A Rare Association Of Swyer-James-Macleod Syndrome With COPD And Bronchiectasis: A Case Report

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***Abstract*—Swyer-James-MacLeod syndrome (SJMS) is rare and occasionally diagnosed at a late age. The disease is characterized by a lack of pulmonary arterial flow in one lung, and the unilateral lung appears hyperlucent in imaging. The disease is usually diagnosed in childhood, after an infection or other respiratory complication. We have an adult man with chronic respiratory symptoms suggestive of chronic obstructive lung disease. However, on CAT scanning, it was also confirmed that Swyer-James-MacLeod syndrome and bronchiectasis.**

Keywords—Swyer-James-MacLeod syndrome, COPD, Bronchiectasis, Hyperlucent lung, Hypoplasia of the pulmonary artery.

INTRODUCTION

Commonly known as an acquired childhood disease, Swyer-James-MacLeod syndrome is characterized by agenesis or hypoplasia of the pulmonary artery and hypoperfusion of the unilateral lung secondary to recurrent infective bronchiolitis [1]. The recurrent childhood respiratory infections lead to post-infectious bronchiolitis obliterans and vascular wall inflammation, leading to localized bronchial narrowing, reduced ventilation, vasoconstriction, chronic hypoxia and pulmonary hypoplasia [2-4]. Patients are usually symptomatic in childhood, and rarely do they remain asymptomatic until a later age. At later ages, they are incidentally diagnosed with other chronic respiratory diseases, as in our case, who presented with COPD symptoms and was diagnosed as well. The prevalence of SJMS is around 0.01% based on a retrospective survey of 17,450 chest radiographs [5].

CASE PRESENTATION

A 56-year man, a chronic smoker (30 pack years), presented with increased cough, shortness of breath and blood-tinged sputum for one month; however, he has had a chronic mild cough, dyspnea and wheezing for a long time. On examination, his chest revealed bilateral expiratory wheezing and right basal crackles. His routine blood tests were remarkably normal except for raised white cell counts ($12.03x10^3$/mcL). The chest x-ray suggested right lower zone bronchiectatic changes with superadded infection. It also revealed a hyperlucent right lung compared to the left lung (Fig. 1).

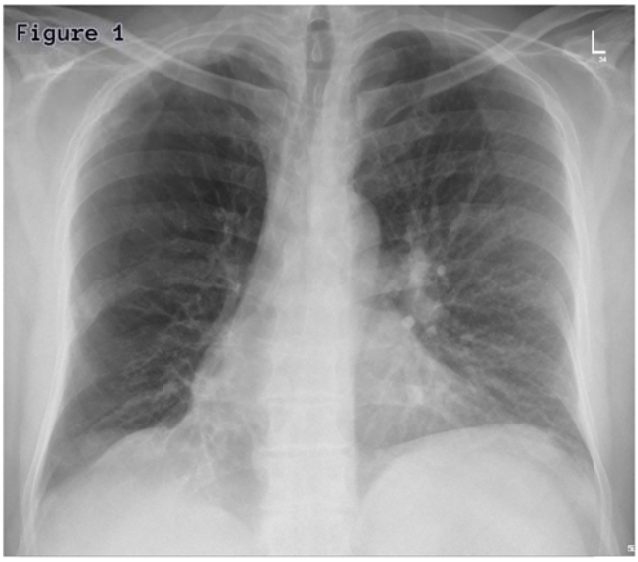

Fig. (1): Chest X-ray PA view showing relatively hyperlucent right lung, decreased vascular markings in right lung and tiny cystic-like areas in the right lower zone.

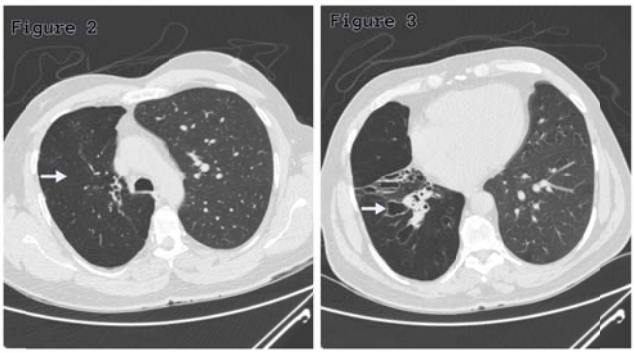

Fig. (2): CT section of the chest showing Hyperlucent right lung and attenuated vessels.
Fig. (3): CT section of the chest showing bronchiectasis and mucus plugging.

The CECT chest showed severe tubular bronchiectasis involving the right lower lobe, with the near total destruction of the anterior basal segment. Evidence of active infection is seen in the form of mucus plugs occluding most of the bronchi of the right lower lobe, with some showing air-fluid levels (Fig. 2, 3).

