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Case Report

A surgical case of Swyer-James Macleod Syndrome (SJMS) discovered in adults due to respiratory distress, which was followed up as childhood asthma

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Abstract...

Background: SJMS is a relatively rare disease characterized by unilateral lung or lobe permeability on chest X-rays and narrowing of pulmonary blood vessels at the site. The treatment method has not been decided, and there are few reports of surgical cases.

Case presentation: A 44-year-old man had been followed up for asthma since childhood, but was admitted to our hospital with a complaint of respiratory distress. Chest X-ray showed hyperinflation of the right middle and lower lobes and left lung. Chest Computed Tomography (CT) showed mottled hyperpermeability in the right middle and lower lobes, and the left lower lobe had finer intrapulmonary blood vessels more permeability than other lobes. Pulmonary blood flow scintigraphy revealed a defect in the left lower lobe, and the diagnosis was SJMS. The left lower lobe was resected for the purpose of improving respiratory distress. The upper left lobe swelled, but the right middle and lower lobes swelled slightly, so it did not improve dramatically. However, preoperative respiratory distress has clearly improved, and lung volume reduction surgery for SJMS is considered to be meaningful.

Conclusions: Lung volume reduction surgery for SJMS may help improve respiratory distress.

Keywords: Swyer-James Macleod Syndrome; Obstructive Bronchiolitis; Pulmonary Blood Flow Scintigraphy; Thoracoscopic Lobectomy; Increased Lung Permiability.

Abbreviations: SJMS: Swyer-James Macleod syndrome; CT: Computed Tomography; PSL: Prednisolone; LAMA: Long Acting Muscarinic Antagonist; mMRC: modified Medical Research Counsil dyspnea scale; VC: Vital Capacity; FVC: Forced Vital C apacity; FEV1.0: Forced Expiratory Volume in one second; 99mTc-MAA: 99mTc-labeled macroaggregated albumin; HLA: HyperLucent Area;M: Male; F: Female.

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Background

SJMS is a rare disease caused by acquired lung hypoplasia and is not the target of surgical treatment in most cases. It has been reported that surgery is indicated when lung hyperinflation is found on one side, but surgical treatment is performed in cases where hyperinflation is found in both lung fields and blood flow defects are found on both sides even in lung scintillation. Since there are few cases of surgery, the judgment must be made with caution.

Case presentation

A 44-year-old man had been followed up for asthma since childhood, but was admitted to our hospital with a complaint of respiratory distress. On chest CT, a hyperlucent area (HLA) was observed in the bilateral lower lung fields, and bronchiolitis obliterans was considered bronchitis. Prednisolone (PSL) and long-acting muscarinic antagonist (LAMA) was administered but did not improve. The radiologist pointed out SJMS in the shadow when evaluated by CT again, so the possibility of improving respiratory distress by surgery was discussed. It was difficult for the patient to maintain a standing position, and the modified medical research counsil dyspnea scale (mMRC) was considered to be grade 4. Leukocytes were as high as 12000/μL in blood sampling, and Vital Capacity (VC) 0.89 L, %VC 22.1%, forced vital capacity (FVC) 0.94 L, forced expiratory volume in one second (FEV1.0) 0.36 L, % FEV1.09.9%, and FEV1.0% and 38.3% in respiratory function tests revealed hypopnea function. Chest X-ray showed hyperinflation in the right lower lobe and left lung (Figure 1a), chest CT showed mottled HLA in the right middle and lower lobes, and the left lower lobe had finer intrapulmonary blood vessels and more permiability than other lobes (Figure 1b). In pulmonary blood flow scintigraphy using 99mTc-labeled macro aggregated albumin (99mTc-MAA), there was a mottled defect in the right middle and lower lobes, and only a small amount of blood flow was observed in the left lower lobe (Figure 2). Regarding treatment, there were lung lesions on both sides, and it was unpredictable how much improvement in respiratory function would be obtained after resection of the left lower lobe, which was not functioning. The left lower lobe was emphysema-like, and both the interlobar pulmonary artery and the lower pulmonary vein were fine enough to suspect hypoplasia, but there were no abnormal blood vessels. When treating the blood vessels and bronchi, there were some adhesions, which were difficult, but these were dissected, and the lower left lobe was removed. Histological findings showed thinning of the alveolar wall and poor adhesion between the alveolar septum and the pleura (Figure 3a). Mild muscular thickening of the pulmonary artery intrapulmonary branch was observed, and an inflammatory reaction was observed mildly around the bronchioles. Elastica Wangison staining (Figure 3b) showed that the elastic fibers of the thinned alveolar septum were significantly fragmented. The report by Yoshida et al. [1] also cited inflammation around the bronchioles and emphysematous changes as the characteristics of SJMS, and this case was also diagnosed as SJMS. The postoperative course was uneventful, and within a few days after the operation, mMRC improved to grade 3. Respiratory function, especially FVC, FEV1.0 and % FEV1.0showed improvement 6 months after the operation, but remained unchanged thereafter. In addition, FEV1.0% remained almost unchanged before and after surgery, and mMRC remained grade 3 (Figure 4).

