



Case Report

Ventilation-perfusion SPECT/CT in Diagnosing Swyer-James (MacLeod) Syndrome

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Abstract

We present the characteristics of a ventilation-perfusion scan correlating to both planar and single photon emission computed tomography and computed tomography (SPECT/CT) images of the lungs in a patient with Swyer-James (MacLeod) syndrome. A Tc-99m MAA lung perfusion scan was recorded with multiple projections and SPECT/CT. In the ventilation scan, initially dynamic acquisition of aerosol Tc-99m DTPA was obtained with an interval of 0.4 seconds per frame, followed by multiple static projections and SPECT/CT. Anatomically co-registered SPECT/CT of perfusion images demonstrated diffusely decreased perfusion without CT chest lesions over the affected radiolucent left lung. The SPECT/CT of ventilation scan showed heterogeneous radiotracer distribution and multiple defects resembling airflow obstruction over the left lung. A dynamic ventilation scan revealed diminished inhaled radioisotope in the left lung compared with the normal right lung. In contrast to over aeration of the left lung observed in the same area on CT, the dynamic hypoventilation implies the presence of air trapping. The SPECT/CT of ventilation-perfusion study showed matched defects in the left lung excluding a residual area of normal density, and permitted a specific reconstructive display to exhibit the pathophysiology of Swyer-James (MacLeod) syndrome with anatomic and functional co-registration. (*Tzu Chi Med J* 2010;22(4):213–218)

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1. Introduction

The ventilation-perfusion (V/Q) scan is a well-established examination in the survey of many pulmonary diseases, such as pulmonary thromboembolism,

and in pre-thoracic surgery evaluation of lung function. In contrast to detecting anatomical changes on computed tomography (CT), a V/Q scan using radio-labeled aerosol Tc-99m diethylene triamine penta-acetic acid (DTPA) and radiopharmaceuticals such as

Tc-99m macroaggregated albumin (MAA), provides a functional image showing changes in pulmonary ventilation and perfusion.

A new technology, single photon emission CT and CT (SPECT/CT), has been used in many nuclear studies to co-register anatomical and functional images. In V/Q scans, SPECT/CT fusion images are applied in diagnoses such as pulmonary embolism and pulmonary emphysema. Here, we present an image study of Swyer-James syndrome with the utilization of V/Q planar and SPECT/CT in the differential diagnosis of unilateral radiolucent lung. The correlation of matched V/Q scan defects with air trapping in SPECT/CT demonstrated the inherent nature of Swyer-James syndrome in obliteration of both the bronchioles and the pulmonary capillary bed.

2. Case report

A 67-year-old man presented to this hospital with a chronic cough for years. He was a carpenter and had no history of smoking. The coughing was especially exacerbated at night. He claimed exercise intolerance since he was a child. Spirometry revealed a forced vital capacity (FVC) of 2.57 liters (61.6% of predicted value), first second forced expiratory volume (FEV_1) of 1.43 liters (44.2% of predicted value), and a FEV_1/FVC of 55.6%. Chest radiography showed an essentially normal right lung parenchyma but hyperlucency of the entire left lung with decreased lung markings (Fig. 1). Contrast-enhanced CT showed a prominent right pulmonary artery and relatively hypoplastic left pulmonary artery in the secondary carina levels. The lung window of the same levels showed hyperlucency with air trapping in the left lung and bronchiectasis in the left lower lobe (Fig. 2). Neither endobronchial tumor nor foreign body obstruction was noted. Nuclear ventilation study (V) with Tc-99m DTPA aerosol (25 mCi) and perfusion study (Q) with Tc-99m MAA (5 mCi) showed global decreased activity with V/Q matched defects in the peripheral region of the left lung (Fig. 3). However, the acquisition of the V/Q scan with SPECT/CT revealed that the diminished perfusion and ventilation areas matched the sites of diminished lung markings in the left lung on the low-energy (2.5 mA) CT. In addition, there was a residual area of normal density in the left B3 segment near the hilar area with normal perfusion and ventilation shown on V/Q SPECT/CT (Figs. 4 and 5). The dynamic time activity curve of ventilation of Tc-99m DTPA aerosol inhalation in the left lung compared with the right lung (Fig. 6), although the left lung seemed to have a larger volume on CT (Fig. 6A). The results of the V/Q scan in combination with SPECT/CT demonstrated the unique pathophysiology and anatomical changes of Swyer-James (MacLeod) syndrome.



