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Swyer James Macleod Syndrome -A rare cause of hemoptysis in 31 year old man

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ABSTRACT

Swyer James Macleod syndrome (SJMS) is a seldom reported manifestation of post infectious obliterative bronchiolitis in childhood. This cause obliteration of peripheral airways and vascularity of the infected lung and impede lung development. We report SJMS in 31 year old male who presented with complaints of exertional dyspnea, productive cough and hemoptysis and history of recurrent childhood respiratory infections. He had hypoplasia of right lung and right pulmonary vasculature with bronchiectasis as per CT pulmonary angiography while ventilation perfusion scan showed matched V/Q defect diagnosed as SJMS.

Keywords: Pulmonary artery hypoplasia, Swyer James Macleod syndrome, matched V/Q defect.

1. INTRODUCTION

A case of unilateral translucent lung with diminished vascular markings in a child was first reported by Swyer and James in 1953 subsequently in 1954 Macleod reported similar findings in a series of nine patients (Swyer, 1953; Macleod, 1954). SJMS is due to of post infectious obliterative bronchiolitis in childhood with incidence of 0.01% (Michelson, 1977). Infections due to adenovirus, paramyxovirus etc are commonly reported with SJMS (Cumming et al., 1971; Stokes et al., 1978). Adult manifestations include dyspnea, cough, hemoptysis, bronchiectasis and recurrent pneumonia (Sen et al., 2014). Imaging includes unilateral hyper lucent lung, diffusely decreased ventilation, and matching decreased perfusion due to hypoplastic pulmonary artery (Chuang et al., 2010). We report SJMS in a young male with dyspnea, hemoptysis and similar imaging.

2. CASE REPORT

A young male, 31 years old came to respiratory medicine OPD with exertional dyspnea (grade 2 MMRC), productive cough and mild hemoptysis on and off since 8 years presented to us with increase in symptoms for last 10 days. His past history suggestive of repeated respiratory infections and had prior hospitalization in 2019 with complaint of massive hemoptysis which was managed in intensive care by blood transfusion and discharged. Routine

blood investigations and vitals were normal. Chest X- ray (fig.1) showed right mid zone bronchiectatic changes with compensatory hyperinflation of left lung. Previous CT chest showed diminished right side vasculature. Hence CTPA (fig. 2) done which showed hypoplasia of right pulmonary vasculature and right pulmonary vein, emphysematous changes in right lung along with bronchiectasis with reduced lung volume and hyperinflation of left lung. His Ventilation/ Perfusion scan (fig. 3) showed matched V/Q defects. Spirometry showed mixed pattern. The clinical presentation and imaging confirmed Swyer James Macleod syndrome. His two dimensional echocardiography was normal. He had obstructive sleep apnea confirmed by polysomnography with Apnea Hypoapnea Index of 50; he was managed with i.v. antibiotics, nebulized bronchodilators, chest physiotherapy and continuous positive airway pressure therapy. Patient improved symptomatically and was discharged after 7 days. He is presently on inhaler therapy, completed his Flu, Pneumococcal and COVID-19 vaccination and on regular follow up.

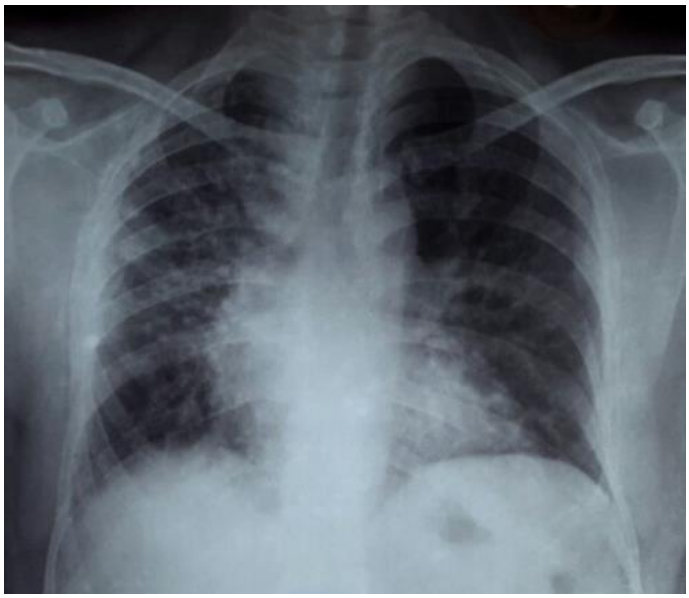

Figure 1 Chest Xray- Right mid zone bronchiectatic changes with compensatory hyperinflation of left lung.

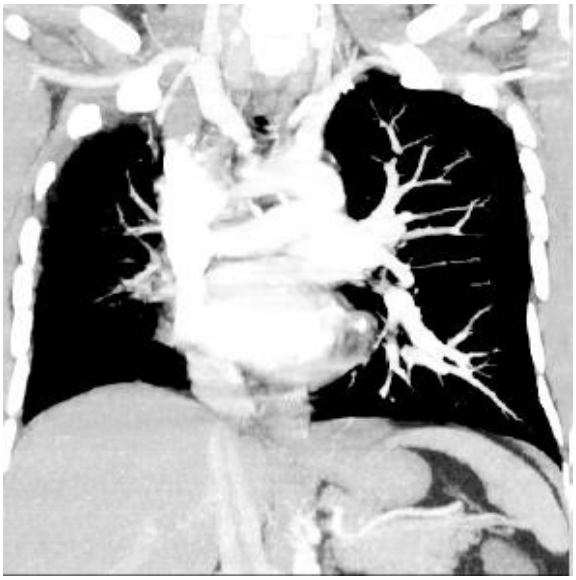

Figure 2 CTPA-Hypoplasia of right Pulmonary artery and vein.