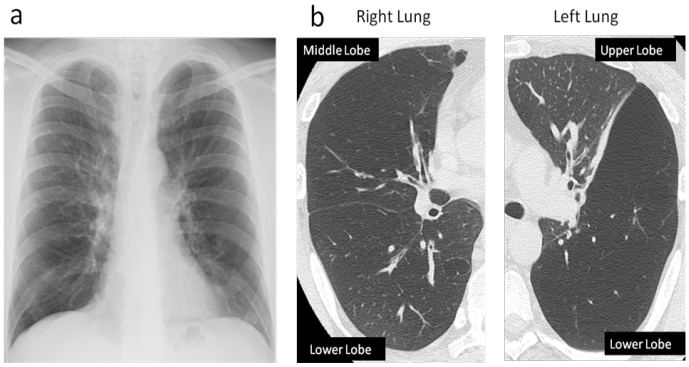

Figure 1: (1a) Chest X-ray showed hyperinflation in the right lower lobe and left lung.
(1b): Chest CT showed mottled HLA in the right middle and lower lobes, and finer intrapulmonary blood vessels and more permiability in the left lower lobe than other lobes.

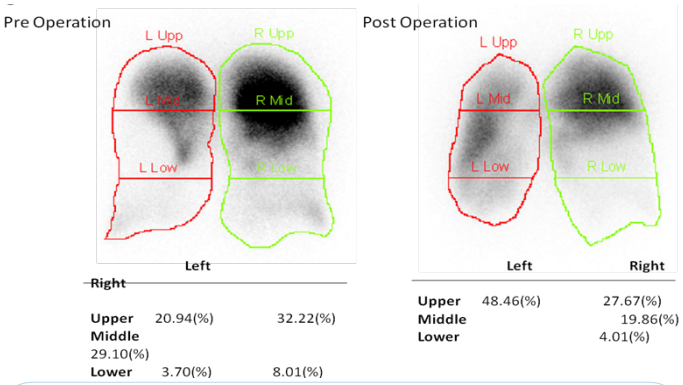

Figure 2: Pulmonary blood flow scintigraphy showed a mottled defect in the right middle and lower lobes, and only a small amount of blood flow in the left lower lobe.

