Swyer-James-Macleod syndrome as a rare cause of unilateral hyperlucent lung

Three case reports

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Abstract

Rationale: Swyer-James-Macleod syndrome (SJMS) is a rare lung disorder characterized by unilateral hyperlucent lungs which arises as a complication of bronchiolitis obliterans. It is typically diagnosed during childhood, although some patients may only be diagnosed in adulthood, often as an incidental finding, but others due to recurrent chest infections.

Patient concerns: Three patients were referred to our institution with complaints of dyspnea on exertion, chronic productive cough, and recurrent pulmonary infections. Two of them had a history of lower respiratory tract infections during childhood.

Diagnosis: A computed tomography scan was performed and showed unilateral reduced density of the lung and bronchiectasis in the 3 patients. Based on the clinical presentation and radiologic features, the diagnosis of SJMS was established.

Interventions: Patients started inhaled corticosteroids and long acting beta agonist, vaccines to prevent respiratory infections were administered and airway clearance techniques were taught.

Outcomes: Two patients became asymptomatic with inhaled therapy and no recurrent chest infections were observed over a 3-year follow-up period, being discharged from our institution to the general practitioner. The 3rd patient had some improvement in the frequency of pulmonary infections and dyspnea, without improvement in respiratory function tests, maintaining vigilance at our center.

Lessons: These cases highlight the importance of being aware of this condition and its frequent association with bronchiectasis, which may adversely affect the prognosis, to manage patients appropriately and prevent recurrent pulmonary infections.

Abbreviations: BMI = body mass index, CT = computed tomography, FEV1 = forced expiratory volume in 1 second, FVC = forced vital capacity, RFTs = respiratory function tests, SJMS = Swyer-James-Macleod syndrome.

Keywords: bronchiectasis, hyperlucent lung, Swyer-James syndrome

1. Introduction

Swyer-James-Macleod syndrome (SJMS) is a rare lung disorder characterized by unilateral hyperlucent lungs. The syndrome occurs due to a vascular and parenchymal development impairment in an area affected by bronchiolitis obliterans in childhood, resulting in hypoplastic vascular regions and emphysematous pulmonary areas. It is typically diagnosed during childhood, although some patients may only be diagnosed in adulthood, often as an incidental finding.[1,2] The clinical, imaging, and prognostic features of 3 patients diagnosed with SJMS in adulthood are presented.

2. Case reports

2.1. Case 1

A 53-year-old Caucasian male was referred with complaints of dyspnea on exertion and productive cough. He was an overweight (body mass index [BMI] of 27kg/m²), former smoker with no significant lower respiratory tract infection in childhood. On physical examination diminished breath sounds and widespread wheeze were noted. The chest X-ray showed hyperlucency of the left lung (Fig. 1) and the computed tomography (CT) scan exhibited cystic bronchiectasis on the left lower lobe and a reduced density in the same area. His echocardiogram was normal and respiratory function tests (RFTs) revealed a reversible moderate obstructive pattern, with pre- and postbronchodilator values of forced expiratory volume in 1 second (FEV1) and forced vital capacity (FVC) of 69%/83% and 77%/95%, respectively. Based on clinical presentation and radiologic features a diagnosis of SJMS was established. The patient started inhaled corticosteroids and long acting beta agonist, and vaccines to prevent pulmonary infections were administered. He became asymptomatic with inhaled therapy, normalization of RFTs was observed and no recurrent chest infection occurred during a 3-year follow-up period, time after which he was discharged from our center to the general practitioner.

2.2. Case 2

A 41-year-old Caucasian male was referred with complaints of dyspnea on exertion and productive cough. He was an overweight (BMI of 27kg/m²), former smoker with no significant lower respiratory tract infection in childhood. On physical examination diminished breath sounds and widespread wheeze were noted. The chest X-ray showed hyperlucency of the left lung (Fig. 1) and the computed tomography (CT) scan exhibited cystic bronchiectasis on the left lower lobe and a reduced density in the same area. His echocardiogram was normal and respiratory function tests (RFTs) revealed a reversible moderate obstructive pattern, with pre- and postbronchodilator values of forced expiratory volume in 1 second (FEV1) and forced vital capacity (FVC) of 69%/83% and 77%/95%, respectively. Based on clinical presentation and radiologic features a diagnosis of SJMS was established. The patient started inhaled corticosteroids and long acting beta agonist, and vaccines to prevent pulmonary infections were administered. He became asymptomatic with inhaled therapy, normalization of RFTs was observed and no recurrent chest infection occurred during a 3-year follow-up period, time after which he was discharged from our center to the general practitioner.

