Swyer-James syndrome associated with asthma and a giant bulla

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Asthma, giant bulla, Swyer-James syndrome.

Introduction
Swyer-James syndrome is a rare pediatric lung disease with unilateral pulmonary hyperlucency on a roentgenogram, but there are no reports about an association with giant bullae. Here, we report a surgical case of Swyer-James syndrome complicated by a giant bulla and asthma.

Case Report
In June 2006, a 13-year-old girl was referred to Kyoto University Hospital with dyspnea on exertion and wheezing. There was a history of pneumonia in infancy and asthma during early childhood, which had remitted after she started primary school. She did not have atopy or allergies. Both the chest X-ray and the chest computed tomography showed a giant bulla in the right upper lobe and hyperlucency of the right lower lobe because of diminished vasculature due to a hypoplastic right pulmonary artery. There was no evidence of bronchiectasis. The X-ray taken before referral to our hospital was not available. She was admitted to Kyoto University Hospital in March 2007 for further evaluation.

The exhaled nitric oxide level (27.9 ppb) was elevated compared with that of normal children (<20 ppb). The sputum eosinophil count was increased to 2.5%. Respiratory function tests (performed in July 2006) showed a severe obstructive pattern (Table 1).

A methacholine challenge test was not performed because of severe airflow limitation, but peak expiratory flow (PEF) monitoring showed diurnal variation of 19% from 210 l/min in the morning to 250 l/min in the evening. A pulmonary perfusion scan demonstrated that arterial flow was decreased on the right side (right : left ratio = 1:3).

Based on the detection of unilateral pulmonary hyperlucency and the absence of other possible causes, including congenital cysts, congenital pulmonary artery abnormalities, and embolization of the main pulmonary artery, she was diagnosed as having Swyer-James syndrome with a giant bulla. In addition, her response to an inhaled bronchodilator, the diurnal variation of PEF, and the detection of eosinophilic airway inflammation suggested the recurrence of her asthma. Although surgical resection of the giant bulla was considered, medical treatment of the patient’s asthma was given precedence to maximize her respiratory function. After treatment with an inhaled...
corticosteroid preparation and tulobuterol patch, her exertional dyspnea showed marked improvement, and the exhaled nitric oxide level and PEF also improved. From August 2009, however, she complained of right-sided chest pain and her exertional dyspnea recurred. Simultaneously, the giant bulla in the right upper lobe enlarged further and the patient’s inspiratory capacity decreased (Figure 1).

Because her chest pain and exertional dyspnea continued to worsen despite higher doses of asthma medication, surgical resection of the giant bulla was performed in January 2011. Under general anesthesia, the patient was placed in the left lateral position. A massive bulla with a small pedicle in the right S1 was completely resected using an Endo-GIA 45-mm stapling gun (Covidien, Tokyo, Japan) and three cartridges via three access ports: (1) an 11.5 mm port in the seventh intercostal space at the posterior axillary line, (2) a 5.5 mm port in the sixth intercostal space at the inferior angle of the scapula, and (3) an 11.5 mm port in the fifth intercostal space at the anterior axillary line. Histopathological examination of the resected giant bulla and the surrounding lung parenchyma showed mild emphysematous change and proliferation of smooth muscle in the peripheral airways. There were no constrictive changes of the bronchioles or dysplasia of the pulmonary vessels and inflammatory changes were minimal.

After surgical resection, her exertional dyspnea improved dramatically and the right-sided chest pain resolved. In August 2011, she was being treated with a leukotriene receptor antagonist alone.

**Table 1. Changes of pulmonary function tests and the BODE index.**

| Date          | Before surgery | After surgery | | | | | | |
|---------------|----------------|---------------|---------------|---------------|---------------|---------------|---------------|---------------|---------------|
|               | July 2006      | March 2008    | October 2010  | April 2011    | August 2011 (before inhalation) | August 2011 (after inhalation) |
| FVC (L) (% predicted) | 2.31 (83.2) | 2.51 (92.3) | 2.43 (74.5) | 2.66 (80.1) | 2.78 L | 2.95 L |
| FEV1 (L) (% predicted) | 1.42 (47.5) | 1.52 (49.7) | 1.65 (52.9) | 1.69 (53.3) | 1.76 | 1.97 |
| FEV1/FVC (%) | 61.9 | 60.6 | 67.9 | 63.5 | 63.3 | 66.8 |
| VC (L) (% predicted) | 2.23 (79.4) | 2.47 (90.8) | 2.47 (75.8) | 2.64 (79.5) | – | – |
| IC (L) | 1.67 | 1.64 | 1.51 | 1.64 | – | – |
| FRC (L) (% predicted) | 1.97 (104.8) | 2.15 (112.0) | 1.95 (100) | 2.27 (114.6) | – | – |
| RV/TLC (%) | 31 | 34.9 | 33.2 | 30.1 | – | – |
| DLCO (mL/min/mmHg) (% predicted) | 22.7 (119.7) | 22.3 (114.6) | 22.4 (113.9) | 21.3 (106.1) | – | – |
| BODE index | 3 | 3 | 3 | 2 | – | – |
| FEV1 (% predicted) | 2 | 2 | 1 | 1 | – | – |
| 6 MWD | 0 | 0 | 0 | 0 | – | – |
| MMRC | 0 | 0 | 1 | 0 | – | – |
| BMI | 1 | 1 | 1 | 1 | – | – |

BMI, body mass index; BODE index, index derived from the BMI, severity of airflow obstruction and dyspnea, and exercise capacity; DLCO, diffusing capacity for carbon monoxide; FEV1, forced expiratory volume in 1 sec; FRC, functional residual capacity; FVC, forced vital capacity; IC, inspiratory capacity; MMRC, Modified Medical Research Council dyspnea scale; RV, residual volume; TLC, total lung capacity; VC, vital capacity; 6MWD, distance walked in 6 min.

**Discussion**

This was a rare case of Swyer-James syndrome complicated by a giant bulla and asthma. Medical treatment for asthma was initially effective, while subsequent resection of the giant bulla abolished the patient’s chest pain and also improved both exertional dyspnea and respiratory function.

Swyer-James syndrome was first reported in 1953 [1], and in 1954, Macleod reported nine adults with similar findings [2]. This syndrome features unilateral hyperlucency because of a decrease in pulmonary vascularity and air trapping that usually follows viral or bacterial respiratory infection during infancy or early childhood. Because of airflow limitation, patients are sometimes misdiagnosed as having asthma, so Swyer-James syndrome should be remembered as an important differential diagnosis in pediatric patients with airflow limitation. Our patient was diagnosed as having both Swyer-James syndrome and asthma based on an episode of pneumonia in infancy and typical radiological findings, as well as the response to asthma medication and the presence of eosinophilic airway inflammation. Hyperplasia of airway smooth muscle hyperplasia was also detected in the resected lung tissue.

Occurrence of spontaneous pneumothorax sometimes leads to the diagnosis of Swyer-James syndrome, suggesting that small bullae may develop in patients with this syndrome [3, 4]. However, to the best of our knowledge, the present patient is the first case of Swyer-James syndrome...
complicated by a giant bulla. The mechanism underlying the development of our patient’s giant bulla could not be clarified, but air trapping might have been involved. Patients with Swyer-James syndrome are usually managed conservatively and surgical intervention is only considered for problems such as recurrent infection or uncontrolled pneumothorax. In a professional athlete with this syndrome, it was reported that lung volume reduction surgery achieved the improvement of respiratory function [5]. The outcome in the present case suggests that surgery may also be appropriate for patients with Swyer-James syndrome and a giant bulla.

Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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