

## Swyer-James-Macleod Syndrome: A Case Report

### Swyer-James-Macleod Sendromu: Olgu Sunumu

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#### Summary

We report a case of Swyer-James-MacLeod Syndrome who was admitted to our hospital with an intermediate probability ventilation/perfusion scintigraphy for pulmonary embolism. There was a diffuse matched ventilation/perfusion defect on the left lung. Decreased volume and hyperlucency of left hemithorax, smaller than normal left hilus were seen in chest X-ray. Spiral computerized tomography of thorax revealed marked decreased vascularity together with decreased volume and hyperlucency of left hemithorax, left hypoplastic pulmonary artery and diffuse saccular bronchiectasis at lingula and left lower lobe. Pulmonary embolism was not observed. The patient was diagnosed as Swyer-James-MacLeod Syndrome due to clinical and radiological findings together with ventilation/perfusion scintigraphy. We recommend that this syndrome should be considered in case of unilateral matched ventilation/perfusion defect and spiral computerized tomography of thorax is an important procedure in differential diagnosis. (*Archives of Lung 2007; 8: 65-7*)

**Key words:** Swyer-James-MacLeod Syndrome, ventilation/perfusion scintigraphy, pulmoner embolism, computerized tomography

#### Özet

Ventilasyon/perfüzyon sintigrafisinde orta olasılıklı pulmoner emboli düşünülerek hastanemize sevk edilen bir Swyer-James-MacLeod sendromu olgusu sunulmuştur. Sintigrafide sol akciğerde diffüz, uyumlu ventilasyon/perfüzyon defekti saptanmıştır. Akciğer grafisinde solda volüm kaybı, saydamlık artışı mevcuttu ve sol hilus normalden küçüktü. Toraksın spiral bilgisayarlı tomografisinde sol akciğerde saydamlık artışı, volüm kaybı ile birlikte vaskülaritede belirgin azalma, sol pulmoner arterde hipoplazi ve sol alt lob ve üst lob linguler segmentlerde yaygın sakküler bronşektazi izlendi. Pulmoner emboliyi düşündürecek bulguya rastlanmadı. Ventilasyon/perfüzyon görüntüleri ile radyolojik ve klinik bulgular birlikte değerlendirildiğinde hastaya Swyer-James-MacLeod sendromu tanısı kondu. Sunumuzda sintigrafide tek taraflı, uyumlu ventilasyon/perfüzyon defekti saptandığında, Swyer-James-MacLeod sendromunun da düşünülmesi gerektiği ve ayırıcı tanıda spiral toraks bilgisayarlı tomografinin önemi vurgulanmak istendi. (*Akciğer Arşivi 2007; 8: 65-7*)

**Anahtar Kelimeler:** Swyer-James-MacLeod sendromu, ventilasyon/perfüzyon sintigrafisi, pulmoner emboli, bilgisayarlı tomografi

#### Introduction

Swyer-James-MacLeod Syndrome (SJMS) is known as unilateral emphysema or unilateral hyperlucent lung (1). It is characterized by hyperlucency, decreased number and size of pulmonary vascular structures and bronchiectasis (2-4). This syndrome is thought to develop secondary to childhood

infectious diseases. Diagnosis is based on radiological signs rather than clinical. Endobronchial lesions, unilateral bullous diseases and pulmonary arterial pathologies must be considered in differential diagnosis. Computerized tomography (CT) of thorax, pulmonary angiography, bronchography and ventilation/perfusion (V/Q) scintigraphy are the diagnostic methods (2,4-6). CT is superior to other methods with regard

to detection of parenchymal changes in the affected lung, status of the opposite lung and accompanying diseases and is also used for differential diagnosis (2-4).

We reported this case since the syndrome is rarely seen and the patient was referred as pulmonary embolism.

## Case

The patient was a 56 years old woman admitted to our department with an intermediate probability V/Q scintigraphy for pulmonary embolism. The scintigraphy revealed a diffuse matched V/Q defect on the left lung. Her complaints were dyspnea on exercise, cough and sputum increasing at winter times but not hemoptysis. There has been an occasional pain and swelling on the left leg for eight years. She had no history of smoking or pneumonia. She had medication with the diagnosis of asthma for many times. There was nothing remarkable in her family history.

On physical examination, blood pressure was 110/70 mmHg, pulse was 82/min, fever was 36.4°C, respiratory rate was 21/min. Rales were heard on left infrascapular region and rhonchi bilaterally. Examination of other systems was unremarkable

