Multimodal imaging findings in an adult case of Swyer-James-MacLeod syndrome

Hitoshi Kawasuji, Kensuke Suzuki, Hideaki Furuse, Takeshi Tsuda, Yasuaki Masaki & Hirokazu Taniguchi

Department of Internal Medicine, Toyama Prefectural Central Hospital, Toyama, Japan.

Keywords
Multimodal imaging, Swyer-James-MacLeod syndrome, three-dimensional CT reconstruction.

Abstract
Swyer-James-MacLeod syndrome (SJMS) is an uncommon obliterative lung disease that is radiologically characterized by hyperlucency of a part of or the entire lung. A 33-year-old man presented to our hospital for chest tightness. A chest X-ray revealed unilateral hyperlucency of left lower lung, and contrast-enhanced computed tomography (CT) of the chest disclosed a hyperlucent left lung without vascularity. Three-dimensional CT reconstruction and ventilation–perfusion scan findings were concordant with SJMS. We herein report a case of SJMS in a patient who showed the characteristic multimodal imaging findings.

Introduction
Swyer-James-MacLeod syndrome (SJMS) is an uncommon obliterative lung disease, and the radiological findings include hyperlucency of a part of or the entire lung, decreased number and diameter of ipsilateral peripheral pulmonary vessels, and low visibility of the arterial network and unobstructed bronchial system.

In SJMS, the involved lung or portion of the lung does not grow normally. Fibrosis and obstruction of the terminal and respiratory bronchioles, likely caused by respiratory infections in early childhood, prevent the normal development of the alveolar bud [1].

This case report describes the diagnostic approach used in a patient with SJMS, which included multimodal imaging using three-dimensional chest computed tomography (CT) and ventilation–perfusion scintigraphy.

Case Report
A 33-year-old man presented to our outpatient clinic following a 1-week history of chest tightness and dyspnoea at rest. He had no medical history of pulmonary infection or bronchial asthma during childhood or as an adult, and he had a 5-pack-year history of smoking.

On physical examination, his respiratory rate was 18 breaths per minute and oxygen saturation was 96% while breathing room air. Pulmonary auscultation demonstrated decreased breath sounds over the left lung. Findings from complete blood counts, urinalysis, D-dimer test, and blood chemistry were all within normal limits. Respiratory function tests showed an obstructive pattern with a forced expiratory volume of 2.76 L in 1 sec (64% of predicted value) and a forced vital capacity of 4.93 L (110.8% of predicted value).

Chest X-ray revealed unilateral hyperlucency of the left lower lung with a small hilar shadow and reduced bronchovascular markings (Fig. 1A). Contrast-enhanced computed tomography (CECT) showed a hyperlucent left lung without anteroposterior gradient attenuation, hypoplasia or absence of pulmonary artery and peripheral vascular bed, and an overall reduction in the left inspiratory lung volume compared to the right (Fig. 1B). CECT revealed no mass or findings suggestive of pulmonary embolism and there were no patchy areas of focal bronchiolitis, hyperlucency or bronchiectasis in the right lung suggesting bilateral involvement. Three-dimensional CT reconstruction...
showed reduced airway branching, and cylindrical and cystic bronchiectasis scattered over the entire left lung, especially in the left lower lobe (Fig. 1C).

Fiberoptic flexible bronchoscopy was performed, and bronchial stenosis was not observed.

With a strong suspicion of SJMS, a ventilation study was performed with xenon-133. An initial single-breath view was obtained, revealing non-segmental ventilation defects in the periphery of the left lung and slightly diminished ventilation in the periphery of the right upper lobe (Fig. 2A). There was delayed washout of xenon uniformly throughout the left lung (Fig. 2B,C).

A perfusion scan was performed with technetium-99m macroaggregated albumin (Tc-99mMAA). The scans showed non-segmental perfusion defects throughout the left lung, matching the ventilation defects (Fig. 2D). The imaging findings confirmed the diagnosis of SJMS.

