Clinical Case Reports: Open Access

Yumed Text
OPEN ACCESS JOURNALS

Case Report | Vol 5 Iss 2
ISSN: 2582-5038
https://dx.doi.org/10.46527/2582-5038.210

Incidental Diagnosis of Swyer-James-Macleod Syndrome: Case Report

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Received: March 19, 2022; **Accepted:** April 04, 2022; **Published:** April 11, 2022

1. Introduction

Swyer-James-Macleod syndrome (SJMS) is a rare constrictive bronchiolitis often diagnose incidentally on thoracic imaging and is thought to arise from childhood pulmonary infections, resulting in air-flow obstruction and vascular distortion. The syndrome is characterized by the pathological radiological finding of unilateral hyperlucency. In this case report, we present a case of SJMS found incidentally after the patient underwent imaging to evaluate their COVID positive status.

2. Case Report

A 29-year-old male without significant past medical history presented to our emergency department with right lower quadrant pain of one day duration. On physical examination, he was vitally stable and only had tenderness and guarding in the right lower quadrant of his abdomen. His labs were significant for a WBC count of 13.27 × 10*9/L with a high neutrophil count of 9.74 × 10*9/L and a CRP of 58.1 mg/L. A computerized tomography (CT) scan of the abdomen with IV and oral contrast was done and was suggestive of acute appendicitis. As per hospital protocol, a nasopharyngeal COVID-19 swab was taken prior to undergoing laparoscopic appendectomy, for which he was found to be positive. He was completely asymptomatic for COVID-19, however, a chest x-ray was done and revealed an incidental finding of decreased lung markings over the left lower chest likely attributed to hyperinflation of a portion of the left lower lobe, for which Radiology recommended a CT chest for further evaluation. CT angiogram showed an incidental left lower lobe parenchymal destruction and bronchiectasis, multifocal narrowing of the left lower lobe bronchus, paucity of the left lower lobe segmental and subsegmental pulmonary vessels with compensatory emphysema of the left upper lobe. The radiological findings were consistent with a diagnosis of Swyer-James-

Macleod syndrome. His case was later discussed in the advanced lung disease multidisciplinary team meeting, and it was concluded that since the patient was completely asymptomatic from a pulmonary aspect, no surgical intervention was required (FIG.1-3).

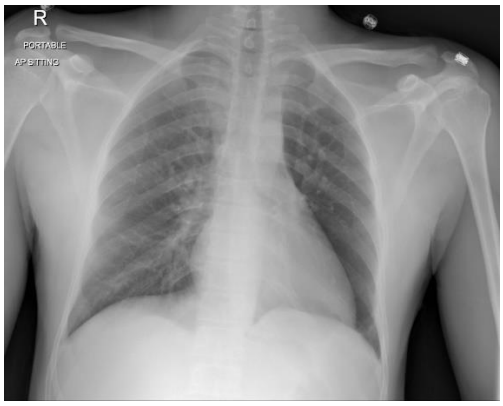

FIG. 1. **Chest x-ray revealing paucity of lung markings over the left lower chest is likely due to hyperinflation of a portion of the left lower lobe.**

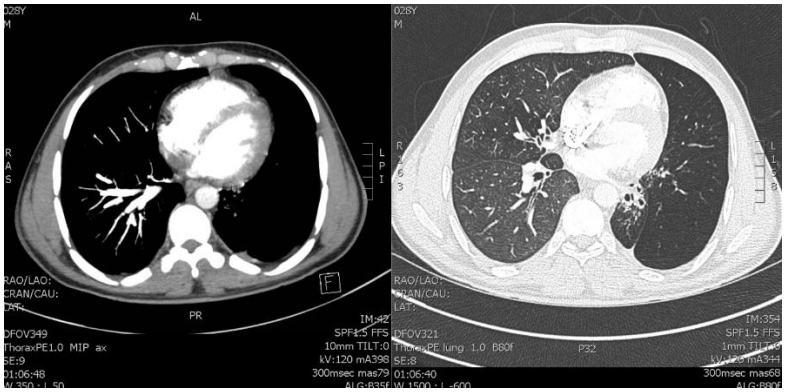

FIG. 2. **Chest CT (mediastinal windows) revealing parenchymal destruction and bronchiectasis; associated multifocal narrowing of the left lower lobe bronchus and paucity of left lower lobe segmental and subsegmental pulmonary vessels.**

