doi: 10.30934/kusbed.1426717 **e-ISSN:** 2149-8571

Kocaeli Üniversitesi Sağlık Bilimleri Dergisi

Olgu Sunumu / Case Report

http://dergipark.org.tr/kusbed

UNILATERAL HYPERLUCENT LUNG: SWYER JAMES MACLEOD SYNDROME

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Geliş Tarihi / Received: 29.01.2024 **Kabul Tarihi / Accepted:** 29.07.2024 **Yayım Tarihi / Published:** 04.09.2024

Abstract

Swyer James Macleod Syndrome, also known as unilateral hyperlucent lung syndrome, is a rare lung condition characterized by the underdevelopment of one lung. This syndrome typically occurs in childhood and is often discovered incidentally during imaging studies for unrelated conditions. Individuals with Swyer James Macleod Syndrome may experience symptoms such as recurrent respiratory infections, shortness of breath, and decreased exercise tolerance. Diagnosis is typically made through imaging studies such as chest X-rays,computed tomography or ventilation-perfusion scans.

Keywords: *Swyer James Macleod Syndrome, ventilation perfusion scan, chest x ray, exercise tolerance.*

Introduction

Swyer-James Macleod Syndrome (SJMS) is a radiological condition characterised by hyperluxence appearance and decreased vascularity with excess ventilation of a single lung lobe without obstruction of the main bronchial airway[1]. It is usually diagnosed after recurrent lung infections. Characteristics such as unilateral air trapping, hyperlucency, a small or normal sized lung, reduced number and size of pulmonary vessels can be observed on radiographs after excluding the potential causes of unilateral radiolucency. It is usually asymptomatic and detected incidental . Although the etiology of SJMS is not known exactly, it is thought to develop after frequent infections especially in childhood[2]. The most common infectious agents are Adenovirus types 3, 7 and 21, *Paramyxovirus*, *Bordetella pertusis*, *Mycobacterium tuberculosis*, *Mycoplasma pneumoniae*, *Haemophilus influenza*, *Streptococcus pneumonia* and *Staphylococcus aureus*. Here, we present a 23-year-old male patient with this rare syndrome.

Case Report

A 23-year-old man with cough and sputum was admitted to our clinic. The patient had a history of frequent lung infections and antibiotic use. The patient was diagnosed with chronic obstructive pulmonary disease, regular use of inhaler medication, and no smoking history. Upon physical examination, it was noted that the respiratory sounds were within normal limits at auscultation and no other abnormalities were observed. The patient was receiving colchicine treatment and was diagnosed with Familial Mediterranean fever (FMF). Pulmonary function test showed obstructive-type ventilatory disorder (FEV1: 1.35 lt/s 34%, FVC: 1.98 lt 42%, FEV1/FVC: 68%). A posteroanterior chest radiograph showed a decrease in the left lung volume and hyperlusensia compared with the right lung (Figure 1). Conventional pulmonary computed tomography (CT) was performed for the differential diagnosis of unilateral hyperlusensia. Pulmonary computed tomography angiography showed hypogenesis in the pulmonary branches of the left lung (Figure 2), and ventilation/perfusion scintigraphy was performed to confirm the diagnosis. Spect-CT showed no parenchymal activity in the left lung, except for a small area of parenchymal activity in the apicoposterior segment (Figure 3). On the basis of these findings, the diagnosis of SJMS was confirmed, and the inhaler treatment was stopped. The patient, who was informed that there is no effective treatment for SJMS, was successfully managed by implementing a vaccination programme (influenza or pneumonia) for preventive purposes of relevant infection.