Fig. 1 — Chest radiography reveals unilateral hyperlucency of the entire left lung with decreased lung markings. The nodular lesion inferior to the left hilum proved to be cryptogenic organizing pneumonia.

3. Discussion

Swyer-James (MacLeod) syndrome, named after the authors who first described this disease in 1953 and 1954 (1), is characterized by hyperlucency of a unilateral lung with scarce lung markings (2). The principal pathologic findings are decreases in the number and size of branches of the pulmonary arteries, leading to so-called hypoplastic vessels, on the affected side (3). Swyer-James syndrome is considered a form of bronchiolitis obliterans (4,5). Submucosal and peribronchial fibrosis destroys and scars the small airways (6). Accordingly, the disease may affect pulmonary function and sometimes causes bronchiectasis. Occasionally, patients with this syndrome are discovered incidentally while receiving chest radiography for other causes or health examination, or are misdiagnosed with chronic obstructive pulmonary disease.

Other etiologies causing unilateral hyperlucent lung include pulmonary vascular occlusion, pulmonary artery agenesis, post-lobectomy lung, mastectomy, and bronchial obstruction from benign or malignant entities, including foreign bodies, mucus plugs, and intrabronchial neoplasms (1,7). The approach to the differential diagnosis of the chest radiographic findings needs both anatomical and functional evaluation. In pulmonary vascular occlusion and pulmonary artery agenesis, the perfusion scan should be defective but the ventilation scan is normal. In post-lobectomy lung or mastectomy, the V/Q scan is essentially normal without air trapping. Bronchial obstruction from benign or malignant entities usually exhibits complete absence of ventilation to the whole lung in a proximal lesion or diminished ventilation to a selected segment or lobe in a peripheral lesion with or without a localized perfusion defect, or generally diminished