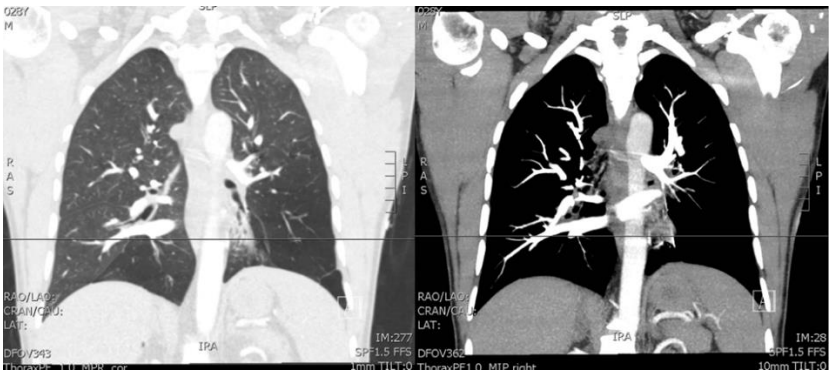

FIG. 3. **Chest CT (lung windows) revealing parenchymal destruction and bronchiectasis; associated multifocal narrowing of the left lower lobe bronchus and paucity of left lower lobe segmental and subsegmental pulmonary vessels.**

3. Discussion

Swyer-James-McLeod syndrome (SJMS) is a rare pulmonary disease and is characterized by hypoplasia of pulmonary arteries to a lung lobe or segment. This in turn causes the lungs to appear hyperlucent on imaging. SJMS was first described in 1953 and the literature review to-date only consists of a handful of case reports, therefore, true incidence/prevalence of this syndrome is not known; although, in one study of 17,450 chest radiographs, investigators reported a prevalence of 0.01% [1]. The disease has predilection to involve the left lung more frequently which is also depicted in our case [2].

The aetiology of SJMS has often been linked with postinfectious bronchiolitis obliterans and pneumonitis during childhood. Infections caused by viruses including the *Paramyxovirus morbillivirus*, influenza A and adenovirus types 3, 7 and 21 as well as the bacterial infections such as *Bordetella pertussis*, *Mycobacterium tuberculosis* and *Mycoplasma pneumoniae* have been implicated in pathogenesis of SJMS [3].

The pathogenesis involves recurrent or severe respiratory infections in childhood, which affect the terminal bronchi and bronchioles, cause pulmonary damage, and prevent the normal development of alveolar ducts, leading to submucosal fibrosis and complete obliteration of small airways. The obstruction of airways results in air trapping, which in turn causes results in emphysema and/or bronchiectasis [4]. The fibrosis of the anterior alveolar septae due to same infectious process causes obstruction of pulmonary capillary beds and hence, leads to reduced blood flow to pulmonary arteries. Overtime, the pathology results in pulmonary arterial development to be hindered during early childhood, causing the lung to appear hyperlucent on radiographs later in life [5].

As most patients with SJMS are asymptomatic, the diagnosis is usually made incidentally. In our case the patient was admitted for acute appendicitis but had no other acute or chronic respiratory symptoms. In a minority of cases, the presentation is manifested by recurrent respiratory infections, exertional dyspnoea, productive cough, wheeze or pleuritic chest pain [1].

SJMS can be diagnosed on routine chest x-ray, which demonstrates a hyperlucent segment or lobe of the lung and reduction in bronchovascular markings [6]. Computed tomography scan of the chest (CT scan) helps ascertain the extent of the disease and complications such bronchiectasis, and atelectasis as well as ruling out other differential diagnoses such as emphysema. Ideally, both inspiratory and expiratory scans should also be obtained to demonstrate air trapping [7]. CT pulmonary angiography in SJMS reveals the hypoplasia of the pulmonary arteries in the affected lobe or segment, however it is not necessary for the diagnosis [3]. A technetium Tc 99 m lung scan or a ventilation/perfusion scan (V/Q scan) can also be used as diagnostic tools as they can show localized perfusion loss in 1 lung. Some authors have suggested that a V/Q scan can be used as first-line test after a chest x-ray [8]. Pulmonary function tests are non-diagnostic and can be normal or mildly restrictive in asymptomatic or obstructive in patients with significant bronchiectasis and air trapping [9,10].

Since most patients with SJMS are asymptomatic, a conservative approach to management is usually all that is needed. For symptomatic patient's the medical management is similar to that with patients with bronchiectasis with focus on airway clearance and reduction of exacerbation frequency. Hence, the treatment modalities include mucolytics, chest physiotherapy, pulmonary rehabilitation, and inhaled bronchodilators [3]. Surgical pneumonectomy, a lobectomy or segmentectomy are all possible treatments, reserved for patients with persistent/recurrent respiratory tract infections, refractory to medical

management [11,12]. Bronchiectasis is the commonest complication of SJMS can lead to significant morbidity. Other significant complications include formation of bullae and increased risk of pneumothorax [10].

Unsurprisingly, the life expectancy in asymptomatic patients with SJMS is likely to be normal and main cause of morbidity is due to development of bronchiectasis and obstructive airways disease [13].

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