

# An Experience with the Modified Total Pleural Covering Technique in a Patient with Bilateral Intractable Pneumothorax Secondary to Lymphangioliomyomatosis

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**We present a case of bilateral intractable pneumothorax associated with a modified form of lymphangioliomyomatosis (LAM), successfully treated with a previously reported surgical procedure, the total pleural covering (TPC) technique, under video-assisted thoracic surgery. The patient was a 28-year-old woman with bilateral pneumothorax secondary to LAM who had undergone thoracoscopic surgery in another hospital. We performed bilateral TPC modified with a preceding coverage of air leak points with polyglycolic acid sheets for reinforcement. Although a minor air leak after the surgery necessitated a mild pleurodesis on the right side, the bilateral pneumothorax was well controlled, and no recurrence has been observed for 9 months. We believe that TPC is a safe and reliable procedure for the management of intractable pneumothorax secondary to LAM. It also has the potential to reduce risk of excessive bleeding in lung transplantation. (Ann Thorac Cardiovasc Surg 2010; 16: 439–441)**

**Key words:** thoracoscopy, pregnancy, lymphangioliomyomatosis, pneumothorax

## Introduction

Lymphangioliomyomatosis (LAM) is a rare disease that occurs mainly in women of reproductive age. It is characterized by the proliferation of abnormal smooth muscle-like cells (LAM cells) in the lung, axial lymph nodes, and elsewhere. Patients with pulmonary LAM suffer from progressive respiratory impairment on the basis of a destruction of lung parenchyma resulting from a proliferation of LAM cells, and lung transplantation represents the mainstay of therapy for selected patients with advanced pulmonary LAM. Widespread pulmonary proliferation of LAM cells leads to diffuse cystic destruction

of the lung parenchyma, and pneumothorax frequently occurs because of a disruption of the fragile cysts, representing one of the primary symptoms leading to diagnosis.<sup>1)</sup> The incidence of pneumothorax in LAM patients is higher than that found in other chronic pulmonary diseases. Kitaichi and co-workers reported that pneumothorax occurred in 18 of 46 LAM patients (39%).<sup>2)</sup> Chu and co-workers also reported that pneumothorax was observed in 24 of 35 LAM patients (69%), 19 of whom (79%) underwent either pleurodesis or pleurectomy.<sup>3)</sup>

Pleural interventions, such as pleurodesis and pleurectomy, have been primarily employed in the management of pneumothorax in patients with LAM, especially in those with recurrent pneumothorax. However, severe and broad pleural adhesion as a result of the pleural interventional procedures sometimes causes serious bleeding when the patients undergo lung transplantation. This is especially true when the transplantation requires extracorporeal membrane oxygenation. In recent years, minimally invasive video-assisted thoracic surgery (VATS) has become a standard form of surgery for pneu-

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**Fig. 1.** A thoracoscopic view showing fragile cysts in the right sides during video-assisted thoracic surgery. These cysts were observed throughout the lung surface.



**Fig. 2.** A thoracoscopic view showing lung surface covered with regenerative oxidized cellulose mesh and fibrin glue.

mothorax. The total pleural covering technique (TPC), which refers to a surgical procedure to cover the entire visceral pleura using regenerative oxidized cellulose mesh (ROC) followed by a spray of fibrin glue to coat the ROC, is a VATS procedure initially proposed by Kurihara and co-workers as a treatment for pneumothorax secondary to LAM.<sup>4)</sup> TPC has a potential advantage to reduce pleural adhesion compared with other pleural interventions because it has been reported that no pleural adhesion was observed in 2 VATS patients with recurrent pneumothorax after TPC, and the visceral pleural surface was thickened and covered with collagen fibers in a microscopic examination of resected specimens.<sup>4)</sup> We present here a patient with bilateral intractable pneumothorax resulting from LAM that was successfully treated with bilateral TPC. We modified the original TPC to cover air leak points with polyglycolic acid (PGA) sheets at first, and TPC was then performed. The surgical technique and the perioperative course are described.

