BALNEO RESEARCH JOURNAL
English Edition
eISSN 2069-7619 pISSN 2069-7597 ROMANIAN ASSOCIATION OF BALNEOLOGY

Challenges in multidisciplinary medical rehabilitation - Swyer-James-MacLeod Syndrome: case presentation and short literature review

ANTON Adelina[1], GHERGHISAN Ioana Sinziana[2], OMER (Gheorghe) Angela[1], IONESCU Elena Valentina[3,4], ILIESCU Madalina Gabriela[3,4], BAZ Radu[5]

Corresponding author: **Ioana Sinziana Gherghisan**, E-mail: ioana.husaru@gmail.com

Balneo Research Journal DOI: http://dx.doi.org/10.12680/balneo.2019.281 Vol.10, No.4, December 2019 p: 457–461

[1]Department of Pulmonology and Internal Medicine, County Clinical Hospital of Constanta, Romania
[2]Department of Pulmonology, Clinical Pulmonology Hospital of Constanta, Romania
[3]Department of Medical Rehabilitation, Faculty of Medicine, Ovidius University of Constanta, Romania
[4]Romania Balneal and Rehabilitation Sanatorium of Techirghiol, Romania
[5]Department of Radiology, Faculty of Medicine, Ovidius University of Constanta, Romania

Abstract

Introduction. Swyer-James-MacLeod syndrome (SJMS) or unilateral hyperlucent lung syndrome is a rare disorder caused by infectious bronchiolitis obliterans and pneumonitis occurring in childhood. It is characterized by hypoplasia and/or agenesis of the pulmonary arteries resulting in pulmonary parenchyma hypoperfusion. **Materials and methods.** We report the case of a 27 years-old female patient who presented with progressive dyspnea, productive cough, fever and chills. **Results and discussion.** Chest radiography showed unilateral loss of left lung volume with hyperlucency. Unilateral reduction in vascularity with reduced caliber of the left pulmonary artery was revealed on CT scan of the chest, final diagnosis of SJMS being confirmed by angiography. **Conclusions.** This case strongly supports the recommendation of considering SJMS within the differential diagnosis workup of bronchiectasis, the syndrome being usually underdiagnosed.

Key words: *pulmonary, rehabilitation, multidisciplinary,*

Introduction

Swyer-James-MacLeod syndrome is a rare entity characterized by hypoplasia and/or agenesis of the pulmonary arteries resulting in pulmonary parenchyma hypoperfusion. It is considered an acquired disease secondary to viral bronchiolitis and pneumonitis during childhood and is etiologically associated with viruses, atypical germs or Mycobacterium tuberculosis (1-4). Its incidence was 0.01% in an X-ray study conducted on 17450 patients (5,6). SJMS was firstly described in 1953 by Swyer and James, by documenting the case of a child with unilateral hypertransparent lung without atelectasis (7). One year later, MacLeod published nine similar cases (8).The aim of the paper is to describe the clinical and paraclinical features of one patient with Swyer-James-MacLeod syndrome and to do a short literature review on the topic.

Material and methods

A twenty-seven year old female patient, Caucasian, nonsmoker, presented to the Emergency Department of our hospital for a ten-day history of progressive dyspnea, productive cough, fever (39^0C) and chills. Empiric antibiotic therapy was prescribed with progressive worsening of her symptoms. For a comprehensive examination, the patient was hospitalized in the Internal Medicine Department, stating no significant family medical history. She disclosed recurrent episodes of pulmonary infections during childhood and adolescence, which were treated with antibiotics and bronchodilators.

Results

The physical examination was unremarkable, except lung auscultation, which revealed crackles on the left side. The complete blood count showed leukocytosis (14720 cells/µl). The patient tested negative for alpha1-antitrypsin deficiency and sweat chloride test, whereas serum immunoglobulins levels were within normal limits. Sputum cultures were positive for Citrobacter spp. Pulmonary function tests suggested relevant irreversible moderate obstructive disease: post-bronchodilator values of forced expiratory volume in one second (FEV_1) forced vital capacity (FVC) and FEV_1/FVC ratio were: 44%, 63% and 60% respectively. The chest radiography showed small left lung and apical lucency area in the left lung. Thoracic computed

tomography (CT) scans revealed a small, hyperlucent left lung with cystic bronchiectasis, compensatory hyperinflation of the right lung and reduced caliber of the left pulmonary artery (figure 1). The CT angiography confirmed diffuse hypoplasia of the left pulmonary artery (figure 2). Patient management included bronchodilators, targeted antibiotics and expectorants. Seven days later, due to symptomatic improvement under current treatment, the patient was discharged with recommendations of regular follow-ups and influenza and pneumococcal vaccinations.

Discussions

Swyer-James-MacLeod syndrome is a rare lung disease, also known as "unilateral translucent lung" or "unilateral emphysema". The hallmark of the syndrome is pulmonary arteries' hypoplasia and/or agenesis, resulting in pulmonary parenchyma hypo-perfusion, with characteristic radiological pattern - translucent or hyperlucent unilateral lung (6).