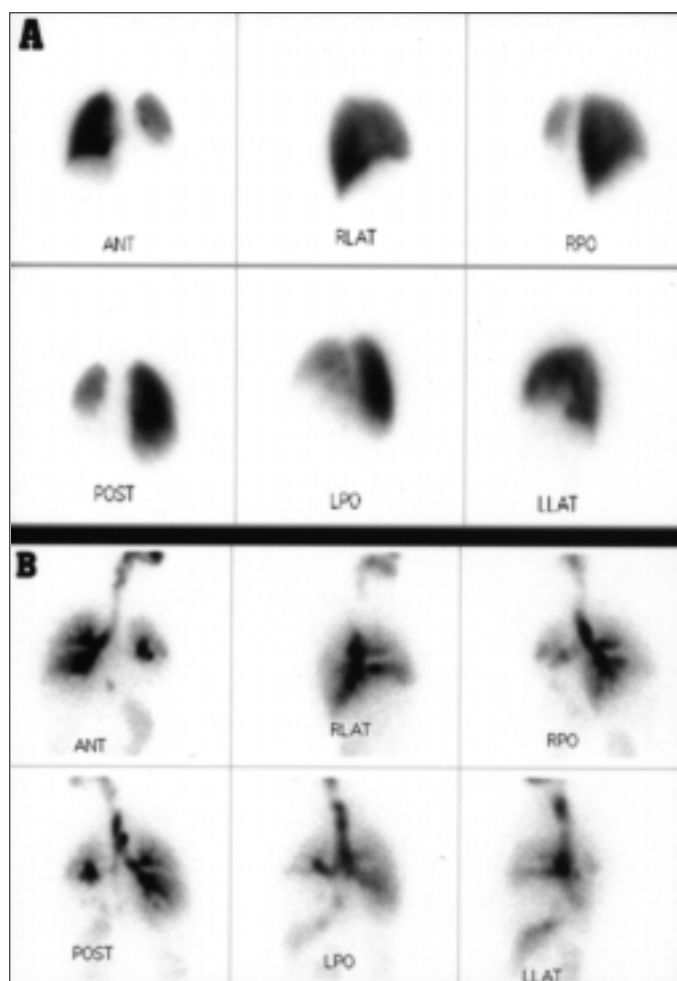


Figure 1. Perfusion scan (A) showing decreased perfusion to the left lung, ventilation scan (B) showing significantly diminished ventilation to the left lung matching the perfusion scan seen in 1A.

and there was no finger clubbing. Erythrocyte sedimentation rate was 29 mm/hr. Complete blood count and biochemical evaluation of blood were normal. Decreased volume and hyperlucency of left hemithorax, smaller than normal left hilus were seen in chest X-ray. V/Q scintigraphy showed diffuse matched defect on the left lung (Figure 1A, 1B). Spiral CT of thorax revealed marked decreased vascularity together with decreased volume and hyperlucency of left hemithorax, diffuse saccular bronchiectasis at lingula and left lower lobe and left hypoplastic pulmonary artery (Figure 2A, 2B and 2C). Pulmonary embolism was not observed. Venous doppler ultrasonography of left lower extremity demonstrated deep venous insufficiency at the level of popliteal vein. On evaluation of arterial blood gases, pH:7.42, pCO<sub>2</sub>:39 mmHg, pO<sub>2</sub>:79 mmHg, O<sub>2</sub> saturation: 97%. Moderate reversible obstructive and restrictive ventilatory defects were detected on spirometry. Pulmonary embolism was excluded and the patient was diagnosed as SJMS.

## Discussion

This rarely seen case of unilateral hyperlucency syndrome was first reported by Swyer and James in 1953 and MacLeod one year later. Prevalence of the syndrome was reported to be 0.01% in a survey of 17,450 chest radiographs (1). Patients are usually asymptomatic and radiological findings are noticed coincidentally. It may involve one or more lobe of one lung and one or more segment of one lobe, whereas it is rarely bilateral (2,3,5). In our case, the lesion was localized in lingula and left inferior lobe.

It is thought to be postinfectious form of bronchiolitis obliterans where there is destruction and obliteration in small airways together with submucosal and peribronchiolar fibrosis. It is reported to be secondary to childhood infections such as adenovirus, measles, pertussis, RSV, influenza, tuberculosis and mycoplasma infections (4). Bronchiectasis is reported to be present in some cases with SJMS. Adult patients are usually asymptomatic, whereas particular patients have dyspnea on exercise, chronic cough and sputum, wheezing and hemoptysis (2,4,5,7,8). In our case who does not have a history of childhood infectious disease, there was cough, sputum, dyspnea on exercise and wheezing increasing in winter times secondary to bronchiectasis but no hemoptysis. SJMS was reported to be seen together with lung cancer, Goodpasture syndrome, myocardial bridge, spontaneous multi-vessel coronary dissection in the literature (9-12).