2.3. Case 3

A 45-year-old Caucasian male was referred with complaints of dyspnea on exertion and productive cough. He was an overweight (BMI of 27kg/m²), former smoker with no significant lower respiratory tract infection in childhood. On physical examination diminished breath sounds and widespread wheeze were noted. The chest X-ray showed hyperlucency of the left lung (Fig. 1) and the computed tomography (CT) scan exhibited cystic bronchiectasis on the left lower lobe and a reduced density in the same area. His echocardiogram was normal and respiratory function tests (RFTs) revealed a reversible moderate obstructive pattern, with pre- and postbronchodilator values of forced expiratory volume in 1 second (FEV1) and forced vital capacity (FVC) of 69%/83% and 77%/95%, respectively. Based on clinical presentation and radiologic features a diagnosis of SJMS was established. The patient started inhaled corticosteroids and long acting beta agonist, and vaccines to prevent pulmonary infections were administered. He became asymptomatic with inhaled therapy, normalization of RFTs was observed and no recurrent chest infection occurred during a 3-year follow-up period, time after which he was discharged from our center to the general practitioner.
emergency department with a respiratory infection. He was obese (BMI of 31 kg/m²), nonsmoker, with a history of left pneumonia in childhood. Even without infection, the patient complained of effort dyspnea and crackles were noted at the lower left hemithorax on pulmonary auscultation. The chest X-ray revealed hyperlucency of the left lung field and the CT scan showed a decrease in density of the left lung with a decrease in density more pronounced on the left lower lobe, a small left pulmonary artery, cylindrical bronchiectasis and a mosaic perfusion defect in the right lung (Fig. 2). Echocardiogram was unremarkable and RFTs demonstrated a reversible mild to moderate airflow obstruction (pre- and postbronchodilator values of FEV1 and FVC were 61%/69% and 88%/96%, respectively). Due to the clinical presentation and investigations a diagnosis of SJMS was established. The patient initiated inhaled corticosteroids and long acting beta agonist, and the importance of regular vaccination to prevent respiratory infections and airway clearance was emphasized. He had symptomatic and lung function improvement with no recurrent chest infections during 3 years, being discharged from our institution.

2.3. Case 3
A 70-year-old Caucasian non smoker female, with obesity (35 kg/m²), obstructive sleep apnea treated with continuous positive airway pressure and a recently diagnosed asthma, was referred due to recurrent pulmonary infections requiring hospital admission. She had a history of lower respiratory tract infections during childhood and complaints of dyspnea on exertion and chronic cough. Crackles were audible on the entire left hemithorax. The chest X-ray showed hyperlucency of the left lung and CT scan demonstrated volume loss in the left lung with a decrease in density more pronounced on the left lower lobe, a small left pulmonary artery, cylindrical bronchiectasis and a mosaic perfusion defect in the right lung (Fig. 3). CT angiogram was negative for pulmonary embolism and she performed a bronchoscopy that was unremarkable. The echocardiogram showed indirect signs of pulmonary hypertension and RFTs revealed a nonreversible moderately severe mixed airflow pattern (FEV1 of 58% and FVC of 63%). Based on the above clinical presentation and investigations a diagnosis of SJMS was made. The patient maintained previously prescribed inhaled corticosteroids and long acting beta agonist, inhalation technique was revised, vaccines to prevent pulmonary infections were administered and airway clearance techniques were taught. She had some improvement in the frequency of pulmonary infections and dyspnea, stable RFTs and no hospital admission in the last 2 years, maintaining vigilance at our center.

3. Discussion
The SJMS is a rare lung condition. In a review of 17,450 chest radiographs, it was found in only 0.01% of cases. As in all our patients, SJMS is usually symptomatic for a prolonged period of time before detection. Symptoms include chronic cough, dyspnea on exertion, wheezing, hemoptysis, and chest pain. Most commonly there is a history of pulmonary infections in childhood, although this was not the case in one of our 3 patients. Recurrent lung infections and bronchiectasis are also present in a large number of patients, which may adversely affect prognosis. Physical examination may reveal decreased chest

Figure 1. Chest radiography of case 1 showing a hyperlucent left lung.

Figure 2. Computed tomography scan of case 2 showing low density areas with reduced vascularity (white arrows) and cystic bronchiectasis in the left lung (black arrow).
expansion and breath sounds, hyperresonance, crackles, or wheeze. However, symptoms and physical examination are nonspecific and may be present in many chronic lung diseases. Frequently patients are diagnosed as having asthma or chronic obstructive pulmonary disease due to their RFTs, which usually reveal a mild-to-moderate obstructive airflow pattern.

The differential diagnosis of SJMS include pneumothorax, congenital lobar emphysema, pneumatocele, bronchogenic cyst, bullous lung disease, pulmonary embolism, endobronchial foreign body, or postlobectomy compensatory emphysema, with radiologic studies being of extreme importance for their distinction.

The diagnosis is made through imaging examinations. SJMS may be identified by chest X-ray, showing decreased density in the lung field of the affected side with reduced bronchovascular markings, a smaller hilus, and reduced lung volume with slight displacement of the mediastinum to the affected side. CT scan is more sensitive in detecting these features and assessing the extent and distribution of the disease. Others findings can include bronchiectasis, pulmonary artery hypoplasia, oligemia, air trapping, and mosaic perfusion defect. Unilateral decreased density and reduced bronchovascular markings with bronchiectasis localized in that area were observed in all our patients. The last patient also presented with pulmonary artery hypoplasia and mosaic perfusion defect. Based on these findings and clinical presentation the diagnosis of SJMS was made. Interestingly the affected side is in most cases the left one, as in the 3 cases we described. In addition, pulmonary hypertension has also been reported to be associated with the syndrome.

Our 3 cases had bronchiectasis, and although not identified in all patients with SJMS, this feature affects the clinical manifestations and prognosis of the disease, with patients having more severe exacerbations and recurrent infections than those who do not have bronchiectasis.

Treatment of SJMS is usually conservative and involves chest physiotherapy, inhaled bronchodilators and low-dose inhaled corticosteroids, management and prevention of pulmonary infections. In selected cases, lung resection may improve lung function and quality of life, mostly in patients who suffer from severe and repeated respiratory infections associated with lung damage.

In the subgroup of patients with postinfectious bronchiectasis, those who have SJMS can have a different prognosis, due to the associated vascular changes. This syndrome is probably under recognized, but easily diagnosed based on the characteristic radiologic features, and should be considered early in the differential diagnosis of a unilateral hyperlucent lung. Physicians should be aware of this condition and its frequent association with bronchiectasis, to manage patients appropriately, prevent recurrent pulmonary infections, and achieve a better prognosis.

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