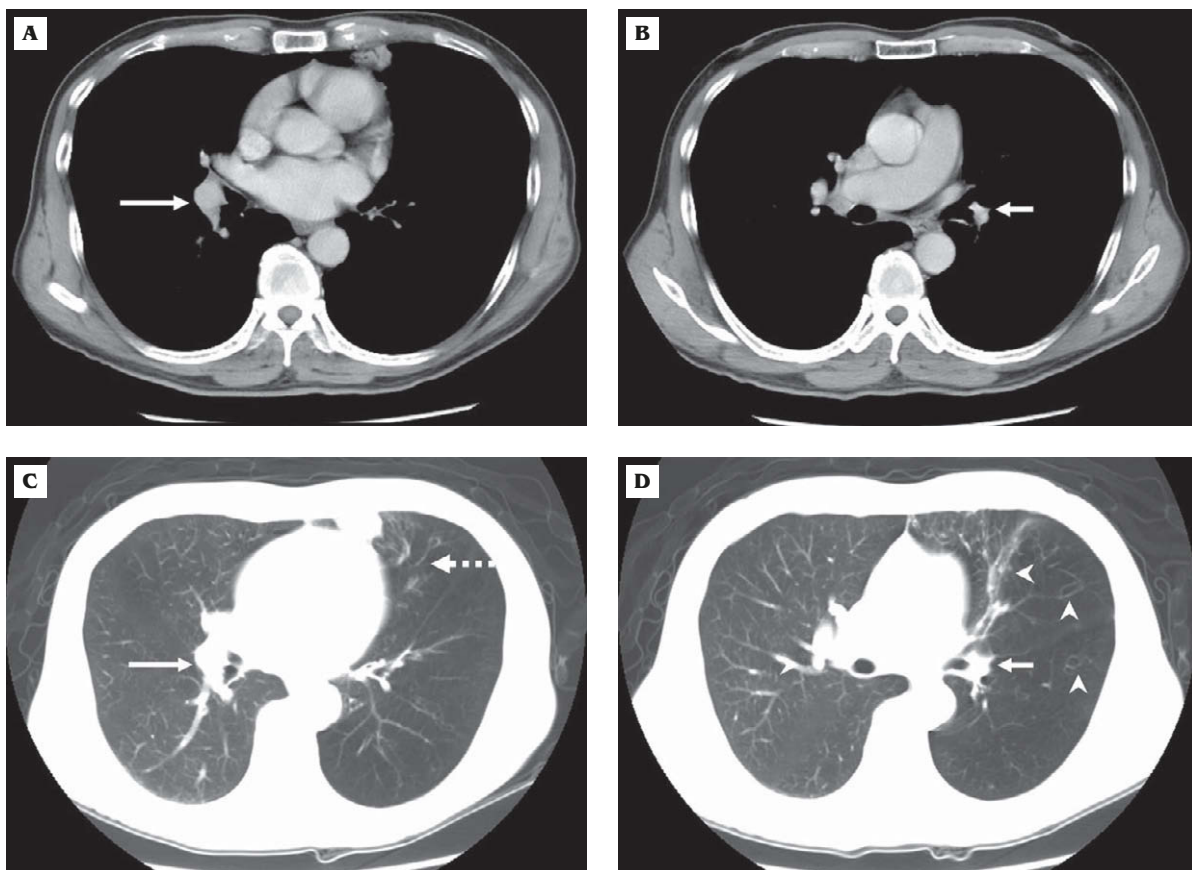


Fig. 2 — (A) Contrast-enhanced CT shows a prominent right pulmonary artery in the secondary carina (arrow). **(B)** Contrast-enhanced CT shows a relatively hypoplastic left pulmonary artery in the secondary carina (arrow). **(C)** Lung window of panel A shows hyperlucency of the left lung with residual normal lung markings in the left B3 segment (dotted arrow). **(D)** Lung window of panel B shows bronchiectasis (arrowhead) in the left lower lobe.

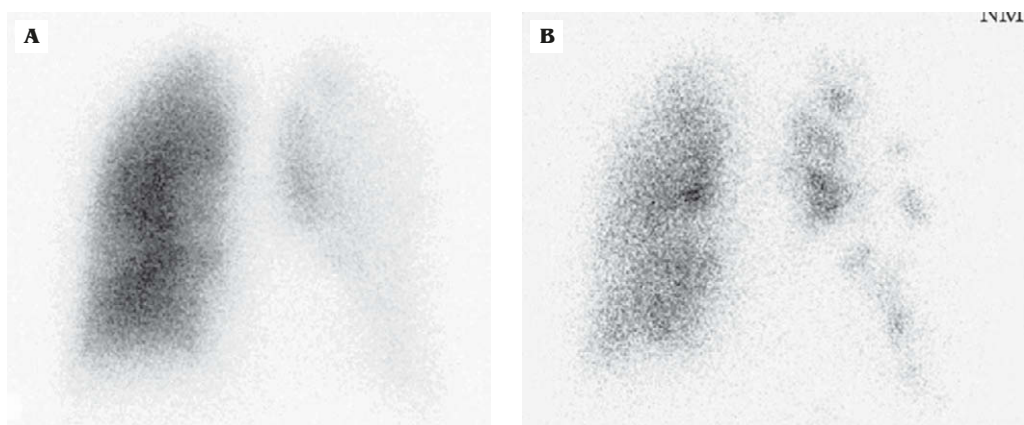


Fig. 3 — (A) Perfusion study with Tc-99m MAA shows diffusely diminished perfusion to the left lung and normal uptake in the right lung without any perfusion defects. There is some perfusion to the medial side of the left lung, indicating residual normal pulmonary vasculature. **(B)** Ventilation study with Tc-99m DTPA aerosol shows multiple defects over the left upper and lower lung. The peripheral and medial uptake to the left lung implies a preserved ventilation area there. The right lung is homogeneously ventilated without any filling defects.