Diagnosis depends on radiological findings such as lobar or unilateral hyperlucency, airway obstruction during expiration, decreased vascularization, smaller sized central and peripheral pulmonary arteries. Involved lung is smaller than the normal lung during inspiration and mediastinum shifts to the uninvolved hemithorax during expiration (2,3,7,13). Size of the involved lung depends on the age that bronchiolitis develops. In case of the infection occurring in earlier ages, development of the lung is incomplete, whereas it may reach normal size after an infection in late childhood (5). In pulmonary angiography, pulmonary arteries are decreased in number, size and diameter (4,13). In V/Q scintigraphy, matched defect is seen at the involved region of the lung and air trapping in

ventilation scintigraphy in the same region (3,4,6,14). On bronchography, diffuse bronchiectasis is seen with "pruned tree" appearance due to radiopaque filling defect (5). Central airway obstruction, parenchymal cysts or bulla, pulmonary vascular diseases may be considered in differential diagnosis of SJMS where CT of thorax is very helpful and superior to

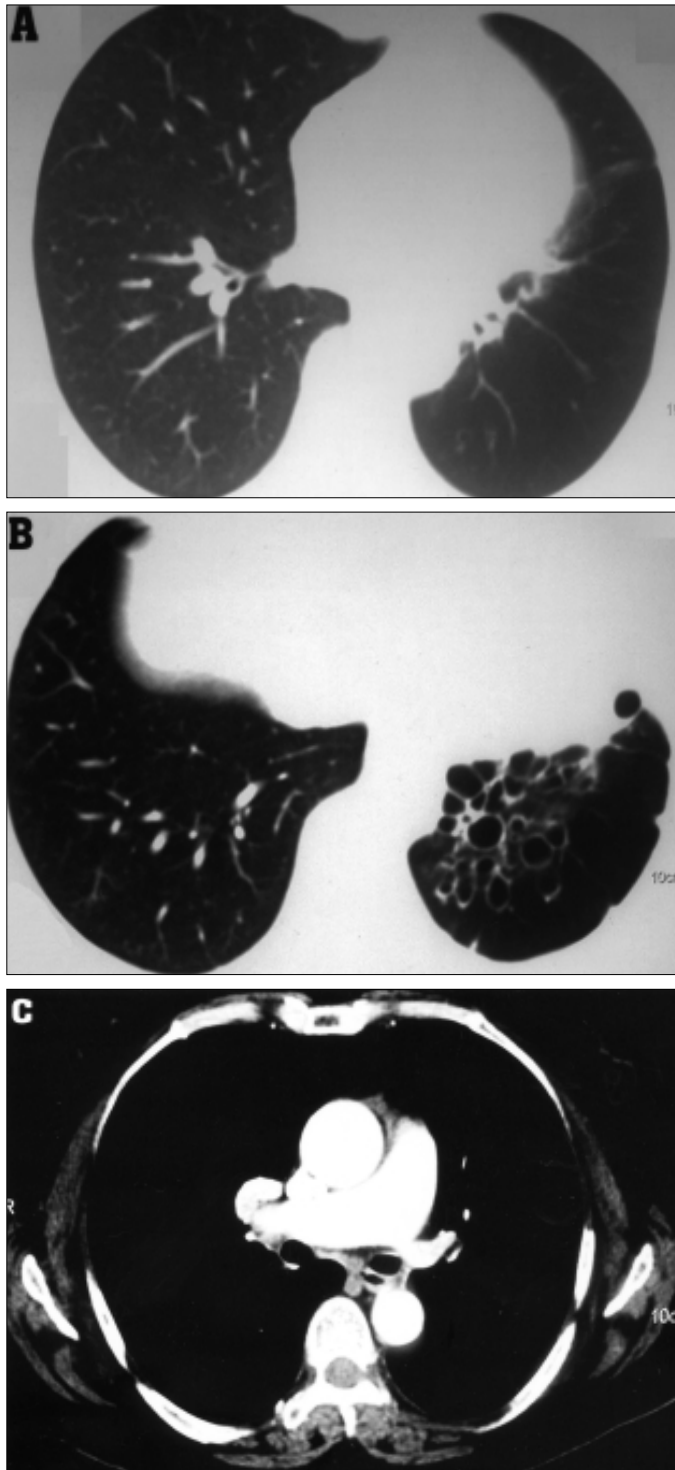


Figure 2. CT scan of the chest with contrast showing peripheral wedge-shaped densities, diminished lung volume and hyperlucency, decreased vascularity (A) and saccular bronchiectasis (B) in the left lung and left hypoplastic pulmonary artery (C).

V/Q scintigraphy with regard to determining hyperlucency, extent and localization of the involvement. It is also useful in diagnosis of bronchiectasis (2-4,7). On the other hand, spiral CT is preferred to V/Q scintigraphy in the evaluation of pulmonary embolism due to detection of parenchymal lesions and exclusion of other pathologies (15). Spiral CT was able to diagnose SJMS, exclude the diagnosis of pulmonary embolism and detect bronchiectasis in our case with an intermediate probability ventilation/perfusion scintigraphy for pulmonary embolism. Treatment is symptomatic in SJMS. Surgery is indicated in case of recurrent hemoptysis and infections. Surgical methods are segmental or lobar resection, pneumonectomy and occlusion of main bronchus (14,16). Surgery was not considered in our case due to absence of recurrent infections and hemoptysis. Inhaled bronchodilator and corticosteroid medication was started since pulmonary function test revealed reversible airway obstruction. Influenza and pneumococcal vaccines were also recommended.

As a conclusion, spiral CT of thorax is valuable for differential diagnosis and evaluation of lung parenchyme of SJMS where there is diffuse matched V/Q defect and superior to V/Q scintigraphy in diagnosis of pulmonary embolism.

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