
Swyer-James-Macleod Syndrome in a Sixty Year Old Patient

Patricia McWalter

Amal Al-Shmassi.

MRCGP

MRCP

Abstract:

Swyer-James-Macleod syndrome (SJMS) is an uncommon, acquired condition characterised by unilateral hyperlucency of the lung and associated with the development of emphysema, bronchiectasis and/or bronchiolitis obliterans ⁽¹⁾. It is thought to be due to sequelae of childhood respiratory infections ⁽²⁾. In SJMS, the involved lung or portion of the lung does not grow normally and is slightly smaller than the opposite lung ⁽³⁾. This case demonstrates the importance of considering SJMS in patients with recurrent respiratory infections and unilateral lung hyperlucency on chest x-ray.

We report the case of a 60 year old man who has a long history of recurrent sinusitis and respiratory tract infections. Although he had been diagnosed with bronchiectasis at a younger age, it wasn't until the age of 60, that Computed Tomography (CT) of chest confirmed Swyer-James syndrome. This case demonstrates the importance of considering SJMS in patients with recurrent respiratory infections and unilateral hyperlucency on chest x-ray.

Key words: Respiratory system, Swyer-James-Macleod Syndrome Ear, nose & throat

Case report:

This Saudi male patient was first diagnosed with Swyer-James-Macleod syndrome (SJMS) at age 60 at King Faisal Specialist Hospital and Research Centre in 2007.

Prior to this diagnosis, the patient had a long history of allergic rhinitis and conjunctivitis. We were unable to find records of childhood infections but the patient remembers being prone to respiratory tract infections as a child. He has a history of hiatus hernia and peptic ulcer disease. He has been an intermittent light smoker over the years. He is allergic to penicillin. Later, he developed recurrent chest infections and was told he had bronchiectasis and emphysema after having CT chest at another hospital.

When seen in November 2007, he was complaining of a productive cough and rhinorrhoea.

Clinical examination showed normal observations except an oxygen saturation of 94%. Chest exam revealed bilateral inspiratory and expiratory rhonchi with right basal crepitations.

Cardiac, abdominal and neurological examinations were normal. Routine haematological and coagulation studies were within normal ranges. Blood sugar, liver function and renal function tests were also within normal ranges.

Radiology Investigations:

Pulmonary function tests showed signs of early obstructive airway disease. CT chest (see **figure 1**) was carried out at our hospital as he started to have intermittent haemoptysis-it was reported as follows: 'The left lung appears smaller and more lucent, particularly the left lower lobe. Bronchiectasis of the left lower lobe with some opacified dilated bronchioles representing either mucus plugging and/or superimposed infection. There is minimal bronchiectasis of the left upper lobe with compensatory hyperinflation and minimal bronchiectasis of the right lower lobe and the right middle lobe. Non-enlarged calcified mediastinal and right hilar lymph nodes. The overall appearance is consistent with Swyer-James syndrome'. **Figure 2** shows a recent chest x-ray. This patient has left lung

involvement which is usually the case in SJMS. Chest x-ray showed a hyperlucent but small left lung with diffuse scarring and peribronchial thickening of the left lower base.

Differential diagnosis:

Other causes of chronic unilateral hyper lucent lung include localised emphysema, congenital hypoplastic pulmonary artery, previous massive pulmonary embolism, bronchial carcinoma, sequelae of radiation therapy, and benign intra bronchial neoplasm ^(2,3).

Treatment:

The patient was referred to Pulmonology for further assessment and investigations but he did not attend. He was treated medically. He was commenced on symbicort (budesonide/formoterol) and salbutamol inhalers as well as antibiotic therapy for intercurrent infections. He was strongly advised to stop smoking. He remains stable.

Discussion:

This condition was first described by Swyer and James in 1953⁽⁴⁾ and by Macleod in 1954 ⁽⁵⁾. It is also called Bret's syndrome and Janus syndrome. The syndrome appears to be an acquired disease that develops after viral bronchiolitis and/or viral pneumonia in childhood ⁽⁶⁾. It is a rare constrictive bronchiolitis with inflammatory characteristics as reported in a study by Bernardi et al when bronchoalveolar lavage from two patients with SJS showed an expansion in situ of immunocompetent cells ⁽⁷⁾. Constrictive bronchiolitis is a bronchiolar airway disease that leads to fibrotic concentric narrowing and luminal obliteration ⁽⁸⁾. The fibrosis and obstruction of the terminal and respiratory bronchioles, likely caused by respiratory infections in early childhood, prevents normal development of the alveolar bud⁽⁹⁾ Organisms linked to SJMS include respiratory syncytial virus, influenza virus, *Mycoplasma pneumonia*, *Bordetella pertussis*, staphylococcal and streptococcal infections. Patients with SJMS can be asymptomatic and the condition may be an incidental finding on chest x-ray⁽¹⁰⁾.

Others may present with recurrent pulmonary infections and chronic respiratory symptoms as our patient did. Clinical manifestations and prognosis depend mainly on the presence of other lesions, for example bronchiectasis ⁽¹¹⁾. Patients without bronchiectasis or cylindrical bronchiectasis had a lower incidence of pneumonia episodes than those with saccular bronchiectasis ⁽¹²⁾.

One of the reasons to explain difficulty in diagnosis is that when patients develop little bronchiectasis, and therefore, few symptoms, then this syndrome may not be recognised until adulthood. It can mimic other pulmonary disorders which may lead to incorrect diagnosis and inappropriate therapy ⁽¹²⁾. The pulmonary hyperlucency is caused by overdistension of the alveoli in conjunction with diminished vascularity ⁽¹³⁾.