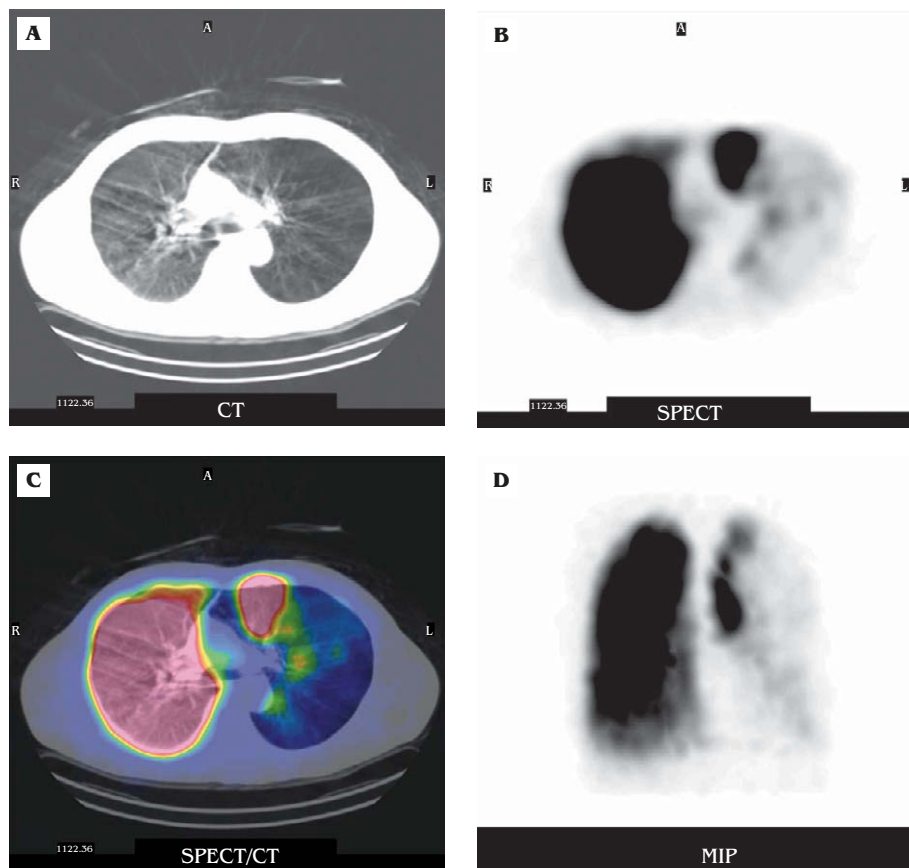


Fig. 4 — The low-energy (2.5 mA) non-diagnostic CT (A), SPECT (B), fused SPECT/CT (C) and MIP view (D) in the perfusion study performed after the multiple planar views. SPECT/CT demonstrates diminished radiouptake over the areas of decreased lung markings in the anatomic and functional co-registered images. Notice the increased left lung volume shown on panel A with a right shift of the mediastinum. Residual perfusion of the left medial lung is compatible with the residual normal lung markings (on CT) in the left B3 segment. This reveals the inherent characteristic hypoplastic vascular tree of the unilateral hyperlucent lung on radiology.

perfusion defect corresponding to the lesion on a lung scan. Swyer-James syndrome typically shows matched V/Q defects with air trapping, diffusely diminished blood flow to the hyperlucent area, and also non-segmental defects in the peripheral area of the affected lung on a single breath image with xenon ventilation.

Air trapping is an important manifestation of Swyer-James syndrome, reflecting the pathophysiologic features of bronchiolitis obliterans (5). Correlation with a history of pulmonary infection such as adenovirus, measles, pertussis, tuberculosis, or mycoplasma in childhood or foreign body inhalation may be helpful in the diagnosis of Swyer-James syndrome. These insults might lead to inhibition of the normal growth of the affected lung (8). Those with bronchiectasis could have episodes of pulmonary infection or hemoptysis, and may even require surgical lobectomy (9). It is impossible to make a conclusive diagnosis of this disease based solely on either a V/Q scan or chest CT. In our case, the combination of V/Q scan and SPECT/CT