SJMS is an acquired illness secondary to childhood infectious bronchiolitis and pneumonitis, which are important clues, when found during anamnesis. Possible etiology may include viruses (Paramyxovirus, Morbillivirus, Influenza A, Adenovirus types 3, 7 and 21), atypical germs (Bordetella pertussis, Mycoplasma pneumoniae) and Mycobacterium tuberculosis (MTB) (5, 9). Infection with MTB is still very common in our country, overlapping with other rare conditions (10) and resulting in tuberculosis active disease in immunosuppressive conditions (11-14). Repeated infection causes an inflammatory reaction with consecutive obliteration of the peripheral airways and of the vessels, affecting the development of the organ. The affected lung becomes smaller than the healthy one, destruction of the alveolar walls leading to bronchiectasis and emphysema (5, 9). Unlike the majority of respiratory diseases, in which tobacco use or environmental exposures represent the most important risk factors (15-19), SJMS is not usually associated with smoking. Only the clinical assessment of smokers is not enough, but adding a biological evaluation will give the great picture of the problem (20).

Patients with SJMS are either asymptomatic, or, more frequently, they present non-specific respiratory symptoms (hemoptysis, dyspnea, chest pain, chronic cough, wheezing) which are commonly present in many other respiratory conditions (18). SJMS may also associate recurrent respiratory infections. Physical examination is nonspecific: decreased chest expansion, wheezing, bronchial rales, crackles, hyper resonance (9, 21).

Respiratory function tests (RFTs) usually reveal a mild to moderate obstructive pattern: decreased FEV_1/FVC ratio, decreased FEV_1, bronchial hyper responsiveness, decreased DLCO, severe air-trapping, increased RV/TLC ratio (residual volume/total lung capacity), and normal/slightly decreased lung volume. Frequently, these patients are misdiagnosed with chronic obstructive diseases such as asthma or chronic obstructive pulmonary disease (COPD) (21).

Diagnosis is mostly based on imaging studies. Radiographic changes may appear in a few months to a few years after the etiological infection and include: unilateral, unique hypertransparent lung, decreased pulmonary markings, small hilum, and mediastinum shift to the affected side. Furthermore, the involved lung fails in growing and exhibits minimal volume changes during breathing (9). CT-scans are mandatory for a complete positive diagnosis, revealing characteristic findings in the involved lung: small size, decreased lung attenuation, hyperlucency, bronchiectasis (in 30% of patients) and a mosaic pattern of air trapping (9, 21). The presence of bronchiectasis reveals severe exacerbations with poorer prognosis. Characteristic changes can also be found during nuclear medicine imaging procedures, such as decreased perfusion and decreased gas exchange during ventilation in the affected lung as compared to the healthy one (6, 9). Angiography is a helpful tool, showing small and/or hypoplastic pulmonary artery and branches in the involved lung (9).

Bronchoscopy is mainly recommended for differential diagnosis purposes, the endoscopic appearance being similar to chronic bronchitis. Bronchoalveolar lavage (BAL) shows inflammatory alveolitis, with increased cellularity (increased number of neutrophils) sometimes lymphocytes), increased number of CD8+ lymphocytes and decreased CD4+/CD8+ ratio. The considerable presence of B-cells (CD19+ cells) in the BAL fluid collected from our patient is an interesting finding, the given subpopulation having a normal concentration when examined by flow cytometry in the peripheral blood. This aspect suggests a hyperimmune reaction occurring in the lung, which might play a role in the subsequent development of the pulmonary impairment following the initial lung infection. A significant increase in the polyclonal B-cell lymphocytes' number can be related to

bronchus-associated lymphoid tissue hyperplasia. Cases of follicular bronchiolitis sustained by latent adenoviral infection have been reported, suggesting that the lungs of these patients respond to persistent (unknown) antigenic stimulation by activating local immunological mechanisms (22).

A positive diagnosis requires positive history of respiratory infection and one of the following imaging requests: unilateral loss of lung volume with associated hyperlucency (chest X-ray), unilateral reduction in vascular density (chest CT) and unilateral diminished arterial perfusion (Tc 99m lung scan) (23).

Differential diagnosis needs exclusion of pulmonary thromboembolism, asthma, and COPD, lung imaging, PFTs, and absence of therapeutic response being important helpful tools. A hyperlucent lung is an alarming radiographic finding, case in which congenital lobar emphysema, bullous emphysema, pneumothorax, bronchiectasis with air trapping, bronchial stenosis and obliterative bronchiolitis must all be taken into consideration. It is also mandatory to exclude other vascular pathologies, such as congenital pulmonary agenesis or hypoplasia and acquired stenosis or compression of the main pulmonary vessels. None of these abnormalities would both produce diffuse, peripheral ventilatory defects on the single breath image and unilateral lung hypoplasia. Furthermore, air trapping and perfusion modifications are both missing in primary arterial defect and pulmonary thromboembolism (9).

