings into consideration, a diagnosis of early lymphangiomyomatosis was made. Thoracoscopic management of pneumothorax in young women can facilitate the early diagnosis of this condition. (Ann Thorac Cardiovasc Surg 2005; 11: 405–7)**

**Key words:** pulmonary lymphangiomyomatosis, video-assisted thoracoscopic surgery, pneumothorax, bulla, early diagnosis

### Introduction

Pulmonary lymphangiomyomatosis (LAM) is a rare disease that affects women of child-bearing age. The occurrence of progressive dyspnea and intractable pneumothorax lead to a poor prognosis. LAM is characterized by the excessive growth of smooth muscle in the walls of the bronchioles and alveoli, sites where there is little muscle tissue in the normal lung. The diagnosis of LAM used to be made by open lung biopsy, transbronchial lung biopsy, or autopsy. In the present patient, LAM was diagnosed after a thoracoscopic operation for recurrent pneumothorax.

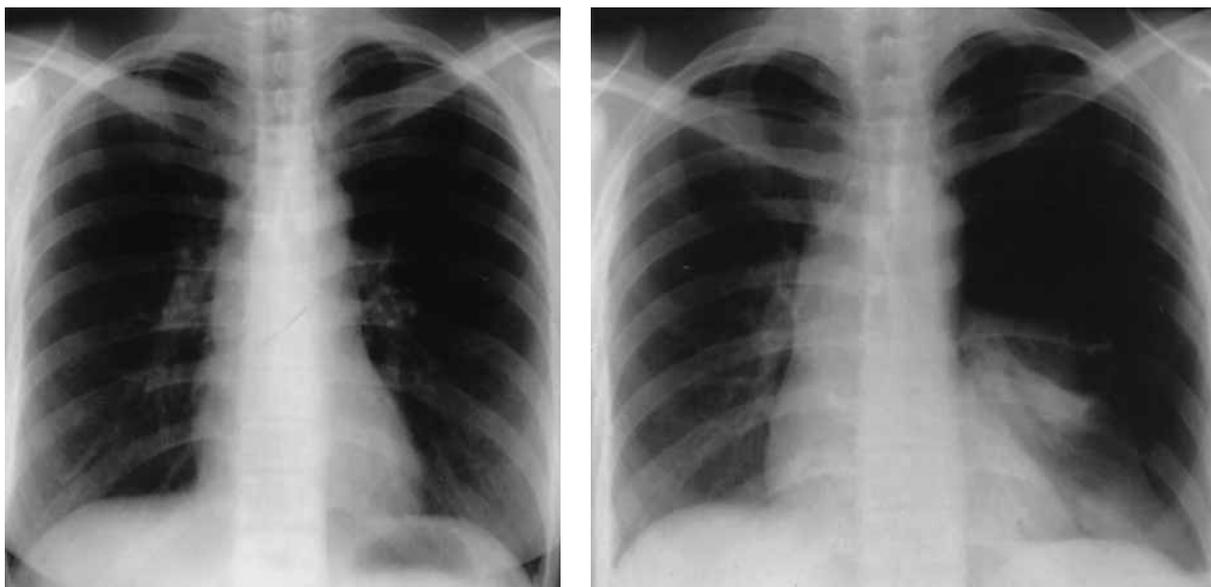
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### Case Report

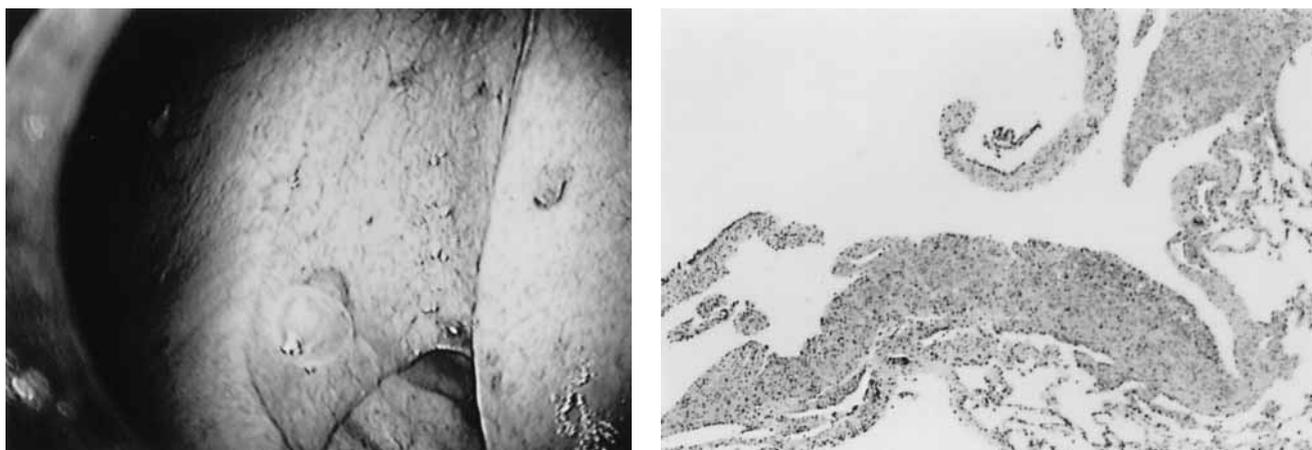
A 28-year-old woman first presented to a local clinic with left-sided chest pain and chest X-ray film showed left-sided pneumothorax. Placement of a chest tube allowed the lung to re-expand. She was subsequently referred to our hospital because of recurrence of the pneumothorax. Her past medical history was unremarkable. She was not menstruating. There was no family history of a neurocutaneous syndrome. On chest X-ray film showed a left-sided pneumothorax chest CT scans did not reveal any bullae on the left side (Fig. 1A). After placement of a chest tube, the lung re-expanded fully again. However, pneumothorax recurred after removal of the chest tube so she underwent video-assisted thoracoscopic surgery (VATS).