were helpful in the diagnosis. A V/Q scan provides diagnostic assistance by showing matched defects, and invasive procedures such as bronchoscopy, bronchography and angiography can be avoided. SPECT/CT of perfusion images demonstrates the co-registered anatomic features of the hypoplastic vascular tree and functional hypoperfusion area. A dynamic Tc-99m DTPA aerosol ventilation scan is acquired first, and the time activity curve demonstrated globally diminished ventilatory function of the left hyperlucent lung. Most importantly, SPECT/CT of ventilation images demonstrated that the hyperaerated area of the lung (hyperlucency) just co-localized to the functional hypoventilated area, a typical picture of air trapping. These imaging findings disclosed the inherent nature of Swyer-James syndrome and indicated the disease etiology to be the obliteration of the bronchioles and pulmonary capillary bed with secondary air trapping (7). The residual peripheral uptake to the left lung implying distal air flow might indicate an inhomogeneous obliteration of the airway and the nature

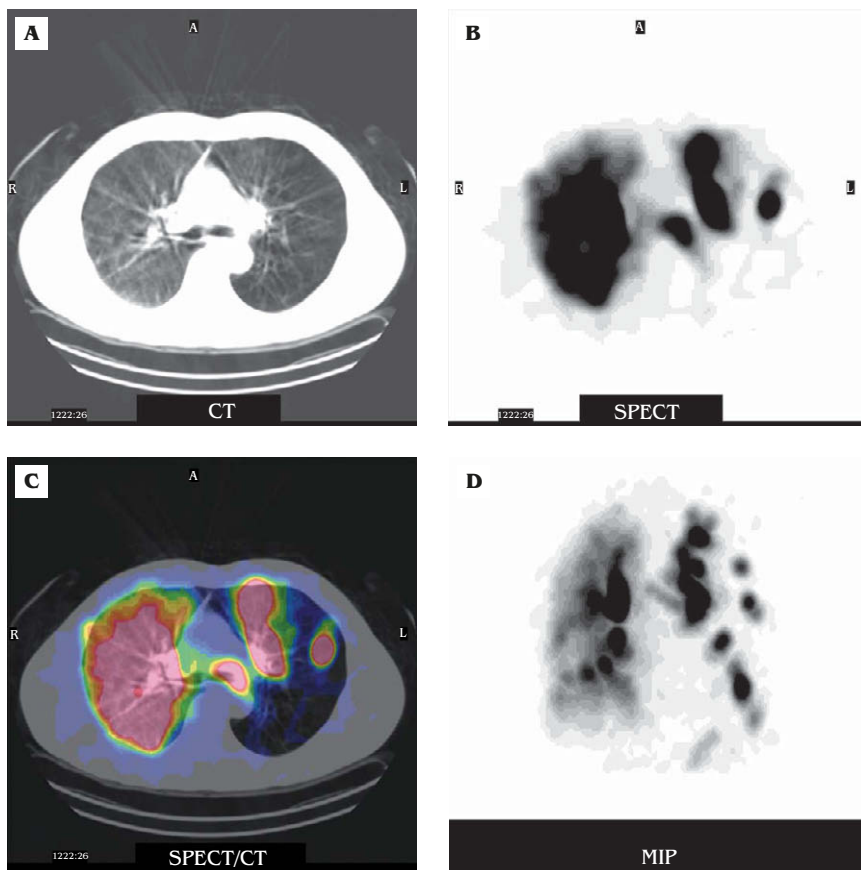


Fig. 5 — The low-energy (2.5 mA) non-diagnostic CT (A), SPECT (B), fused SPECT/CT (C) and MIP view (D) in the ventilation study performed after the multiple planar views. SPECT/CT demonstrates multiple ventilation defects over the areas of decreased lung markings in the left lung in the anatomic and functional co-registered images. Residual ventilation of the left medial lung is compatible with the residual normal lung markings (on CT) in the left B3 segment. This reveals the inherent characteristics of airflow obstruction in the bronchioles in the unilateral hyperlucent lung on radiology.

