

# Swyer-James-MacLeod Syndrome

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## ABSTRACT

This case report describes a patient with Swyer-James-MacLeod Syndrome (SJMS) in an adult male diagnosed on the basis of findings on X-ray chest, high resolution CT (HRCT) of chest and radionuclide perfusion lung scan. This rare syndrome is considered to be an acquired disease due to repeated pneumonias in early childhood.

**Key words:** Swyer-James syndrome. Chest X-ray. High resolution computed tomography. Perfusion scan.

## INTRODUCTION

Swyer-James syndrome also known as MacLeod syndrome, first described in 1953, is an uncommon disease characterized by hyperlucency of one lung, lobe or part of lobe due to pulmonary vascular abnormalities and alveolar overdistension.<sup>1</sup> Emphysema, bronchiectasis or bronchiolitis obliterans are the cardinal pathological features.<sup>2</sup>

The syndrome is of unknown etiology although repeated episodes of viral bronchiolitis or viral pneumonias are the speculated cause.<sup>1</sup> Patients usually present as adults and prior history of childhood infections is not obtained in most cases.<sup>3</sup>

This report describes a case of Swyer-James syndrome diagnosed in an adult male.

## CASE REPORT

A 30 years old man presented with history of gradually progressive dyspnea, now grade II, for last 6 months. There was no history of associated cough, sputum, fever, anorexia or weight loss. He had no past history of tuberculosis, asthma, childhood pneumonias, whooping cough or measles. He had received complete coverage of Expanded Program of Immunization (EPI) in early childhood. He was a non-smoker and driver by occupation, working in Saudi-Arabia. There was no history of exposure to pets, farm animals, organic or inorganic dust. He was married for the last 3 years with no issue and there was no history of extra-marital contacts.

On examination, he had respiratory rate of 28/minute with tracheal shift to the right, hyper-resonant

percussion note and decreased breath sounds on the left side of chest. On investigations, Montoux's test, C-reactive protein (CRP) and sputum for AFB were negative. ESR was 7 mm at the end of first hour. Serum alpha-1 antitrypsin levels were within normal limits. Analysis of arterial blood gases showed pH of 7.55 with PaO<sub>2</sub> of 143.5 mmHg, PaCO<sub>2</sub> of 22.8 mmHg and arterial oxygen saturation of 99.2%. Spirometry showed restrictive ventilatory defect.

X-ray chest PA film showed marked hyperlucency of left lung with passive collapse of right lung and mediastinal shift to the right (Figure 1).

HRCT of chest showed pan lobular and bullous emphysema of left lower lobe with marked air trapping, small sized left pulmonary artery, mediastinal shift to right, partial loss of volume left upper lobe and right lung as shown in Figure 2.

Radionuclide perfusion lung scan done with Tc 99m showed absent perfusion in left lung and sub-optimal perfusion in right lung as shown in Figure 3.

He was given supportive treatment including bronchodilators, antibiotic courses as and when required and was advised to have pneumococcal and influenza vaccines on regular basis as per schedule.

Patient showed symptomatic improvement with treatment and was advised to have regular follow-up with his physician in Saudi Arabia.

## DISCUSSION

Swyer-James syndrome is an uncommon cause of localized hyper-transradiance of lung. It is characterized by unilateral hyperlucency of lung on X-ray chest, unilateral reduction in vascularity on chest CT and unilateral loss of perfusion on lung scan.<sup>4,5</sup> Due to unknown factors, it usually involves the left lung.<sup>6</sup>

It is a form of obliterative bronchiolitis with concomitant vasculitis following injury to immature lungs during the first 8 years of life. It usually follows infections with organisms like adenovirus, measles or pertussis. Both

