Pleural Covering Application for Recurrent Pneumothorax in a Patient with Birt-Hogg-Dubé Syndrome

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Birt-Hogg-Dubé syndrome (BHDS) is a rare hereditary disease that presents with multiple lung cysts and recurrent pneumothorax. These cysts occupy predominantly the lower-medial zone of the lung field adjacent to the interlobar fissure, and some of them abut peripheral pulmonary vessels. For the surgical management of pneumothorax with BHDS, the conventional approach of resecting all subpleural cysts and bullae is not feasible. Thus, after handling several bullae by using a stapler or performing ligation as a standardized treatment, we applied to a pleural covering technique to thicken the affected visceral pleura and then to prevent recurrence of pneumothorax. We herein report the successful application of a pleural covering technique via thoracoscopic surgery to treat the recurrent pneumothorax of a 30-year-old man with BHDS. This technique is promising for the management of intractable pneumothorax secondary to BHDS.

Keywords: Birt-Hogg-Dubé syndrome, multiple lung cysts, recurrent pneumothorax, thoracoscopic surgery

Introduction

Birt-Hogg-Dubé syndrome (BHDS), a hereditary genodermatosis caused by a germline folliculin (FLCN) mutation, was first described in 1975 and 1977.1,2) The thoracic manifestations of this syndrome are multiple lung cysts and pneumothorax with frequent recurrences.3) These cysts appear predominantly at the lower-medial zone of the lung field and adjacent to the interlobar fissure but sometimes abut peripheral pulmonary vessels.4,5) Thus, the conventional surgical approach in which all subpleural cysts and bullae are resected is not feasible for the management of pneumothorax in BHDS. Here, we describe a patient with BHDS whose recurrent pneumothorax was successfully resolved by applying a pleural covering technique during thoracoscopic surgery.6) This procedure was effective in preventing further recurrence.

Case Report

A 30-year-old male presented at the emergency department of our hospital, complaining of a sudden left-sided chest pain. A chest X-ray and computed tomography showed collapse of the left lung and multiple lung cysts. He was diagnosed with left-sided pneumothorax and treated with the insertion of a portable drainage kit
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(Thoracic Egg®, Sumitomo Bakelite CO., Ltd. Tokyo, Japan), that enables to manage him in a setting of outpatient clinic. One-week after insertion of the tube, resolution of the pneumothorax enabled removal of the chest drainage tube. The patient was an ex-smoker (10 packs/year) but had no medical problems in the past. However, a noteworthy comment was that his father and brother had medical histories of pneumothorax. Since this patient had a family history of pneumothorax and had multiple lung cysts localized predominantly at the lower-medial zone of the lung fields (Fig. 1), a diagnosis of BHDS was strongly suspected. Genetic testing was performed using peripheral blood leucocytes, and then demonstrated that he had an insertion of cytosine (C) in exon 11 of the FLCN gene (c.1285dupC), thereby confirming the diagnosis. Subsequently, after 16 and then 20 months passed, his left-sided pneumothorax recurred. Accordingly, he underwent video-assisted thoracoscopic surgery (VATS) as the traditional option. The operative findings revealed multiple cysts of varied sizes over the whole left lung. These cysts predominated on the lung’s basal and mediastinal sides including the hilum and interlobar areas (Fig. 2a). Additionally, some cysts were adjacent to pulmonary arteries and veins. We concluded that the standardized approach for treating recurrent pneumothorax,7,8 that is, resecting all bullae or performing a partial pleurectomy of the lung’s mediastinal portion (i.e., close to the pulmonary artery, vein and hilum) was not feasible. Thus, we first performed a partial bullectomy which doubled as a lung biopsy of the lingual segment using a stapler (Ethicon ENDOPATH® STAPLER ECHELON FLEX™ Johnson, New Brunswick, NJ, USA), and ligated residual bullae as much as

Fig. 1 Preoperative computed tomographic images of the chest showed multiple lung cysts localized predominantly at the lower-medial zone of the lung field and adjacent to the interlobar fissure. The cysts, which were thin-walled and round- to oval-shaped, sometimes abutted the peripheral pulmonary vessels (arrow).