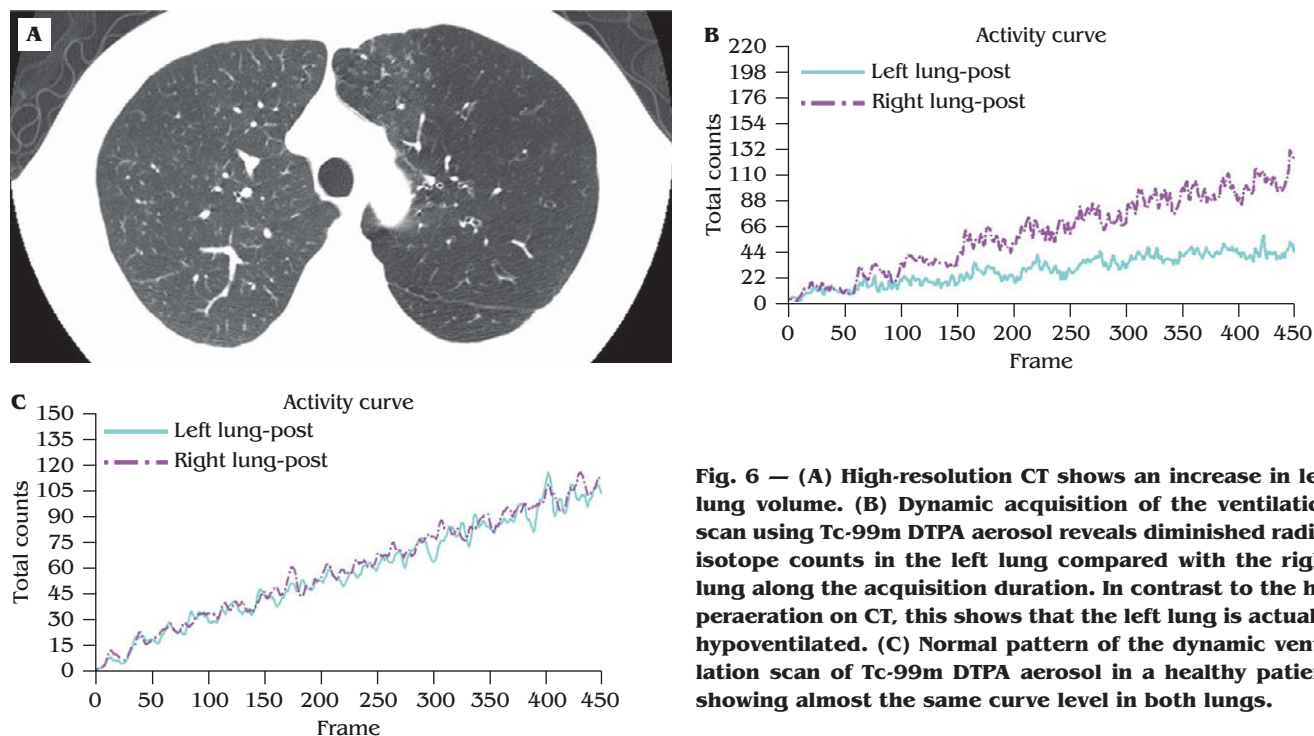


Fig. 6 — (A) High-resolution CT shows an increase in left lung volume. (B) Dynamic acquisition of the ventilation scan using Tc-99m DTPA aerosol reveals diminished radioisotope counts in the left lung compared with the right lung along the acquisition duration. In contrast to the hyperaeration on CT, this shows that the left lung is actually hypoventilated. (C) Normal pattern of the dynamic ventilation scan of Tc-99m DTPA aerosol in a healthy patient showing almost the same curve level in both lungs.

of deposition of the aerosol. In contrast, the obliteration of the pulmonary capillary bed was more homogeneous. Since Swyer-James syndrome is an acquired disease, there could be some preserved portion with residual ventilation and perfusion in the destroyed lung. This cannot be detected solely by CT or planar V/Q scan

Nuclear V/Q scan, a noninvasive procedure, offers diagnostic assistance in differentiating the possible etiologies of unilateral hyperlucent lung shown on chest radiography. However, as in the present case, we used SPECT/CT in combination with V/Q scan to provide both anatomic and functional information in one study. We think this offers additional benefits for clinical judgment and is pathognomic in demonstrating the pathophysiological features of Swyer-James syndrome.

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