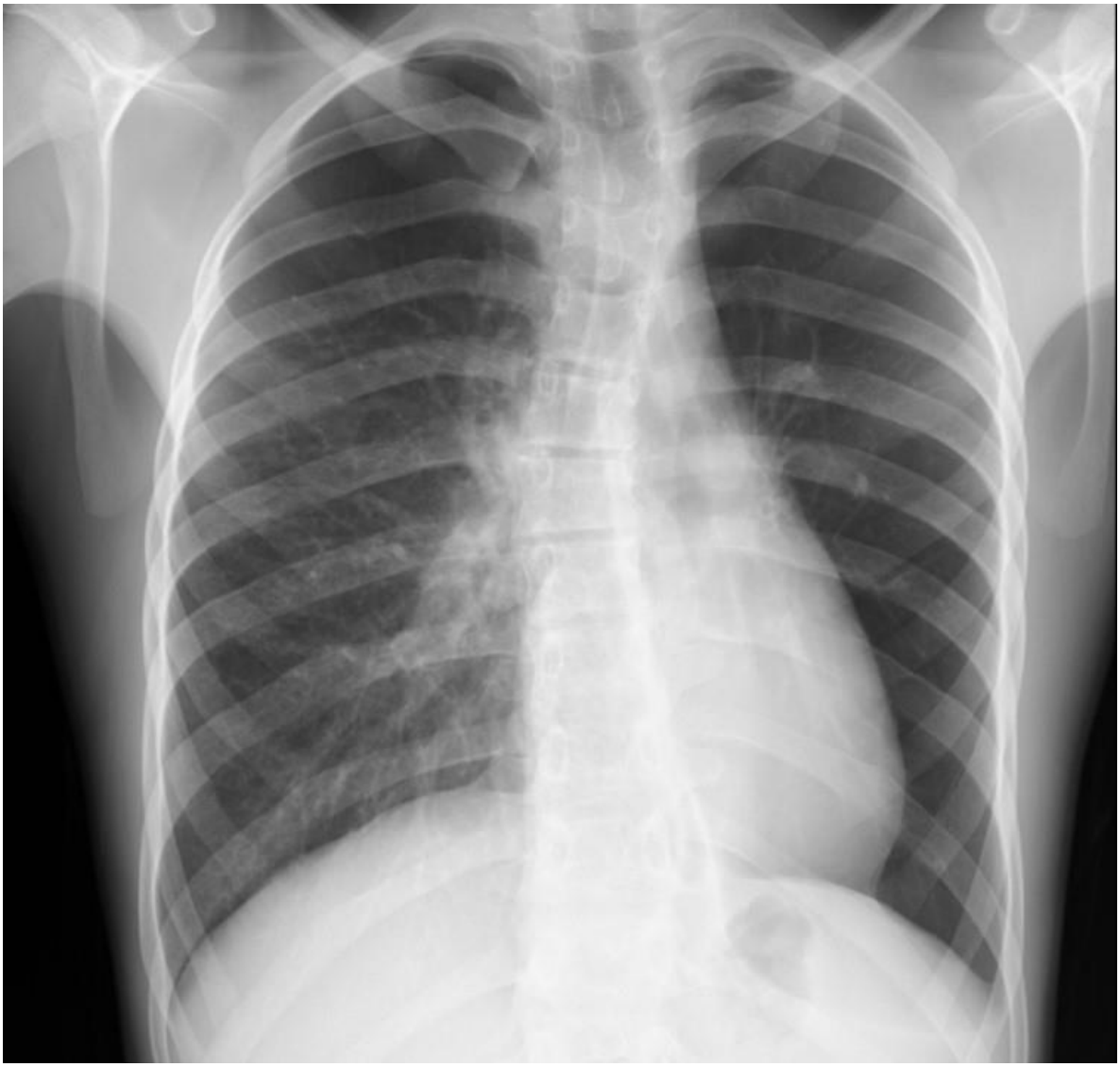

Figure 1. Unilateral hyperlusensia observed in posteroanterior chest radiograph

