ORIGINAL RESEARCH ARTICLE

A Rare Case of Swyer James Macleod Syndrome: Adult Diagnosis

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A B S T R A C T

Introduction: Swyer James Macleod syndrome is an extremely rare condition that presents with unilateral hyperlucent hemithorax. This condition is associated with post infectious bronchiolitis obliterans, usually during the childhood. There is obliteration of the bronchi and vasculature, resulting in hypoplastic pulmonary artery and impaired lung development.
Case report: Here we report a case of a 47-year-old Indian male who presented to the pulmonary medicine department with cough and breathlessness diagnosed as Swyer James Macleod syndrome during workup of unilateral hyperlucent lung.
Conclusion: Although, this syndrome is usually a childhood diagnosis, the patient can have an asymptomatic childhood leading to a delayed presentation as an adult where the syndrome is discovered as an incidental finding.

Keywords: Swyer James Macleod Syndrome, Adult Diagnosis

INTRODUCTION

Swyer James Macleod syndrome (SJMS) is an extremely rare condition that presents with unilateral hyperlucency of one or more lobes or the entire lung that occurs due to loss of pulmonary vasculature and alveolar distension.[1] The common presenting lung findings are unilateral emphysema, bronchiectasis and/or bronchiolitis obliterans. Although SJMS is of unknown etiology repeated episodes of viral bronchiolitis or viral pneumonias are suspected to be the cause.[2]

CASE PRESENTATION

A 47-year-old Indian male patient presented to pulmonary medicine department with primary complaints of cough associated with mucoid sputum and breathlessness that were on and off for the past 10 years with increased severity since the last 10 days. There is no history of diabetes, hypertension and he is not a smoker, alcoholic. There is no significant past medical history and family history. On physical examination there is decreased breath sounds on the left side. Blood pressure was approximately 120/70 mmHg, pulse of 78/min, respiratory rate of 22/min and saturation was 98% at normal room conditions. Rest of the physical examination was unremarkable. His laboratory investigations are Hb of 11 mg/dl, WBC count of 7500, platelet count of ~ 3.5 lakhs. The patient's liver function tests, kidney function tests, and serum electrolytes were all within the normal limits.

Chest radiograph (Figure 1) showed a hyperlucent left hemithorax with decreased bronchovascular and hilar markings. right lung appears normal. there is no evidence of consolidatory changes, pleural effusion on both sides.

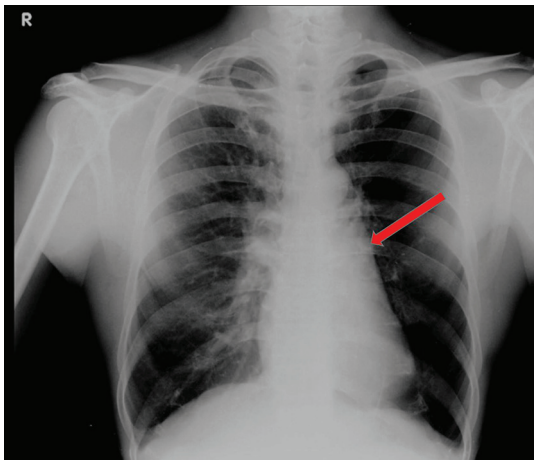

Figure-1: Hyperlucent left hemithorax with decreased bronchovascular and hilar markings. Right lung appears to be normal. Hypoplastic appearing left hilum (Red Arrow)

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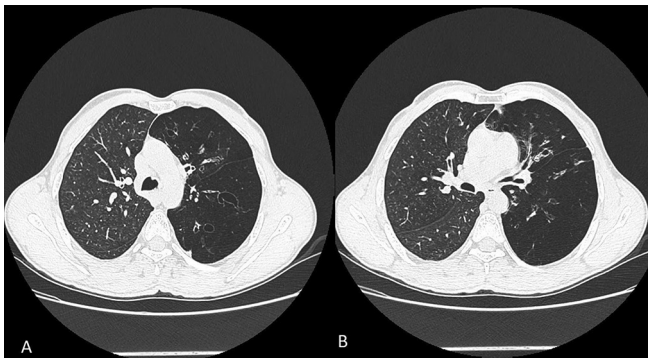

Figure-2: Diffuse cystic and tubular bronchiectasis involving all the segments in both upper (A) and lower (B) lobes. There is decreased attenuation of entire left hemithorax, and the left pulmonary artery appeared to be grossly reduced in caliber.

