A unilateral hyperlucent lung - Swyer-James syndrome: A case report and literature review

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**Abstract**
Swyer-James-Macleod syndrome (SJMS) is a rare etiology of a unilateral hyperlucent hemithorax but an important one, which should be considered in any individual with such findings. Presentation usually occurs in adulthood with an asymptomatic history in many cases or with a history of childhood infections. Clinically, symptomatic patients may present with productive cough, dyspnea on exertion, hemoptysis, decreased exercise tolerance and recurrent pulmonary infections. Many individuals are asymptomatic only requiring conservative management. However, some patients may require surgical intervention. SJMS can be easily misdiagnosed and must be suspected for example, in any patient diagnosed with asthma who does not respond to therapy. We report a case of a 51-year-old African American male diagnosed with SJMS on the basis of his medical history, clinical presentation and x-rays and computed tomography chest scans findings.

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1. Introduction
Swyer-James-Macleod syndrome (SJMS), first reported in 1953 and 1954, is a rare cause of a unilateral hyperlucent hemithorax. It is a relatively uncommon entity, occurring in 3.8% of patients with bronchiolitis obliterans (BO) in one study and in 4.3% of BO cases in another [1]. Most patients are clinically asymptomatic. However, some patients can present with repeated pulmonary infections, dyspnea, or hemoptysis [1]. There are various proposed mechanisms for the presence of bronchiectasis in SJMS including BO leading to atelectasis or scarring which results in bronchial dilatation, bronchiectasis itself being the primary inciting event with distal spread to peripheral small airways leading to obliteration, or the initial viral infection may damage the bronchioles and bronchi simultaneously [1]. The characteristic radiographic findings include a unilateral hyperlucent lung along with decreased bronchovascular markings, a small hilar shadow and slight displacement of the mediastinum to the affected side [2].

2. Case presentation
A 51 year-old African American male patient presented with complaints of cough with scanty mucoid sputum production and exertional shortness of breath. His symptoms had started after a severe respiratory tract infection at the age of 9 years and he was admitted several times with complaints of cough and sputum production during adolescence and childhood and was treated with antibiotics and bronchodilators. The patient has no other significant past medical history, and his family history was noncontributory. He denied tobacco, alcohol and recreational drug use. On examination, he was not in acute distress, with a blood pressure of 117/75 mmHg, a pulse rate of 62/min, a respiratory rate of 16/min and he was saturating 98% on room air. Pulmonary examination was significant for decreased breath sounds at the left lung base. The remainder of his physical examination was normal.

Laboratory findings showed a white cell count of 6200/mm³, a hematocrit of 43% and a platelet count of 334,000/mm³. His serum electrolytes, renal and liver function tests were within normal ranges. The chest radiograph disclosed a hyperlucent left lung with numerous cystic regions of bronchiectasis (Fig. 1).

Computed tomography (CT) scans demonstrated diffuse cystic bronchiectasis involving the left lung with a small caliber left pulmonary artery; findings consistent with Swyer-James-Macleod syndrome (Fig. 2).

There was a mild obstruction on pulmonary function test (PFT), and he tested negative for Alpha-1 Antitrypsin deficiency.

3. Discussion
SJMS also known as unilateral hyperlucent lung syndrome which was first described in 1953 and 1954 is a rare entity
characterized by hyperlucency of one lung, lobe or part of a lobe. There has been much debate regarding the exact etiology of this process however it is associated with childhood infections most commonly post-infectious bronchiolitis obliterans and pneumonitis. It is also associated with various viruses such as Paramyxovirus morbillivirus, Bordetella pertussis, Mycobacterium tuberculosis, Mycoplasma pneumoniae, influenza A and adenovirus types 3, 7 and 21 [3]. Regardless of this association, it is relatively uncommon occurring in 3.8% of patients with bronchiolitis obliterans in one study and 4.3% in another [1]. Most patients have a unilateral involvement of the left lung primarily. In one study, the left lung was involved in all of its patients [1]. Only the left lung in our patient was affected. In most cases, both the small bronchi and bronchioles are affected, and the pulmonary damage that occurs during childhood prevents normal development of the alveolar ducts. Airways develop submucosal fibrosis leading to luminal irregularity and occlusion. Pulmonary vasculature is hypoplastic while the lungs distal to diseased bronchioles become hyperinflated with pan-acinar emphysematous changes in some cases [4].