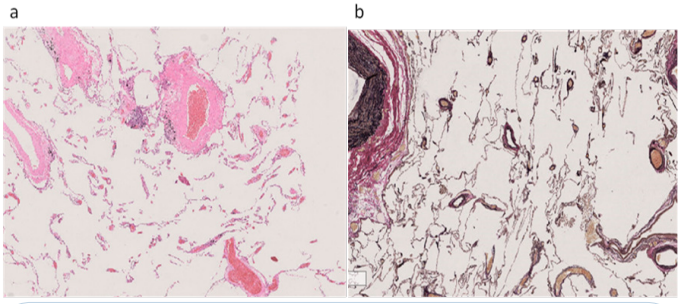

Figure 3: In hematoxylin eosin stainings, histological findings showed thinning of the alveolar wall and poor adhesion between the alveolar septum and the pleura, and mild muscular thickening of the pulmonary artery intrapulmonary branch was observed.
(3b): In elasticawangison staining, an inflammatory reaction was observed mildly around the bronchioles.

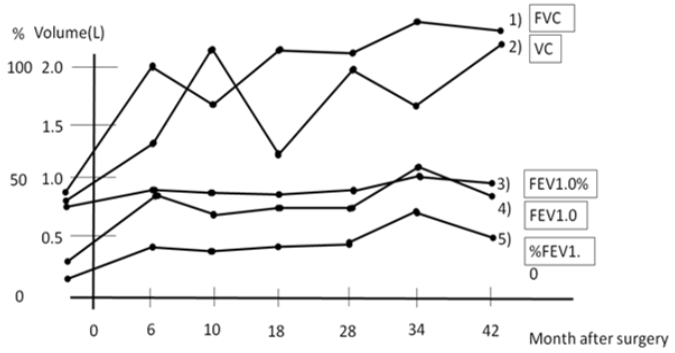

Figure 4: In respiratory function, especially FVC, FEV1.0 and % FEV1.0 showed improvement 6 months after the operation, but FEV1.0% remained almost unchanged before and after surgery.

Discussion

SJMS refers to a group of diseases with hyperpermeability of the chest and decreased secondary pulmonary vascular shadows without a clear cause of central bronchial obstruction. Macleod [3] reported pediatric and adult cases, who became known as SJMS. Regarding the cause of this syndrome, (1) bronchial theory and (2) pulmonary artery theory have been proposed [1,4]. For bronchial theory (1), deformation and stenosis cause obstructive bronchiitis, resulting in emphysematous changes in the peripheral airspace and associated impaired pulmonary blood flow. For pulmonary artery theory (2), secondary bronchial dysplasia and emphysematous changes occurred due to congenital pulmonary artery hypoplasia. In this case, chronic inflammation was observed around the bronchioles, and mild muscular thickening of the pulmonary artery intrapulmonary branch was confirmed, but no congenital hypoplasia was observed in the pulmonary artery, and there was repeated respiratory infection in early childhood. From the above, we considered bronchial theory (1) as the cause of SIMS in our case. There is no standard treatment for SJMS and no clear criteria for surgery. To maintain the current situation, it is necessary to vaccinate and to detect pneumonia at an early stage [5]. Surgery is recommended if recurrent pneumonia, pneumothorax, and giant lung cysts [6], exacerbate symptoms [7]. There were 7 adult cases, including our case, who visited a hospital for respiratory distress and were diagnosed with SJMS and underwent lung volume reduction surgery for the purpose of improving respiratory distress (Table 1) [7-12]. There were 4 males and 3 females, aged 25 to 65 years (median 43 years), and CT showed HLA in one lung in 6 cases. Only in our case, was HLA recognized in the bilateral lungs. Postoperative respiratory function was improved in all 7 cases, and although temporary improvement was observed after surgery in our case, there was little improvement thereafter. Regarding respiratory function, FEV1.0% showed almost no improvement before and after surgery. In pulmonary blood flow scintillation 10 months after surgery, blood flow in the upper left lobe doubled, but in the lower right middle lobe, blood flow was reduced by half (Figure 2). The causes of the stagnation of postoperative respiratory function improvement are as follows: (1) Along with the mediastinal shift to the left, the right middle and lower lobes are more aerated than before the operation, which enhances air trapping and causes residual right middle and lower lobe hyperinflation. (2) The right bronchiole was secondarily excluded and occluded, and deep pulmonary arteriovenous stenosis was caused, so pulmonary blood flow to the right middle and lower lobe decreased. (3) It is possible that the mismatch of ventilated blood flow increased during exertion, resulting in a decrease in PaO_2 and dilation of A-aDO2 [13]. In our case, SJMS in the right middle and lower lobes became apparent as a result of surgery, which may have partially offset the improvement in respiratory function in the left lung. At present, the respiratory distress of mMRC grade 4 does not appear just by standing up as before the operation, but we considered mMRC grade 3 due to stopping after walking 100m. However, at least clinical symptoms have improved, and lung volume reduction surgery aimed at improving respiratory function is considered to be effective for SJMS.