The course of the disease includes recurrent pulmonary infections, bronchiectasis and eventually, respiratory failure (24-27). The prognosis is worse in the presence of bronchiectasis (21, 22).

Treatment of SJMS is usually conservative and consists in preventive respiratory infection therapy and/or curative treatment with antibiotics, bronchodilators associated or not with low-dose inhaled corticoids and pulmonary rehabilitation with complex methods, using even natural therapeutic factors (28-31). In selected cases, such as chronic infection with lung parenchyma destruction, surgical treatment might be useful, including pulmonary lobectomy or pneumectomy (21), multidisciplinary rehabilitation programs after surgery being also necessary (32, 33). Before drastic measures are initiated, multimodal should be assed to guide both short- and long-term treatment options, including pulmonary rehabilitation, trying to find a marker for monitoring the effect of the therapeutic measures for these patients including radiological parameters (34, 35, 36).

Conclusions

We hereby report the case of a patient with positive medical history and characteristic radiological findings. Although he described recurrent respiratory infections during childhood and adolescence each time when admitted to the hospital, the indolent course of the disease and the non-characteristic chest scans led to late diagnosis. We strongly recommend considering SJMS within the differential diagnosis workup of bronchiectasis, the syndrome being underdiagnosed when having in mind the epidemiological data.

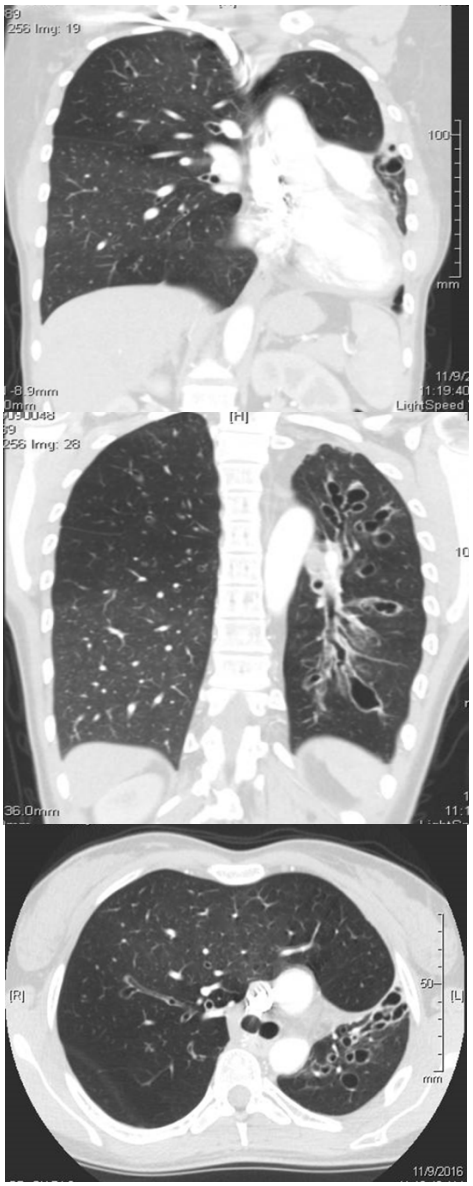

Fig. 1. Thoracic CT scan: small hyperlucent left lung with diminished vascularity and large cystic bronchiectasis, compensatory hyperinflated right lung

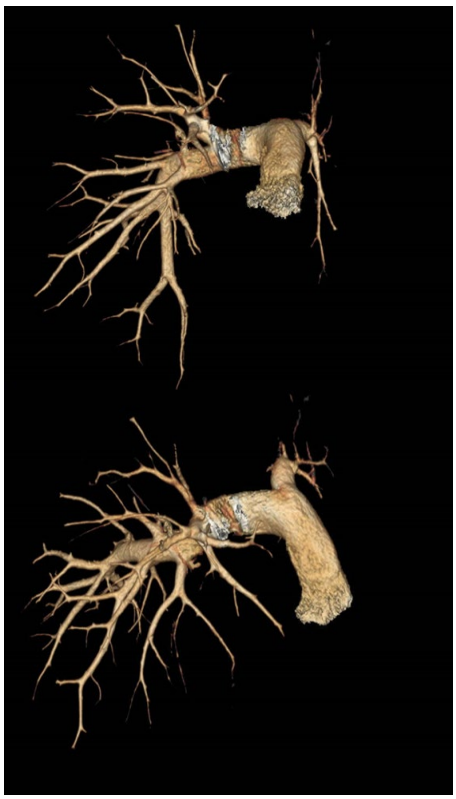

Fig. 2. CT angiography reconstruction: reduced caliber of the left pulmonary artery

Author contributions.
The authors contributed equally to the work.
Declaration of conflict of interests. There is no conflict of interest for any of the authors regarding this paper.
Informed consent. An informed consent was obtained from the patient included in this study.

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