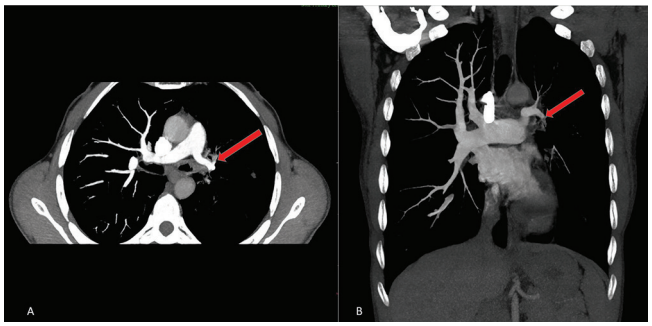

Figure-3: Maximum intensity projection (MIP) Axial section (A) and Coronal section (B) of pulmonary angiogram showing markedly hypoplastic left pulmonary artery (Red Arrows). There is no evidence of pulmonary thromboembolism.

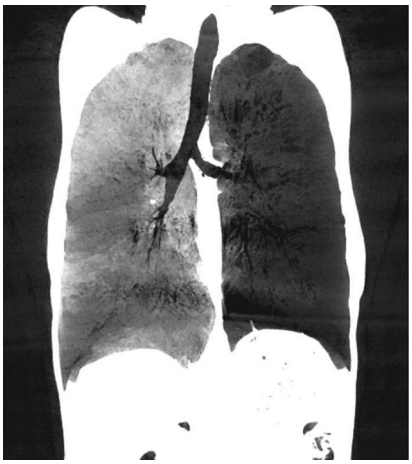

Figure-4: Coronal section of minimum intensity projection (MINIMIP) image showing unilateral hyperlucency of left sided lung field.

Computed tomography scan (Figure 2) revealed diffuse cystic and tubular bronchiectasis involving all the segments in both upper and lower lobes of left lung field. There is decreased attenuation of entire left hemithorax, and the left pulmonary artery appeared to be grossly reduced in caliber. Pulmonary angiogram (Figure 3) showed markedly diminished size of the left pulmonary artery with no evidence of pulmonary thromboembolism.

DISCUSSION

Swyer [physician] and James [radiologist] were first to report this entity in the year 1953 in a 6-year-old boy and later in 1954 the same condition was reported by MacLeod [chest physician] in an adult. Although there have been huge controversies over why the condition develops, it has been widely agreed that the disease is associated with childhood infections most commonly post-infectious bronchiolitis obliterans and pneumonitis.[3] There are less than 20 cases reported since then in Indian literature.

The pathophysiology of Swyer James Macleod syndrome is viral pneumonitis due to adenovirus, influenza or non-viral lower respiratory tract infection like mycoplasma, tuberculosis which causes bronchitis and bronchiolitis of involved lung followed by obliteration of bronchi leading to fibrosis of interalveolar septa. These changes will lead to decreased ventilation and chronic hypoxia causing hypoplastic pulmonary arteries.[4] A radiological diagnosis is made by unilateral hyperlucency of lung on X-ray chest, unilateral reduction in vascularity on chest CT and unilateral loss of perfusion on CT pulmonary angiography. Although the disease can be diagnosed on chest radiographs, CT scans are far more valuable as they provide information regarding emphysematous changes, bronchiectasis, bronchiolectasis and mosaic attenuation due to air trapping. Pulmonary angiography which is not routinely done shows decreased caliber of pulmonary artery and its branches causing hypoperfusion of lungs.[5]

SJMS should be differentiated from other conditions causing unilateral hyper lucent lungs like unilateral absence of pectoralis muscle, congenital unilateral pulmonary artery and ipsilateral lung hypoplasia, congenital lobal emphysema, and left lower lobe collapse. A detailed history, along with attempts to locate and visualize any prior imaging, is essential for an accurate diagnosis.[6]

Treatment of SJMS is usually conservative with chest physiotherapy, immunizations, bronchodilators and managing of respiratory infections. However, for patients presenting with bronchiectasis along with Swyer-James syndrome, leading to recurrent and severe lung infections, pneumonectomy is necessary to provide the patient with an improved quality of life and a prolonged life expectancy.[1]

CONCLUSION

Although Swyer-James-Macleod syndrome (SJMS) is a rare etiology of an unilateral hyperlucent hemithorax, it is an important one that should be considered in any individual with such findings. Inaccurate diagnosis can lead to incorrect management. Typically, this condition is a childhood diagnosis where the child presents with recurrent respiratory infections. However, a few patients with an indolent course of the disease can present as adults either as an incidental finding or due to delayed presentation of symptoms.

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