Conclusion

We experienced one surgical case of Swyer James syndrome. Although there may be no dramatic improvement after surgery in patients with bilateral defects in pulmonary blood flow scintigraphy, lung volume reduction surgery for SJMS may be significant in improving respiratory function.

Declarations

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Authors' contributions: AN wrote this manuscript, and AN, YK and YH performed the operation. HF, YS,YK, YY and YS helped with the perioperative management of the patient. All authors read and approved the final manuscript.

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Competing interests: The authors have no competing interests.

References

1. Yoshida K, Koba H, Igarashi T, Nakagawa A, Nishiyama K, et al. A case of Swyer-James Syndrome-A Morphological Study with Inflated Fixed Lung. The Japanese Journal of Thoracic Diseases. 1994; 32: 856-860.

2. Swyer PR, GC James. A case of unilateral pulmonary emphysema. Thorax. 1953; 8: 133-136. Macleod WM. Abnormal transradiancy of one lung. Thorax. 1954; 9: 147-153.

3. Takaharu Yamada, Yuichi Shibui, Yuji Watanabe. A Case Report of Lung Resection in a Boy With Bilateral Swyer-James Syndrome Due to Multiple Cysts in the Left Upper Lobe and Complete Atelectasis in the Left Lower Lobe. Journal of the Japanese Society of Pediatric Surgeons. 2018; 54: 927-934.

4. Tutar O, Esin D, Samanci C, Bas A. Adult diagnosis of Swyer-James-Macleod syndrome. BMJ Case Reports. 2012.

5. Kayawake H, Sakai H, Matsumoto H, Date H. Swyer-James syndrome associated with asthma and a giant bulla. Respirol Case Rep. 2014; 2: 150-153.

6. Mariana Vigiola Cruz, Gregory D.vTrachiotis. Pneumonectomy in a patient with Swyer-James-Macleod Syndrome. Surgical Science. 2015; 6: 55-58.

7. Sunil K, Ohri S, Guy Rutty, S William Fountain. Acquired segmental emphysema: the enlarging spectrum of Swyer-James/Macleod's syndrome. Ann Thorac Surg. 1993; 56: 120-124.

8. Tasaki A, Nakanishi R. Lung volume reduction surgery for a 7 professional athlete with Swyer-James syndrome. Ann Thorac Surg . 2005; 80: 342-344.

9. Yagi Y, Minami M, Yamamoto Y, Kanzaki R, Funaki S, et al. Pneumonectomy in an Older Patient With Swyer-James-MacLeod Syndrome With a Giant Bulla. Ann Thorac Surg. 2020; 109: e263-e265.

10. Miyakoshi J, Makino T, Shiono S, Anami Y, Tojima H. Swyer-James Macleod Syndrome with Progressive Hyperinflation. Ann Thorac Surg .2021; 4975: 02040-02043.

11. Tauchi S, Uchida T, Sugiyama H, Tobe S. Swyer-James syndrome requiring surgical therapy because of respiratory failure: A case report. The Journal of the Japanese Association for Chest Surgery. 2018; 32: 842-846.

12. Yoshirou Mochizuki. Swyer-James Syndrome. Respiration. 1996; 15: 47-51.