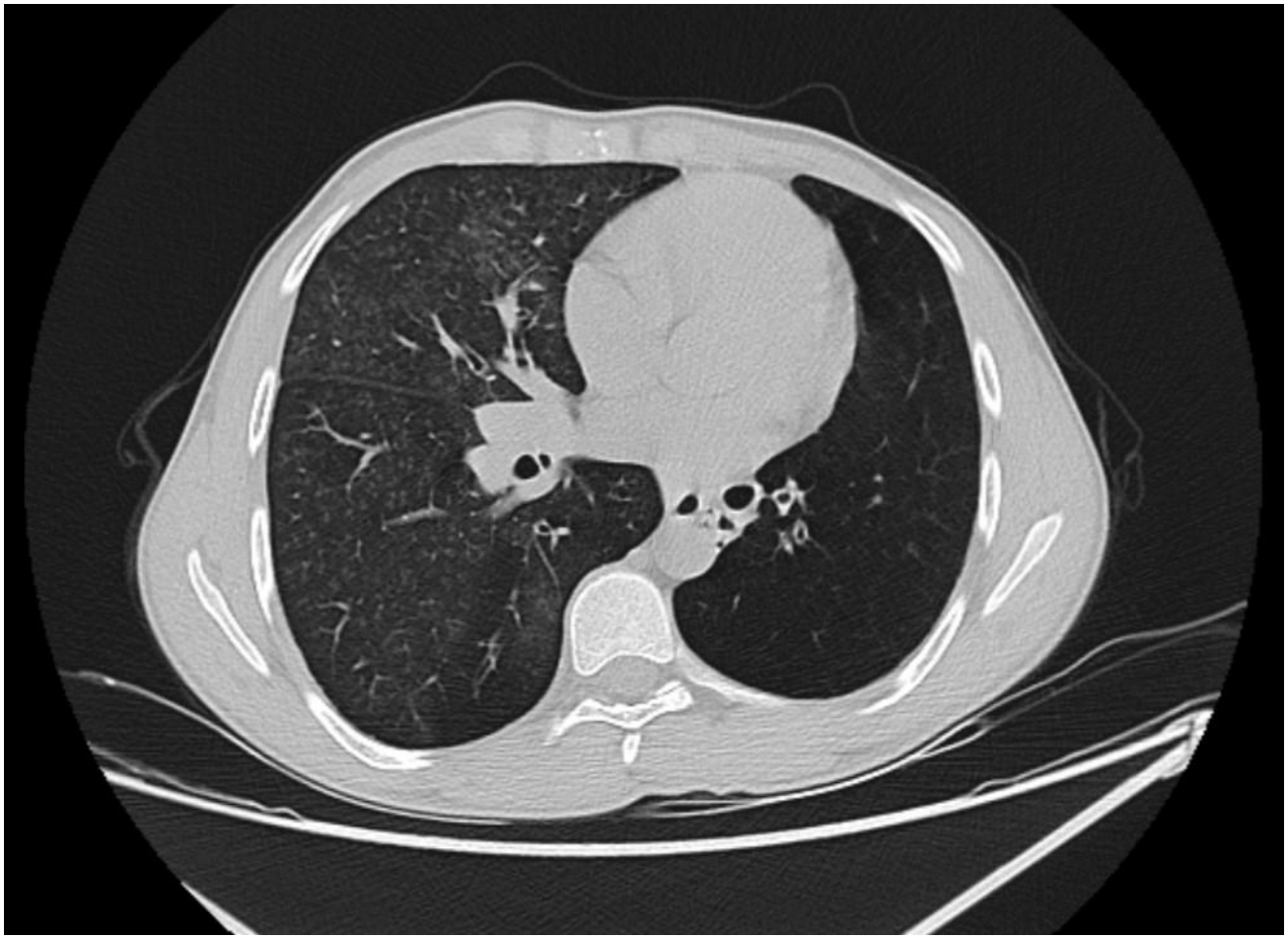

Figure 2. Computed tomography scan showed hypogenesis of the pulmonary artery branches of the left lung.

