Swyer-James Syndrome: An Infrequent Cause Of Bronchiectasis?

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Abstract

Swyer-James syndrome is perceived as an unusual, sporadic phenomenon but may occur more frequent than expected. It is usually identified by the presence of unilaterally increased radiolucency. A case of a 61 year-old white male who presented with dyspnea and revealed a history of a long-lasting pulmonary infection during early childhood is presented. The case is remarkable because classical radiographic features of Swyer-James syndrome were misdiagnosed as congenital pulmonary hypoplasia despite the apparent air trapping and bronchiectasis on radioimaging studies.

ABREVIATIONS

FEV_1 = Forced Expiratory Volume in the first second;

FVC = Forced vital capacity;

RV = Residual volume;

SJS = Swyer-James syndrome.

INTRODUCTION

Unilateral hyperlucent emphysema was first described by Swyer and James in a 6-year old child in 1953.[1] The following year, MacLeod reported nine patients with unilateral pulmonary hyperlucency.[2] This syndrome is characterized radiologically by a small or normal-sized hyperlucent lung with decreased vascularity and air trapping on expiration. The Swyer-James syndrome (SJS) is one of several causes of a unilateral hyperlucent lung. It is now considered that SJS is a consequence of an obliterative bronchiolitis.[3] After an initial pulmonary infection, these patients are prone to develop repeated respiratory infection and bronchiectasis in the involved lung.[4] The occurrence of bronchiectasis is frequent but not universal; nevertheless, the presence and type of bronchiectasis may influence clinical course and prognosis.[3]

The aim of our report is to describe the importance of ascertaining the concomitant finding of bronchiectasis and a hyperlucent, small-sized lung in diagnosing SJS. In addition, we discuss the clinical and radiological features, and the differential diagnosis of the two classical presentations of the SJS.

