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An elderly lady with Swyer-James-Macleod Syndrome; a rare finding in adult pulmonology

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Introduction

Swyer-James-Macleod Syndrome (SJMS) is considered as a rare occurrence in adult respiratory medicine, and it is characterised by unilateral alveolar hyperdistention, reduction in pulmonary vasculature (both leading to hyper-lucent affected lung field) with or without associated bronchiectasis (1,2). It is considered as a postinfectious consequence of childhood bronchiolitis obliterans. The resultant effect of childhood infection leads to hypoplasia and/or agenesis of pulmonary vasculature resulting the hypoperfusion of pulmonary parenchyma, which is in turn more liable to acquire infections and develop bronchiectasis (3-5). The prevalence SJMS is mentioned as 0.01% and occurs mainly in children (4-5). The entity was first described in by an English pulmonologist, William Mathieson Macleod 1954, and by a physician, Paul Robert Swyer and a radiologist, George James in Canada in 1953 (6). Characteristically, the disease is highlighted as a unilateral disease though there are some case reports of bilateral disease in paediatric patients (7). The diagnosis of SJMS is mainly radiological and computed tomography and perfusion scintigraphy can direct to the diagnosis (4-5). We present an elderly patient with bronchiectasis who was found to have SJMS.

Case presentation

A 74-year-old lady without significant comorbidity presented to the outpatient clinic with episodes of recurrent chest infection over few months with a sputum culture which had grown *Pseudomonas aeruginosa.* She had no clubbing and systemic examination was insignificant. Chest auscultation revealed scattered crepitations on left lung-field. She did not have features suggestive of immunodeficiency, chronic obstructive pulmonary disease, connective tissue diseases, inflammatory bowel diseases or significant occupational and industrial exposures, especially for hydrocarbons. She could recall that she has had frequent events of early childhood chest infections, including a few hospital admissions though she could not recall fine details and there were no records available. The results of basic blood investigations, autoimmune screening, fungal screening, and screening for immunosuppression were all essentially normal.

The chest X-ray showed left sided hyperlucent lung fields with reduced pulmonary vascular markings. At a glance, it looks like more vascular marking on the right. However, what is the real abnormality is that she has got less pulmonary vascular markings on the affected left side. (Figure 1). The high-resolution computed tomography (HRCT) shows left sided bronchiectasis, reduced pulmonary vasculature and alveolar hyperdistention (Figure 2). The vascular 3D reconstruction clarifies the reduced left sided pulmonary vasculature (Figure 2). The echocardiogram did not suggest any abnormality including pulmonary hypertension. The CT excluded other common differential diagnosis of unilateral hyperlucent

lung such as asymmetrical emphysema, unilateral pneumothorax, asymmetrical constrictive bronchiolitis and foreign body inhalation. The clinical and radiological diagnosis of SJMS was made and standard bronchiectasis care was implemented along with anti-pseudomonal treatment. The chest clearance technique targeting the left lung was taught by the pulmonary physiotherapist. Current *Pseudomonas* infection/colonization was treated with Intravenous Ceftazidime 2 g thrice daily for 14 days with an intention to eradicate *Pseudomonas* and azithromycin (250 mg 3 times a week) were planned if any further colonization with *Pseudomonas aeruginosa* in the follow up sputum cultures.

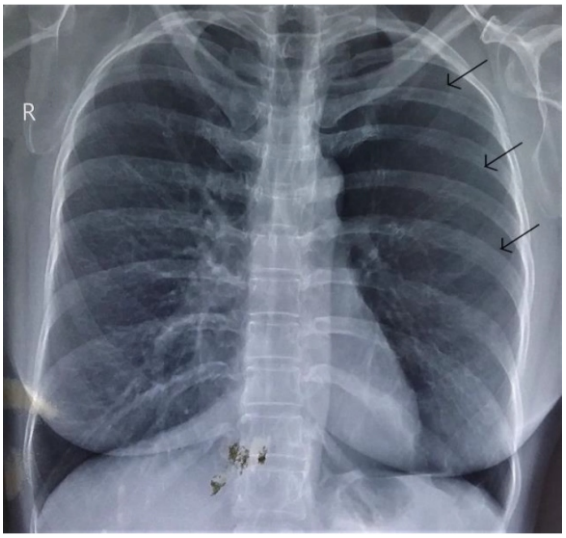

Figure 1: Chest X-ray showing the hyper-lucent left lung due to the reduced ipsilateral vasculature (arrows)