Thoracoscopy revealed capillary engorgement over the entire surface of both the parietal and visceral pleura, as well as slight emphysematous changes of the lung. The



**Fig. 1.**  
A: A chest radiograph reveals left pneumothorax.  
B: A chest radiograph showed recurrent left pneumothorax.

A | B



**Fig. 2.**  
A: A bulla can be seen in the lingula of the left lung.  
B: Photomicrograph reveals emphysema-like lesions and proliferation of smooth muscle in the lung. (H&E stain)

A | B

emphysematous apical segment of her left lung was resected using an automatic stapler, and the postoperative course was uneventful. Examination of the surgical specimen showed no clear histological evidence of a bulla or LAM.

One month postoperatively, she again developed chest pain on the left side and chest X-ray film showed a new left pneumothorax (Fig. 1B). VATS was performed again and a bulla was recognized in the lingular segment of the left lung (Fig. 2A). The affected lung tissue and the bulla

were resected with an automatic stapler. Her postoperative course was again uneventful, but histological examination of the resected specimen indicated a diagnosis of LAM (Fig. 2B).

### Discussion

LAM is a rare diffuse lung disease that occurs in women of child-bearing age, and is characterized by the abnormal overgrowth of smooth muscle in the small airways

and alveoli.<sup>1,2</sup> It presents with dyspnea and recurrent pneumothorax, eventually progressing to respiratory failure and intractable pneumothorax with a poor prognosis. The true incidence and prevalence of LAM are unknown. It is diagnosed at a rate of 1 case per 1,000,000 persons in Europe and the United States,<sup>3</sup> but the actual incidence is probably higher because LAM is often mistaken for asthma, chronic obstructive lung disease, or bronchitis. The first six cases of LAM were reported as lymphangiomyoma by Cornog and Enterline in 1966.<sup>4</sup> In 1974, Silverstein et al.<sup>5</sup> designated this condition as pulmonary lymphangiomyomatosis and many cases have been reported since then. LAM presents with characteristic bilateral diffuse reticular or military changes and overexpansion of the lungs on chest X-ray film, while multiple cysts with uniform thin walls are distributed diffusely through the lungs on chest CT scans.<sup>6</sup> In the present patient, no bulla that seemed to be the likely cause of pneumothorax was detected during the first thoracoscopic operation, but it has been reported that clusters of cysts with minute bullae can cover the surface of the lung in LAM. In a previous report patients with thoracoscopic diagnosis of LAM were first diagnosed clinically from the findings on chest CT scans and pathological confirmation was obtained by thoracoscopy. In the present patient, chest X-ray film and chest CT scans obtained after re-expansion of the left lung failed to demonstrate any of the characteristic features of LAM, including multiple cysts scattered through the lung field or irregular walls of bronchioles and vessels. During the second thoracoscopic operation, a solitary bulla was recognized in the lingular segment of the left lung and was resected, after which a diagnosis of LAM was made by histological examination of the surgical specimen. LAM is characterized by abnormal interstitial proliferation of smooth muscle cells derived from lymphatics around the pulmonary vessels, bronchioles, and alveolar septa. It has been reported that immunohistochemical assessment of progesterone receptor expression, as well as HMB45 expression, is of diagnostic value,<sup>7</sup> but these markers are not necessarily expressed by all LAM cells and both were negative in the present patient. Destruction of the alveolar walls, abnormal interstitial proliferation of smooth muscle cells, and positivity for smooth muscle antibody were demonstrated by pathological examination in the present case, these are unlikely to be physiological findings and are consistent with a diagnosis of early LAM. According to the Armed Forces Institute of Pathology,<sup>8</sup> early LAM presents with evidence of pulmonary emphysema, while dif-

fuse cystic changes occur as the disease progresses and honeycomb lung eventually develops. In the present patient, only slight emphysematous changes were observed thoracoscopically, but there was severe capillary engorgement over the entire lung surface. It has been reported that it is possible to diagnose early LAM by thin-section CT<sup>9</sup> and some authors<sup>6,10</sup> have suggested that histological confirmation is not necessarily required to establish a diagnosis of this disease at an early stage. In the present case, however, early LAM could not be diagnosed by chest CT. In conclusion, thoracoscopic surgery may be useful in young women with pneumothorax by facilitating the early diagnosis of LAM as well as treating the acute problem.

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