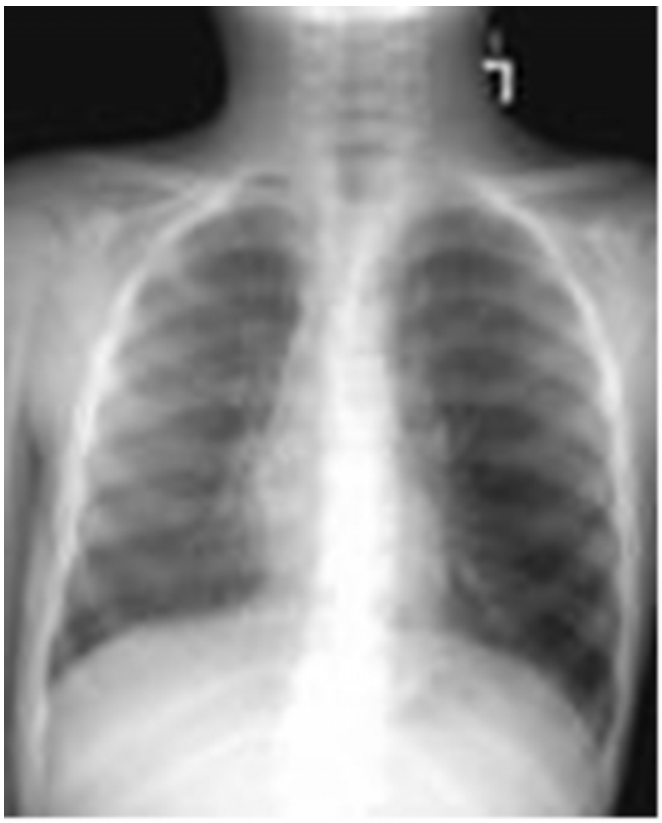
CASE REPORT

A sixty one year-old white male nonsmoker with diagnosis of papillary thyroid carcinoma with lymph node metastasis, status-post total thyroidectomy, was referred to our clinic due to shortness of breath and increased production of purulent sputum for the previous four weeks. His past medical history is significant for an episode of long-lasting pneumonia and frequent respiratory tract infections during early childhood, and at age 36 he was told to have underdevelopment of the right lung when he entered the military service. Since he was discharge from the army, patient has had dyspnea associated with wheezing and repeated respiratory tract infections. In addition, he has hypertension. His father died of emphysema and two sisters have thyroid goiters. Physical examination revealed a well develop man, tachypneic, afebrile, his respiratory rate was 36, blood pressure 146/95, pulse 80. Lung examination showed a thyroidectomy scar. The rest of the examination was unremarkable. Chest radiography revealed hyperlucency in the majority of the left lung with decreased vascularity, small left hilum, an enlarged right hilum, volume loss in the left lung with mediastinal shift to the left, and bronchiectatic changes in the left base. The CT of the chest revealed emphysematous changes with air trapping especially in the left lung with decreased in the vascular and interstitial marking throughout the left lung and some areas of cylindrical bronchiectasis. Pulmonary function studies revealed a decreased Forced Vital Capacity (FVC). The ratio

of forced expiratory volume in the first second (FEV_1) to FVC was decreased (77% predicted) indicating airway obstruction. There was improvement of these parameters following bronchodilators. Residual Volume (RV) was markedly increased (215% predicted), consistent with air trapping. The diffusing capacity was normal.

Figure 1

Figure 1. Chest X-ray PA an lateral views showing hyperlucent left lung suggesting unilateral emphysema.



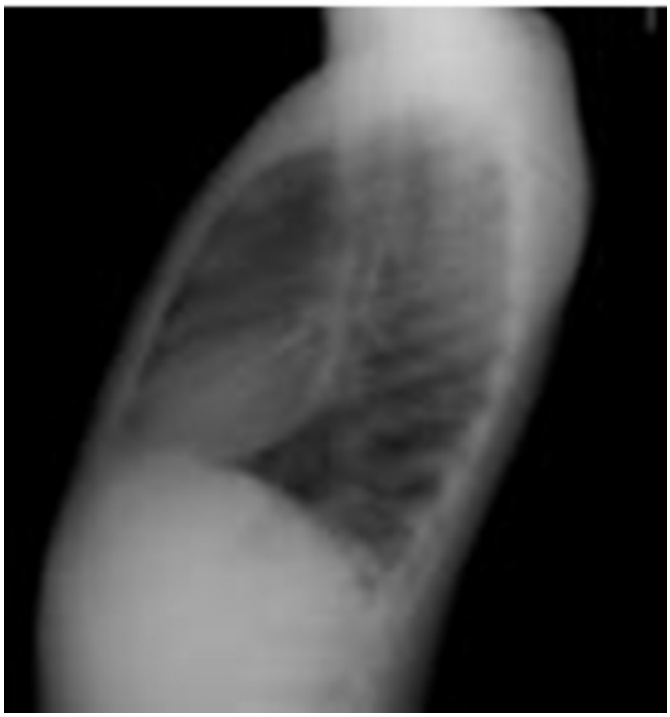
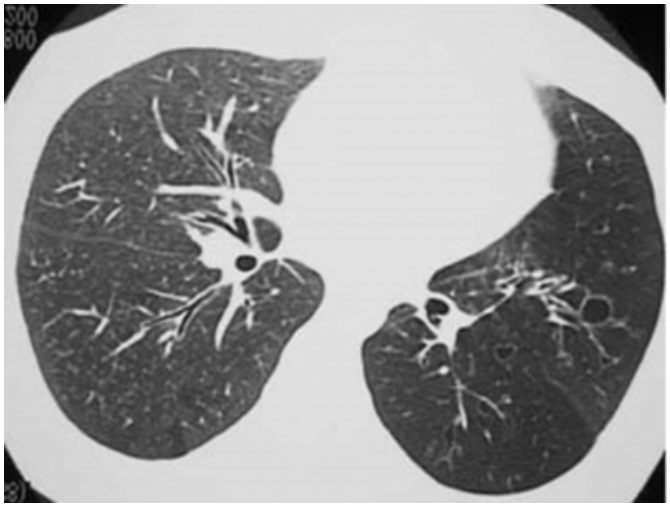


Figure 2

Figure 2. Axial Computed Tomography of the chest reveals a small hyperlucent left lung, atresic vascular structures and evidence of air trapping, characteristic of the Swyer-James syndrome.