Presentation usually occurs in adulthood with an asymptomatic history in many cases or with a history of childhood infections. Clinically symptomatic patients may present with productive cough, dyspnea on exertion, hemoptysis, decreased exercise tolerance and recurrent pulmonary infections. In one study, dyspnea on exertion was the most frequent presenting complaint compared to a prior study in which shortness of breath was not a prominent feature [1].

Diagnosis is made radiographically by x-ray and CT scan and is an incidental finding in some cases. Radiographically, the hyperlucency is usually confined to one lobe or lung, even though bronchography and radionuclide scans show that the abnormality is often patchy and bilateral [5]. The characteristic radiographic findings include a unilateral hyperlucent lung along with decreased broncho-vascular markings, a small hilar shadow and slight displacement of the mediastinum to the affected side [2]. In one retrospective study which examined eight patients, radiographs revealed unilateral disease in seven of their cases and bilateral disease in one, however, both radionuclide and CT scans showed that radiographs underestimated the extent of disease [5]. In this same study, CT was more sensitive than radiography as well as scintigraphy, in detecting hyperlucent lung regions and was also superior in assessing the extent and distribution of the disease [5]. On CT scans, air trapping manifests as persisting low attenuation of lung tissue in affected regions at end exhalation [5]. CT scan findings may also include bronchiectasis, bronchiolectasis, atelectasis and scarring [2]. Bronchiectasis, though not a universal finding, evident in only 30% of patients, affects the clinical manifestations and prognosis of the disease [2]. Individuals with SJMS who have saccular bronchiectasis usually have more severe exacerbations than those who do not have bronchiectasis on high resolution CT scan [2]. Pulmonary angiography is not an essential criteria for the diagnosis of the entity although it can reveal hypoplasia and diminished size of the affected pulmonary artery. Pulmonary angiography however has its limitations, as it cannot discern congenital from acquired etiologies of hypoplastic pulmonary vasculature.

Fig. 1. Chest radiograph showing a hyperlucent left lung with cystic bronchiectasis.

Fig. 2. CT scan axial and coronal views showing diffuse cystic bronchiectasis involving the left lung, with areas of honey-combing.
Treatment is usually individualized ranging from conservative management to surgical intervention. Conservative symptomatic management is the mainstay of treatment for patients diagnosed with SJMS utilizing chest physiotherapy, low-dose inhaled corticosteroids, and inhaled bronchodilators [6]. Patients should also be given pneumococcal and Influenza vaccinations. Long-term oxygen therapy may be appropriate in cases with advanced disease and respiratory failure [2]. Surgical intervention should be considered for patients who have repeated infections and are not responding to optimal medical management. The most common surgical procedure was a pneumectomy and some patients were treated with lobectomy or segmentectomy [6]. Prognosis is dependent on the presence or absence of bronchiectasis [2].

There are several important differential diagnoses that should be considered when evaluating any individual with unilateral pulmonary hyperlucency including pneumothorax, asymmetric emphysema, congenital lobar emphysema, and pulmonary artery hypoplasia [2]. Other differential diagnoses include gastrointestinal herniation, bronchial compression, mastectomy and mediastinal fibrosis [7]. Poland syndrome is another cause of a unilateral hyperlucent hemithorax, which is due to congenital unilateral absence of the pectoralis major and minor muscles, hypoplasia of the breast and nipple and scarcity of subcutaneous tissue [8]. SJMS can be easily misdiagnosed and must be suspected in any patient diagnosed with asthma who does not respond to therapy. One case highlighted this, as their patient was initially misdiagnosed as having asthma [9]. Another case presented an individual with SJMS, who was initially misdiagnosed as having a pneumothorax for which he received multiple chest tube insertions [10].

4. Conclusion

It is important to highlight the various diagnoses that one must be mindful of, when approaching any patient who presents with unilateral lung hyperlucency. Inaccurate diagnoses can lead to inappropriate therapy.

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Conflicts of interest
None.

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