## Case Report

A 28-year-old woman in the 9th week of gestation presented to a local hospital complaining of shortness of breath and right-side chest pain. A chest X-ray showed pneumothorax on that side. A chest computed-tomographic scan revealed small bullae in the apexes of both lung fields. Although the patient was initially treated with chest tube drainage, air leakage was prolonged for 4 days, and the patient was moved to another hospital for surgical treatment. VATS was performed on day 7 after admission. A thoracoscopic view showed multiple small thin-walled

cysts throughout the right lung surface and a ruptured cyst in the apex of the lung, which was resected with an Endo-GIA60-3.5 mm stapler (Auto Suture, Tyco, USA). Pathological examination of the surgical specimen established a diagnosis of LAM. The chest drainage tube was removed on day 5 after surgery; nevertheless, she underwent chest tube drainage again because of recurrent pneumothorax on day 6 after surgery. Left side pneumothorax was further developed on day 12 after surgery, and a chest drainage tube was inserted into the left chest cavity. She eventually was referred to our hospital for the treatment of bilateral intractable pneumothorax. The patient decided to resign childbirth because of anxiety over the rapid progression of LAM during pregnancy. As a result of severe air leakage on the left side and a possibility of offering lung transplantation in the future, we decided to perform TPC on both sides. VATS in the right side revealed diffuse multiple cysts on the lung surface, and a minor alveolar fistula near the staple line formed in the first operation. We coated an area of pleural surface around the air-leak point with PGA sheets (Neoveil, Gunze Ltd., Kyoto, Japan), and we then performed TPC using ROC (Igaki Medical Planning, Tokyo, Japan). First, while keeping the lung deflated, we covered the entire surface of the visceral pleura with ROC, including the interlobular and diaphragmatic surfaces. The lung was then half inflated, and additional coverage was applied to the area becoming uncovered by inflation. We used 10 mL of fibrinogen solution and 10 mL of thrombin solution, both diluted with 30 mL of saline (Bolheal, the Chemo-Sero-Therapeutic Research Institute, Kumamoto, Japan) to spray the entire ROC surface (Fig. 2). Next we

changed the operating posture from the left to the right decubitus position and applied TPC on the left side in a similar manner. Chest X-ray after the operation revealed good reexpansion of both lungs. A minor air leakage was observed on the right side, and a mild pleurodesis with 5 KE of OK-432 and 100 mg of minocycline was performed on day 4 after surgery, and the chest tube on the right side was removed on day 6. No air leak was observed from the left-side chest tube, but slight subcutaneous emphysema was observed in the left chest cervical area. The subcutaneous emphysema, however, gradually subsided, and the left chest tube was removed on day 13 after surgery. The patient was discharged on day 19 after surgery, and no recurrent pneumothorax has been recorded during 9 months after TPC.

## Discussion

We successfully applied TPC in a patient with bilateral intractable pneumothorax secondary to LAM. We modified the original TPC at first by covering a staple line, including an air leak point with PGA sheets, reported to be an ideal material for staple-line reinforcement,<sup>5)</sup> and we believe it is more reliable in controlling air leakage than ROC mesh is. We next applied TPC with ROC mesh, which is less expensive than PGA sheets, and reported that it caused no pleural adhesion.

Surgeries during pregnancy should preferably be delayed until the second trimester to avoid early teratogenic effects. The patient, however, decided to resign childbirth because pregnancy exacerbates progression of LAM and increases the incidence of LAM-related complications. Johnson and co-workers found that the overall incidence of LAM-related complications during pregnancy was 11 times higher than at any other time.<sup>6)</sup>

Conservative therapy for pneumothorax in LAM is associated with high recurrence rates, and pleural interventions such as pleurodesis or pleurectomy are frequently required. One of the most effective therapies for intractable pneumothorax is talc poudrage, which is associated with a reduction in the rate of pneumothorax recurrence when compared with bullectomy under VATS with or without mechanical pleurodesis.<sup>7)</sup> On the other hand, it must be noted that significant pleural symphysis as a result of the talc poudrage increases a risk of excessive bleeding that may lead to intraoperative death.<sup>8)</sup> Recently,

Kurihara and co-workers reported TPC using ROC mesh that causes only a thickening of visceral pleura and does not raise intrathoracic adhesion. ROC coated with fibrin glue appears not to invade fibroblasts in the thoracic wall, and this is what prevents adhesion.<sup>4)</sup> We used a PGA sheet to apply the TPC modified with coverage of air leak points to a patient with intractable bilateral pneumothorax secondary to LAM. Although a minor air leak after the TPC necessitated a mild pleurodesis on the right side, the bilateral pneumothorax was well controlled, and no recurrence has been observed for 9 months. Postoperative lung function is better-preserved by this technique after 9 months, though it is unclear for the reservation of long-term pulmonary functions. We believe that TPC is a safe and reliable procedure for the management of intractable pneumothorax secondary to LAM, which also has future potential to reduce the risk of excessive bleeding in lung transplantation.

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