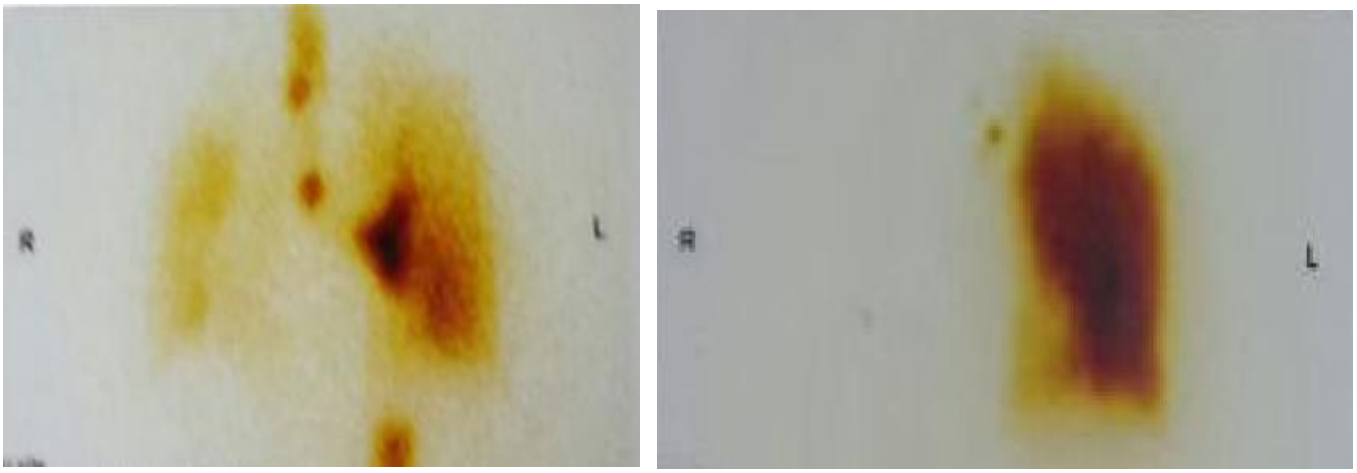

Figure 3 Ventilation/Perfusion scan showed matched V/Q Defect.

3. DISCUSSION

SJMS is seldom reported in literature (Michelson, 1977), it usually arises after childhood viral bronchiolitis and pneumonitis in childhood. It is also due to post Adenovirus, Paramyxovirus, Mycoplasma pneumonia, Mycobacterium tuberculosis infections etc. (Cumming et al., 1971; Stokes et al., 1978). Manifestations include dyspnea, hemoptysis, cough, wheezing, recurrent bronchitis and bronchiectasis usually occur in adult with the recurrent childhood infections (Sen et al., 2014) as in our case who had dyspnea and hemoptysis on off since 8 years with history of recurrent childhood respiratory infections. On auscultation basal crackles heard on right side. His chest X-ray showed right mid zone bronchiectatic changes with compensatory emphysematous on left side and his CTPA showed hypoplasia of right pulmonary vasculature,and empysema with bronchiectasis and reduced lung volume in right lung while there was compensatory hyperinflation of left lung as similar as the cases reported in the study (Sen et al., 2014). His Ventilation/ Perfusion scan showed matched V/Q defects with air trapping similar to the cases in previous studies (Chaung et al., 2010; Zeiger et al., 1977).

The infectious process leads to inflammatory response which causes obliteration of pulmonary vasculature as well as peripheral airways leading to impeded lung development (Chaucer et al., 2016). SJMS diagnosis needs either of the following: unilateral lung volume loss with hyperlucency as seen in chest X ray or unilateral reduced lung vascularity on CT chest, or unilateral reduction in lung perfusion scan. In our patient right lung is affected although most of the case series predominantly reported left lung involvement (De Moura et al., 2017). The SJMS patients frequently suffer from recurrent lung infections (Chaucer et al., 2016). SJMS with pulmonary hypertension is recently reported; but in our case two dimensional echocardiography showed normal pulmonary artery pressure (Yuce et al., 2016).

Our patient had concominant OSA as similar to a case reported (Machado et al., 2019). Conservative management is the treatment of choice with effective antibiotics and chest physiotherapy while surgery is the option for patient with recurrent infections or when the entire lung is affected (Chaucer et al., 2016). Our patient preferred medical management and on regular follow up. It is been observed that SJMS with coexistence of bronchiectasis as negative impact in prognosis (Machado et al., 2019).

4. CONCLUSION

A proper history and radiology can help in clinching the diagnosis. Dyspnea is being a common respiratory symptom which on further evaluation led to SJMS confirmation this case. Although surgical management gives the curative option, conservative management alleviates the symptoms and reduces the disease burden. This report is to revisit SJMS, its clinical, radiological presentations and management.

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Informed Consent

Written & Oral informed consent was obtained from the patient included in the study.

Author Contributions

MJ and MY – Concept and writing of the manuscript.
AR and KL - Data collection

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Conflict of interests

The authors declare that there are no conflicts of interests.

Data and materials availability

All data associated with this study are present in the paper.

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