Computerised tomography (CT) is superior to chest radiography in the diagnosis and differential diagnosis of Swyer-James syndrome ⁽¹³⁾. The following radiological features can be seen: A hyperlucent but small lung with over expansion of the contralateral lung, and with a diffuse pattern of scarring. On CT chest, the bronchi will have a pruned appearance and a mosaic pattern of air trapping in acinii and air trapping during expiration ⁽¹⁴⁾. It is important to exclude a central lesion in cases of unilateral hyperlucent lung. Pulmonary function tests, chest CT-scan, ventilation and perfusion scintigraphy and if necessary bronchoscopy help in the differential diagnosis ⁽¹⁵⁾. Most patients are managed clinically. Management of the syndrome is typically conservative with prevention and early treatment of pulmonary disease and occasionally surgical treatment as well as influenza and pneumococcal vaccination. Patients should avoid inhaling injurious substances, avoid smoking and avoid hobbies and occupations where pulmonary barotraumas are possible.

On review of the literature, a number of cases have been reported from Japan. Most of the cases are

of young adults. A 26 year old asymptomatic man was diagnosed with Swyer James syndrome (SJS) when a screening Chest X-Ray showed increased radiolucency of the left lung and overinflation of the right lung ⁽¹⁶⁾. A case of a 20 year old Japanese man with a pneumothorax was reported ⁽¹⁷⁾. After improvement of the pneumothorax, chest x-ray revealed hyperlucency of the right upper lung. Chest CT revealed cystic spaces in the right upper lobe. The pneumothorax was probably caused by weakness of the cystic spaces. An 18 year old Japanese lady was admitted to hospital with a history recurrent spontaneous pneumothorax ⁽¹⁸⁾. CT confirmed SJS and emphysematous changes in the bronchioles. A case of a 17 year old patient in Taipei with the typical findings of SJS on chest x-ray was reported ⁽¹⁹⁾. He was admitted with cystic bronchiectasis complicated by lung abscess.

There have only been a few cases reported in the older population like our patient. A 65 year old female patient in Japan was admitted with dyspnoea on effort. CT Chest confirmed SJS ⁽²⁰⁾. A male patient aged 70 was being followed for a long time with bronchiectasis-he was finally diagnosed with SJS by radiological studies ⁽²¹⁾. Like our patient he also had a long history of chronic sinusitis.

SJMS should be considered as a spectrum disease. Bronchiectasis is not a universal finding,

The presence and type of bronchiectasis will influence clinical manifestations and prognosis, Studies have emphasised the importance of taking a careful history, the application of computed tomography and scintigraphy in confirming the diagnosis of SJMS and in eliminating other diseases).

This case is of both clinical and radiological interest. It highlights the importance of taking a good history and appropriate use of imaging like computed tomography. Infective exacerbations should be treated early and health promotion measures like smoking cessation advice and vaccination can be offered in the primary care setting.

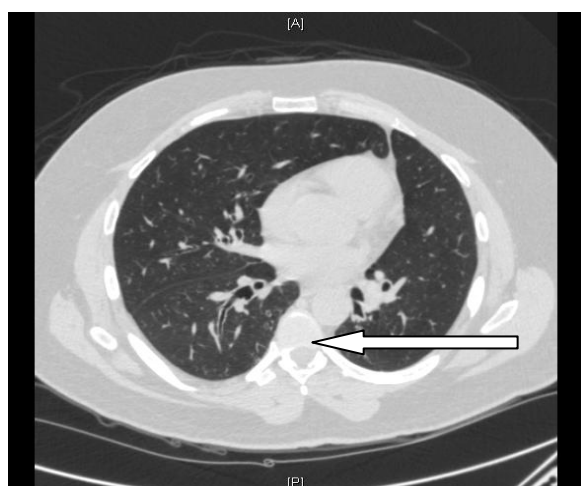


Figure 1: CT Thorax shows a pruned appearance of the bronchi and air trapping in the acinii.

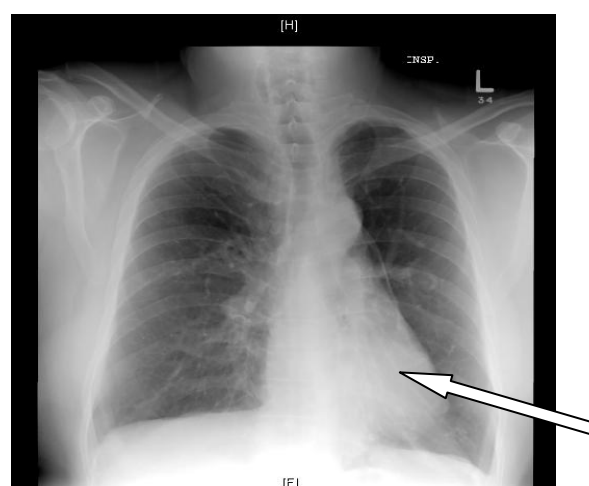


Figure 2: The Chest X-ray shows a hyperlucent but small left lung and diffuse scarring and peribronchial thickening of the left lower base.

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The corresponding author is:

Dr Patricia McWalter, Dept. of Family Medicine,
Consultant Family Physician, King Faisal Specialist
Hospital and Research Centre,
MBC 62, P.O box 3354, Riyadh 11211, Kingdom of
Saudi Arabia. Email-pmcwalter@doctors.org.uk