DISCUSSION

The finding of the chronic infections and volume loss associated with hyperlucency of the left lung in this patient led to the belief that this clinical picture was similar to the one described by Swyer-James, and MacLeod.[1,2] The diagnosis of SJS is explained by the fact that this patient had pneumonia at an early age which lasted a long time creating a great deal of airway obstruction and chronic inflammation (bronchiolitis). In turn, this bronchoilitis may have led to air trapping and overdistention of the air spaces of the right lung with subsequent compression of the developing pulmonary vasculature impeding its development and ending in localized “emphysema” and bronchiectasis. Another mechanism leading to the hypoplastic vasculature of the affected lung is the ultimate fibrosis of the inter-alveolar septae; resulting in obliteration of the pulmonary capillary bed and reduction in pulmonary artery blood flow.[5]

Swyer-James syndrome is usually the consequence of constrictive bronchiolitis that implicate etiological agents, such as adenovirus, measles, respiratory syncytial virus, influenza virus, mycoplasmas, and tuberculosis.

As previously described is cases of SJS, the findings in our patient of recurrent respiratory infections during childhood are found in as many as 60% of the cases.[6] However, few of the patients could be asymptomatic and the diagnosis may be delayed.[3] Moreover, cough, dyspnea, occasional hemoptysis, and clinical manifestations of bronchiectasis also may occur.[4]

The diagnosis is usually made on the basis of moderate hyperlucency of an entire lung or a lobe preferably in the CT of the chest. The hyperlucency results from air trapping and decreased pulmonary vascular marking in the involved area.[3,4] Pulmonary function tests reveal decreased to normal FVC and decreased FEV1/FVC ratio indicating airway obstruction.

Swyer-James syndrome has two classical presentations: as isolated hyperlucent lobe or lung and a hyperlucent lobe with collapsed ipsilateral lobe as in our case. In the latter classical presentation of SJS, which combines a hyperlucent lobe with as ipsilateral paucity containing bronchiectasis (exemplifying 2/3 of these cases), the main differential diagnosis is severe bronchiectasis.[3]

In the differential diagnosis, a number of conditions can create a roentgenographic appearance similar to hyperlucency of the lung, ie.: pulmonary embolism, congenital pulmonary hypoplasia, pulmonary artery hypoplasia, bronchial atresia, and Williams Campbell syndrome.[4,6] However, the closest resemblance would be congenital pulmonary hypoplasia, which presents with only slight unilateral radiolucency; no air trapping, and diminutive normal bronchial tree without bronchiectasis.[4]

In terms of the differential diagnosis for bronchiectasis, we may think of congenital or acquired bronchiectasis, ciliary dyskinesia, cystic fibrosis, allergic broncho-pulmonary aspergillosis, and immunoglobulin deficiencies. In this respect, we suggest that the clinician should consider SJS when evaluating patients with bronchiectasis. Furthermore, the serious condition of foreign body-related air trapping can create roentgenographic signs that are similar to those in SJS, except for the ipsilateral increased lung volume. Therefore, in any patient presenting with an isolated hyperlucent lobe or lung, the presence of a lesion within the ipsilateral main bronchus must be excluded before a diagnosis of Swyer-James syndrome is accepted.[7]

The presence and type of bronchiectasis have been reported to influence the clinical course and prognosis in patients with SJS.[3] Our case represents the cylindrical type of bronchiectasis which has been reported to progress toward repeated, mild respiratory infections but a spontaneous tendency toward improvement without harboring further bronchiectatic changes, in contrast to those patients with saccular or cystic bronchiectasis.[3] These findings may explain the slow progression over time of the bronchial abnormalities seen in our patient.

CONCLUSION

In summary, we report a case of a patient who has demonstrated the classical history and radiological findings of the Swyer-James syndrome, which may be encountered in medical practice more frequent than expected when it is considered within the differential diagnosis of bronchiectasis.

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