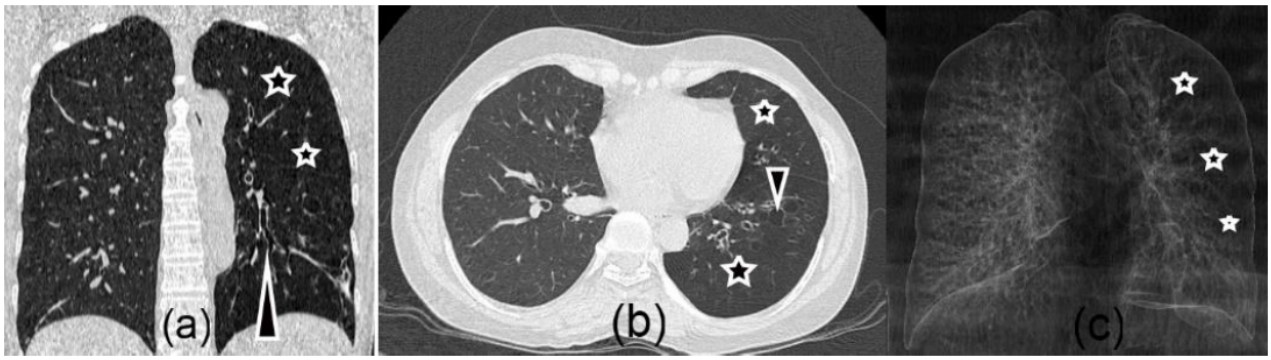

Figure 2: High Resolution CT images in sagittal (a), axial (b) views and HRCT-vascular reconstruction (c) Illustrating the left lung bronchiectasis (arrow head), reduced pulmonary vasculature (asterix)

Discussion

Swyer-James-Macleod Syndrome is a rare finding in adult respiratory medicine. It is usually asymptomatic and can even be under diagnosed (1). The presentation of patients with SJMS can be due to the complications or consequences of it like bronchiectasis and pulmonary hypertension. Therefore, they can present with recurrent chest infections, chronic cough, chronic *Pseudomonas aeruginosa* colonisation, haemoptysis and shortness of breath (1-3). The currently explained aetiology for SJMS is childhood viral bronchiolitis or viral pneumonia (1). Adenovirus, paramyxovirus, measles, Mycoplasma pneumonia and *Bordetella pertusis* has been described in the literature as infective aetiologies, whereas exposure to hydrocarbons is considered as a non-infective aetiology for this entity (1,4). The resultant inflammation of the peripheral airway leads impeded lung development along with the under development of pulmonary vasculature (3).

The radiological findings of SJMS are characteristic because the under-developed lung, it appears as a hyperlucent hemithorax on the chest X-ray. The reduced pulmonary vasculature and distended alveolar structure give rise to the ipsilateral hyper-lucency (4-5). The unilateral reduction of vasculature on CT scan or perfusion scan confirms the diagnosis in most of the patients.

The treatment for the condition is largely conservative with strategies for prevention of recurrent chest infections. However, there are patients described in the literature, who had undergone pneumonectomy, lobectomy and segmentectomy due to the complications of SJMS (1).

Conclusions

This rare disease highlights the importance of vigilant evaluation of chest X-ray which can lead to a diagnosis of underlying uncommon causes. Therefore, even though SJMS is a rare entity, simple chest X-ray can make a significant difference in diagnosis and the management of the patient.

Informed written consent has been obtained from the patient to publish this case report.

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