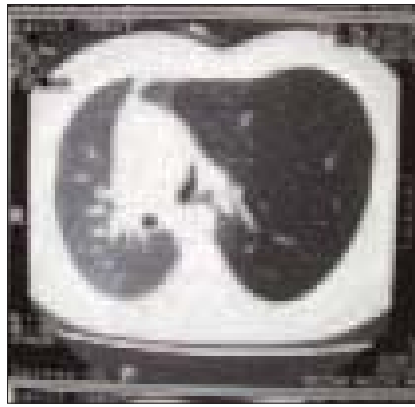
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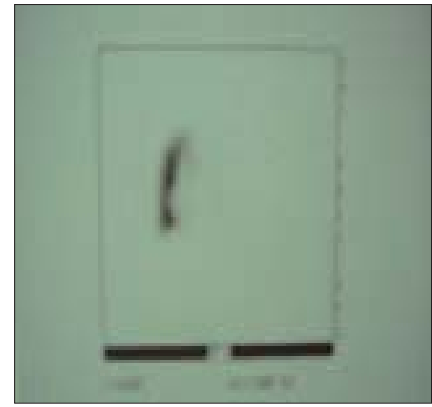
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**Figure 1:** X-ray chest showed collapsed right lung, hyperventilated left lung and mediastinal shift to the right.



**Figure 2:** HRCT chest showing air trapping with reduced vascularity on left side.



**Figure 3:** Perfusion lung scan showing reduced vascularity in the right lung with absent vascularity on the left side.

small bronchi and bronchioles are affected and the lung with abnormal airways remains inflated by collateral air drift.<sup>7</sup> This damage during the early childhood prevents normal development of the alveolar ducts. Airways develop submucosal fibrosis leading to luminal irregularity and occlusion. Pulmonary vasculature is hypoplastic while the lung distal to diseased bronchioles become hyperinflated and sometimes pan acinar emphysematous changes develop.<sup>7</sup>

Patients usually present with gradually progressive dyspnea and repeated chest infections because of associated bronchiectasis, although this patient denied any history of cough with copious purulent sputum.<sup>2</sup>

Spirometry in these patients usually show restrictive defect as in this case, though a significant proportion of patients can have obstructive pattern due to associated bronchiolitis and bronchiectasis.<sup>7</sup>

The disease as assessed radiologically is predominantly unilateral so other causes of hyperlucent lung like pneumothorax, asymmetric emphysema, congenital lobar emphysema (CLE), pulmonary artery hypoplasia, pulmonary embolism and bronchial obstruction due to foreign body or mucus plugs should be ruled out.<sup>1</sup> Hyperlucency in this case is due to reduced perfusion and air trapping. Pulmonary vessels are reduced on the affected side but lung volumes are only slightly decreased. Ipsilateral air trapping is a key finding.<sup>7</sup>

The characteristic radiographic findings in these patients include unilateral hyperlucent lung along with decreased broncho vascular markings, a small hilar shadow and slight displacement of the mediastinum to the affected side. In this case, mediastinum was shifted to the contralateral side because of the long-standing nature of the illness and severe emphysematous changes in the left lung.<sup>10</sup>

CT scan findings of Swyer-James syndrome include patchy areas of low attenuation and hypovascular areas

interspersed with areas of normal attenuation. Air trapping is confirmed on expiratory scans. Other changes on CT may include bronchiectasis, bronchiolectasis, atelectasis and scarring.<sup>8,9</sup> Bronchiectasis, though not a universal finding, evident in only 30% of patients, affects the clinical manifestations and prognosis of the disease. Patients who had saccular bronchiectasis usually have more severe exacerbations than those who do not have bronchiectasis on HRCT. In this case, bronchiectasis was not identified on HRCT chest and this may explain his apparent lack of symptoms for a longtime till he developed gradually progressive dyspnea.

Pulmonary ventilation-perfusion scintigraphy shows marked decrease in ventilation, vascular flow and perfusion in the affected regions.<sup>3</sup> This patient showed similar findings on perfusion lung scan.

Pulmonary angiography, though not essential for diagnosis, shows markedly diminished size of the affected pulmonary artery. It cannot, though reliably differentiate congenital from acquired causes of hypoplastic pulmonary vasculature. Pulmonary angiography was not performed in this patient for this reason.

Treatment is largely supportive with early control of super-added infections along with influenza and pneumococcal vaccination.<sup>1</sup> Bronchodilators may help, especially if the spirometry shows obstructive defect. Long-term oxygen therapy is required in cases with advanced disease and respiratory failure. Surgery in the form of lobectomy or pneumonectomy can be offered to those who had severe symptoms despite optimal therapy. Prognosis largely depends on presence of associated bronchiectasis.<sup>4,10</sup> This patient had severe emphysema but did not have bronchiectasis, so prognosis in this individual case may be good. Regular follow-up and ancillary care is required.

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