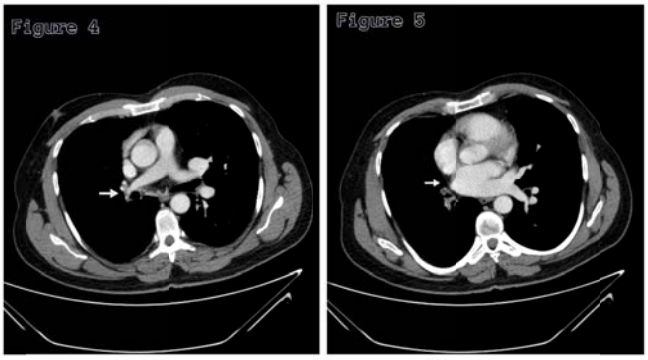

Fig. (4): CT section of the mediastinal window showing complete attenuation of branches of right pulmonary artery.
Fig. (5): CT section of mediastinal window showing complete attenuation of right pulmonary vein.

The CECT chest also revealed pan acinar emphysema involving the right lung, with volume loss of the right hemithorax and absence of the right pulmonary artery and pulmonary vein branches (Fig. 4, 5). The patient underwent bronchoscopy, which showed abnormal bronchial mucosal thickening in the right lower lobe; however, no active bleeding source has been identified. The lavage from the right lower lobe grew the pan-sensitive Hemophilus Influenza. The biopsy from abnormal bronchial mucosa was suggestive of chronic bronchitis.

He was treated with appropriate antibiotics, and on follow-up, his spirometry showed a moderate obstructive pattern without reversibility, suggestive of chronic obstructive pulmonary disease (COPD). He was started on appropriate treatment for COPD and Bronchiectasis.

DISCUSSION

Swyer-James-MacLeod syndrome is an uncommon encounter in respiratory medicine and, in most cases, diagnosed at an early age. This syndrome was discovered first time in a 6-year-old child by Swyer and James in 1953 [6]. The reported prevalence of SJM syndrome is 0.01% [7]. The recurrent childhood respiratory infections such as adenovirus, respiratory syncytial virus, influenza A, measles, tuberculosis, Mycoplasma pneumoniae, and Bordetella pertussis cause post-infective bronchiolitis obliterans which ultimately leads to the development of Swyer-James-MacLeod syndrome [8, 9]. The clinical presentation of SJMS is very non-specific, and it may remain asymptomatic for many years or present with a life-threatening acute respiratory failure [10]. Our patient presented with chronic productive cough with dyspnea and wheezing, consistent with COPD symptoms; however, in some case reports, SJMS was misdiagnosed as COPD [11]. The common differential diagnoses are congenital lobar emphysema, pulmonary hypoplasia, bullous emphysema, and pulmonary embolism [10].

The diagnosis of SJMS can be confirmed by High-resolution CT (HRCT) chest; however, a chest x-ray can also give some clues in severe cases, such as unilateral small lung with hyperlucency and air trapping on expiration [12]. The CT pulmonary angiography can further elaborate the anatomy of pulmonary arteries and branches, which showed hypoplasia or the absence of one or both pulmonary arteries and narrowed attenuated arterial branches in the hyperlucent lung zone. The classic triad for diagnosis of SJMS is unilateral hyperlucent lung, diffusely decreased ventilation, and matching decreased perfusion in the affected lung [13]. In some cases, bronchiectasis may be present; however, this is not a universal finding of SJMS [14]. MR angiography (MRA) is usually not required for making a diagnosis of SJMS; however, if it is done, it may show a small pulmonary artery with diminished vascularity in the periphery [15].

The reported complications of SJMS are recurrent infections [16], lung abscess, pneumothorax [17-19] and rarely pulmonary hypertension [20].

The management of SJMS is conservative in preventing infection by annual influenza vaccination, pneumococcal vaccination and prompt antibiotic course in case of acute infection. In addition, airway clearance techniques can help avoid nidus infection in the bronchiectasis area. Surgery is not a commonly adopted treatment for SJMS; however, this can be an option in recurrent or refractory infections, especially in localized bronchiectasis. The prognosis of SJMS is good in the absence of bronchiectasis.

CONCLUSION

The SJMS is rare in adults and mostly misdiagnosed as COPD or other common respiratory diseases. The history of respiratory infection in childhood and CT chest findings are essential information for diagnosing SJMS. Our case presented with typical symptoms of COPD, and he was diagnosed with SJMS along with COPD and Bronchiectasis. The prognosis is good if the infection part is taken care of promptly.

ACKNOWLEDGMENT

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