Discussion

SJMS was first described in 1953 by Swyer and James and then subsequently in 1954 by MacLeod [2]. Typically, the onset of symptoms occurs during infancy or early childhood in association with frequent respiratory infections. Although SJMS is usually diagnosed during childhood, it can also be diagnosed during adult life with acute respiratory symptoms and recurrent respiratory infections, and in patients who have been diagnosed as asthmatic who appear to be non-responsive to conventional therapy [3].

SJMS is a rare entity associated with post-infectious bronchiolitis obliterans occurring in childhood. The disease starts as acute bronchiolitis with concomitant vasculitis commonly following infections with certain organisms, such as adenovirus types 3, 7, and 21, paramyxovirus, morbillivirus, Bordetella pertussis, Mycoplasma pneumoniae, Mycobacterium tuberculosis, and influenza A [2,4]. Early infection results in inflammation and fibrosis, which causes narrowing of the respiratory bronchiole lumen and reduces blood flow in the main pulmonary artery owing to obliteration of the pulmonary capillary [3]. This reduction in flow creates hypoplasia of the pulmonary arterial bed. These pathophysiologic changes lead to air trapping and hypoperfusion in the affected segment, thus creating radiographic hyperlucent or translucent findings [2].

Diagnosis is made following radiographic evidence of the classic SJMS triad: a unilateral hyperlucent lung, diffusely decreased ventilation, and matching decreased perfusion in the affected lung [1].

The diagnosis of SJMS requires the exclusion of many other causes of unilateral hyperlucent lung on chest radiography, including ventilatory abnormalities such as congenital lobar emphysema, bullous emphysema, bronchiectasis with air trapping, emphysema secondary to bronchial stenosis or bronchospasm, and obliterative bronchiolitis; and pulmonary artery defects such as congenital pulmonary artery agenesis/hypoplasia, acquired stenosis or compression of the main pulmonary vessels, and pulmonary embolism [1–3,5]. None of these ventilatory abnormalities would produce the diffuse, peripheral ventilatory defects on the single breath image and present unilateral lung hypoplasia that the Swyer–James syndrome does.

In pulmonary artery defects and pulmonary embolism, there is no air-trapping on radiological or ventilation-perfusion lung images.
Angiography and bronchography also facilitate the definitive diagnosis, while CT-multiplanar reconstruction can identify most of those afflictions, as in this patient, obviating the need for more invasive procedures in some cases [5].

In the majority of cases with SJMS, treatment is typically conservative and supportive, including close follow-up and management by chest physiotherapy. The most concerning long-term effect of this syndrome is recurrent lung infection, so prompt and effective antibiotic therapy is essential [1,3].

Surgery is an option in patients who experience unremitting infections and is indicated when all other therapies have proven ineffective [2].

In conclusion, we herein reported a case of SJMS with the characteristic presentation on multimodal images. In patients with unilateral hyperlucent lung on X-ray, SJMS should therefore be considered in the differential diagnosis.

**Disclosure Statements**

No conflict of interest declared. Appropriate written informed consent was obtained for publication of this case report and accompanying images.

**Acknowledgment**

We thank Hitoshi Abo, the Department of Radiology, Toyama Prefectural Central Hospital, for providing the radiological suggestions.

**References**

1. Damle NA, Mishra R, and Wadhwa JK. 2012. Classical imaging triad in a very young child with swyer-james syndrome. Nucl. Med. Mol. Imaging 46:115–118.
2. Panagopoulos N, Papavasileiou G, Koletsis E, et al. 2014. VATS bullectomy and apical pleurectomy for spontaneous pneumothorax in a young patient with Swyer-James-McLeod syndrome: case report presentation and literature review focusing on surgically treated cases. J. Cardiothorac. Surg. 9:13.
3. Chaucer B, Chevenon M, Toro C, et al. 2015. Swyer-James-MacLeod syndrome: a rare finding and important differential in the ED setting. Am. J. Emerg. Med. 34:1329.
4. Wojcicki KM, Sindel AD, Berry AC, et al. 2015. An uncommon obliterative lung disease: Swyer-James-MacLeod syndrome. Intern. Emerg. Med. 10:881–882.
5. Chong B, Xiao-lian S, Hui S, et al. 2011. Swyer-James syndrome with peculiar course and ipsilateral pulmonary vein defect. Intern. Med. 50:1829–1833.