Fig. 2 Operative findings of the left lung under VATS. (a) A large, thin-walled, transparent and non-resectable bulla was present, the surface of which had vessels in interlobar septa. Small bullae were also found in interlobar and mediastinal surfaces of the left lung. (b) The entire surface of the left lung was first covered with sheets of ROC mesh, and fibrin glue was then applied drop by drop over that surface. A ★ indicates the left upper lobe. VATS: video-assisted thoracoscopic surgery; ROC: regenerative oxidized cellulose
possible using ENDOLOOP® (Johnson, New Brunswick, NJ, USA). After those procedures, we decided to administer a pleural covering technique using regenerative oxidized cellulose (ROC) mesh (Ethicon SURGICEL® absorbable hemostat gauze, 4” x 8”, Johnson and Johnson, New Brunswick, NJ, USA). After first enclosing the entire surface of the left lung with nine sheets of ROC mesh, our next step was to drop 10 ml of fibrin glue solution (Bolvig®, The Chemo-Sero-Therapeutic Research Institute, Kumamoto, Japan) over the left lung’s entire surface (Fig. 2b). The operative time period was 143 min, and the amount of bleeding was small. Since no air leakage was observed after surgery, no additional post-operative procedures including pleurodesis were required. The chest tube was removed on post-operative day (POD) 4. The patient was discharged on POD6, and no recurrence has been recorded during the one-year post-surgical period.

Discussion

BHDS is a rare inherited autosomal dominant genodermatosis characterized by fibrofolliculomas and trichodiscomas of the skin, renal tumors, and multiple lung cysts. The candidate gene for BHDS was found on chromosome 17p11.2, named the FLNC gene, which is composed of 14 exons and encodes protein called folliculin. The pulmonary multiple cysts predominantly located at lower lung fields, mediastinal sides, and interlobar area of the BHDS-affected lungs. Additionally, these bullae exhibit thin-walled, round- to oval-shaped, various sizes, and some of them abut to peripheral pulmonary vessels. These peculiar properties of BHDS cysts make standard surgical procedures for resecting bullae with a surgical stapler or ligating bullae very complicated. Thus, pneumothorax with BHDS is prone to recur intractably.

In guidelines from the American College of Chest Physicians (ACCP) and British Thoracic Society (BTS), the suggested protocols for recurrent secondary pneumothorax are open thoracotomy or VATS with pleurectomy as a surgical procedure and chemical pleurodesis as a medical treatment. However, these strategies appear to be inapplicable for patients with BHDS. Pleurectomy is not feasible because of the lungs’ close proximity to the pericardial, perivascular and interlobar areas. Moreover, chemical pleurodesis is not effective at such areas, since parietal and visceral pleural surfaces slide continuously with heartbeats and respiratory movement of the diaphragm, preventing the chemical agent for pleurodesis from staying in place. Thus far, the relevant guidelines lack any description of the practical and important questions: (1) which surgical methods, i.e., bullectomy using stapler, ligation, etc. are recommended to provide the least risk of postoperatively regenerating bullae and subsequent recurrence or (2) which agents for pleurodesis are the best in terms of efficacy and safety.

Kurihara et al. reported a new surgical procedure named the pleural covering technique for the additional treatment of pneumothorax and to reduce its recurrence. The pleural covering technique utilizes ROC mesh and fibrin glue to reinforce fragile visceral pleura without adhesions. Recently, this method of enclosing the entire pleural surface was found effective for preventing pneumothorax in patients with lymphangioleiomyomatosis, another cystic lung disease that causes recurrent pneumothorax. In our male patient with BHDS described here, the peculiar nature and locations of multiple bullae prompted us to apply this technique for treating his pneumothorax and for preventing its recurrence on the same side of his lung.

We conclude that our success with this pleural covering technique indicates its effectiveness and reliability for the management of intractable pneumothorax secondary to BHDS. Further evaluation of the pleural covering technique for managing pneumothorax in patients with BHDS is well warranted.

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References


