Swyer-James-MacLeod syndrome – report of two adult cases and review of the literature

Sindromul Swyer-James-MacLeod – raportarea a două cazuri la adult și revizuirea literaturii

Abstract

We describe two adult cases of Swyer-James-MacLeod syndrome (SJMS) localized in the upper right lobe, following clinically well-defined episodes of lower respiratory tract infections. SJMS is considered a sequela of obliterative bronchiolitis, frequently of a viral origin, resulting in the accidental discovery of a pulmonary hyperlucency on chest X-ray, usually localized in a single lobe. Chest CT and ventilation-perfusion scans are used to confirm the diagnosis. We advance the hypothesis that inadequate duration of therapy or lack of recognition of an episode of obliterative bronchiolitis, especially during childhood, may cause the development of SJMS.

Keywords: MacLeod syndrome, Swyer-James syndrome, obliterative bronchiolitis, unilateral lung hyperlucency, unilateral emphysema

Rezumat

Descriem două cazuri de sindrom Swyer-James-MacLeod (SJMS) la adult, localizate în lobul drept superior, secundare unor episoade de infecții ale tractului respirator inferior. SJMS este considerat o sechelă a bronșiolitei obliterative, frecvent de origine virală, care duce la descoperirea accidentală a hipertransparenței pulmonare la radiografia toracică, de obicei localizată într-un singur lob. Tomografia computerizată și scanările de ventilație-perfuzie sunt folosite pentru a confirma diagnosticul. Propunem ipoteza că durata inadecvată a terapiei sau nerecunoașterea unui episod de bronșiolită obliterantă, în special în timpul copilăriei, poate duce la apariția SJMS.

Cuvinte-cheie: sindrom MacLeod, sindrom Swyer-James, bronșiolită obliterantă, hipertransparență pulmonară unilaterală, emfizem unilateral

Introduction

In 1954, MacLeod described nine adult cases of hyperlucency of one lung, without collapse. In 1953, Swyer and James had already described a similar condition in a child. Swyer-James-MacLeod syndrome (SJMS) is also commonly known as “unilateral lung hyperlucency” or “unilateral emphysema”, involving a lobe or an entire lung. This condition, caused by localised bronchiolitis, is rare, with an estimated prevalence, according to the study of Gaensler et al., of 0.01% among 17,450 routine chest X-rays. In some cases, SJMS is diagnosed, or at least suspected, during childhood, when chest X-rays are obtained to investigate recurrent lower respiratory tract infections. In other cases, SJMS is incidentally found on screening chest X-rays. We report and describe two cases of SJMS.

Case 1

A 24-year-old male, originally from China, current smoker of 15 cigarettes per day for 6 years, was diagnosed with latent tuberculosis in 1988, and was treated with isoniazid 300 mg/day for 6 months. In 1989, he was treated for community-acquired pneumonia. In June 1993, he presented with fatigue. A chest X-ray demonstrated hypodensity in the right upper lobe, which was not visible on the chest X-ray from 1988 (Figure 1). A high-resolution chest CT scan (HRCT) showed near absence of lung parenchyma in the right upper lobe (Figure 2). The absence of functional parenchyma was confirmed by a ventilation-perfusion scan. Pulmonary function tests demonstrated a mid obstructive ventilatory defect (FEV1 = 83% predicted), with increased residual volume in body plethysmography (143%), while RV was...
Case 2

A 49-year-old man, originally from Senegal, presented with productive cough, following a bronchitic episode four months prior, which was treated with a macrolide. He reported since adolescence recurrent lower respiratory tract infections during the winter months. He has had a previous history of lower respiratory tract infections during the winter months. The patient was started on a long-term anti-muscarinic agent and oral N-acetylcysteine. 

Figure 2. Case 1. Chest CT-scan confirming the presence an area of complete loss of lung parenchyma in the right upper lobe

Discussion

The diagnosis of SJMS is based on history, pulmonary function tests, chest X-ray and chest CT scan. The clinical presentation of SJMS is variable. Some patients may be asymptomatic, and the syndrome may be incidentally suspected during investigations for lower respiratory tract infections. However, these are usually recurrent in patients with SJMS, starting from childhood. These episodes may present as viral bronchiolitis, often caused by adenovirus, measles virus, or respiratory syncytial virus. Other agents include Mycoplasma pneumoniae and Bordetella pertussis.

Patients with obstructive ventilatory defect may complain of exertional dyspnea. The difference in residual functional capacity measured by plethysmography and by helium dilution in patients with SJMS may be sizable. Diffusing lung capacity for carbon monoxide is usually reduced. Pulmonary function tests normally remain stable over time.

The two cases presented here had the typical radiographic presentation of unilateral loss of broncho-vascular markings, and both had a previous history of lower respiratory tract infections. SJMS always represents a localised sequela of an episode of bronchiolitis obliterans, often of viral etiology and usually occurring in childhood. Less frequently, the originating bronchiolitis can be caused by bronchial tuberculosis, exposure to toxic vapors or by the aspiration of foreign bodies. SJMS can be complicated by pulmonary abscesses or bronchopneumonia.

We advance the hypothesis that inadequate duration of therapy or lack of recognition of an episode of obliterative bronchiolitis, especially during childhood, may cause the development of SJMS. Swyer-James-MacLeod syndrome probably originates from an episode of acute bronchiolitis that causes obliteration of small airways, while distal parenchyma is collaterally ventilated through Kohn’s pores and Lambert’s canals, progressive hyperdistension and gas trapping occur, finally resulting in emphysematous destruction. The contribution from an infectious process is also possible, especially if this persists, in the presence of tobacco smoking, promoting the elastolytic process that results in emphysema. Histologically, the obliteration of bronchioles, with irregular distribution, dilation and panacinar destruction of pulmonary parenchyma are observed.

Chest X-ray shows unilateral hyperlucency. Bilateral involvement is rare. Chest angio-CT scan confirms the reduced number and caliber of vessels in the area involved. The differential diagnosis of unilateral hyperlucency/emphysema is actually limited to SJMS and, with a previous history of infection, the diagnosis is secured. High resolution chest CT scan with inspiratory and expiratory views can be helpful to confirm the typically lobar rarefaction of pulmonary parenchyma, and may show the presence of saccular or cylindrical bronchiectasis. These may be associated with SJMS. Although not typical for SJMS, bronchiectasis may impact the clinical course and the prognosis. As expected, patients with no bronchiectasis exhibit a lower incidence of bronchopneumonic episodes.

Some authors recommend the use of ventilation/perfusion scan to definitely rule out the pulmonary embolism. Bronchography is no longer used.

Regarding the treatment of SJMS, antibiotic and corticosteroid therapies are usually administered in patients with bronchiolitis obliterans, but the incidence of SJMS after such episodes and the duration of therapy necessary to prevent SJMS are not known. Influenza and
pneumococcal vaccinations are recommended when SJMS is established\(^ \text{23}\). In patients with recurrent, clinically significant pneumonias confined to the lobe involved by SJMS, surgical resection may be appropriate\(^ \text{23}\).

**Conclusions**

SJMS is often an incidental finding of localized hyperlucency and gas trapping confined to a lobe, in patients with previous bronchiolitis of infectious origin, especially viral. The radiographic finding of unilateral hyperlucency/emphysea is typical for SJMS, and the diagnosis is further supported by the past history of chest infections, probably poorly treated. The outcome is generally favourable, as long as the remaining lung parenchyma is disease-free, but recurrent lower respiratory tract infections will require close follow-up, targeted antibiotic treatment and, in severe cases, consideration for surgical resection of the affected lobe.

---

**References**