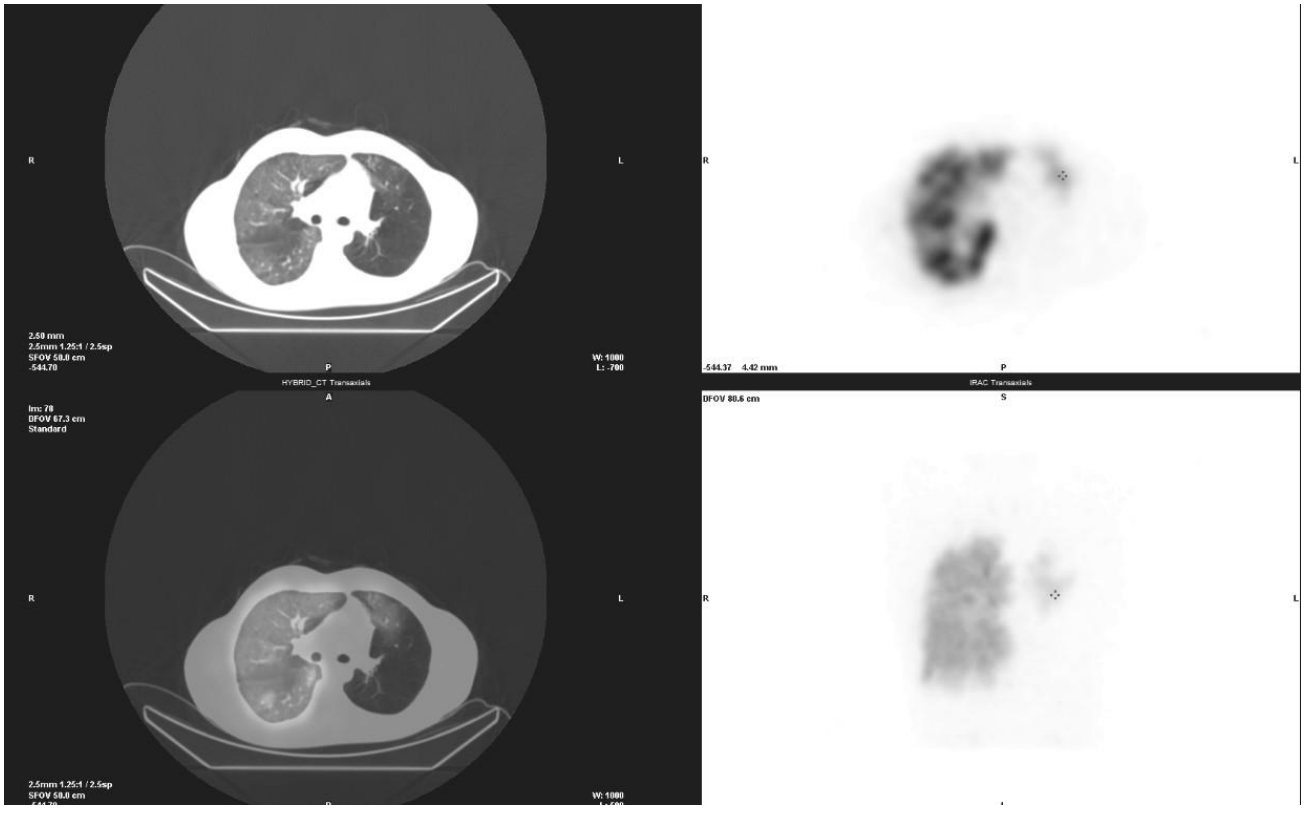

Figure 3. Absence of parenchymal activity in the left lung, with the exception of a small area of parenchymal activity in the apico-posterior segment on Spect CT.

Discussion

SJMS is thought to be associated with post-infectious bronchiolitis obliterans, and the pathogenesis of the syndrome has been attributed to various childhood exposures. These include radiotherapy, measles, pertussis, tuberculosis, mycoplasma, adenovirus infections, and foreign body aspiration. The terminal and respiratory bronchioles are damaged, and the alveoli cannot complete their normal development because of acute bronchiolitis caused by possible viral agents. This retards the development of the affected lung and reduces lung volume and blood flow. In addition to damage to the alveoli, pulmonary circulation is impaired [3-4]. It is usually diagnosed in childhood after recurrent respiratory inflammation, but in very few or none of the patients with bronchiectasis, the clinical picture may be asymptomatic or mild; in such patients, the diagnosis may only be established in adulthood [5]. First described by Swyer and James in 1953 in a 6-year-old boy and reported by Macleod one year later including 9 adults [6]. Inhalation of toxic fumes and gases, aspiration of foreign bodies, radiotherapy, and organ transplants can also cause the syndrome [7,8]. The main pathological mechanism is bronchiolitis with obstruction of the small airways, resulting in alveolar destruction and enlargement of the lung parenchyma and a decrease in peripheral pulmonary vascularity due to inflammation. These pathophysiological changes lead to air trapping and hypoperfusion in the affected segment, resulting in radiographic hyperluxation in the affected lobe [9,10]. In a series of 40 patients in which the causes of unilateral hyperluxation of the lung were investigated, SJMS was found in 45%, localized emphysema in 20%, congenital hypoplastic pulmonary artery in 10%, pulmonary embolism in 10%, bronchial carcinoma in 7.5%, after-effects of radiotherapy in 5%, and benign intrabronchial neoplasm in 2.5% [11]. The disease tends to be asymptomatic, and although the diagnosis may be made incidentally on a chest X-ray taken for other purposes, some patients present to the hospital with cough, reduced exercise tolerance, hemoptysis, and chronic inflammation of the lungs [12]. In a review of 17.459 chest x-rays, this syndrome was found in 0.01% of cases [13]. On chest X-ray, pulmonary vascularization in the affected area was reduced with the appearance of unilateral hyperluxation, and the hilus was found to be smaller than normal. CT is used to establish the diagnosis and severity of bronchiectasis. It is also used to differentiate between lung pathologies such as atelectasis, cavities, and masses[14]. Small, few and narrow pulmonary arteries may be seen on pulmonary angiography. In addition, ventilation-perfusion scintigraphy shows a marked decrease in perfusion on the affected side and can rule out conditions such as pulmonary embolism and pulmonary hypoplasia [15]. In our case, hypoplasia of the pulmonary artery branches and excessive lung aeration on thoracic computed tomography were interpreted as suggesting SJMS. Our patient also had impaired lung perfusion and ventilation on scintigraphy, as seen in SJMS. Pulmonary function tests may show mild to moderate obstructive impairment, as in the present case, and bronchoscopy may be performed to exclude endobronchial obstructive lesions. Treatment of SMJS is conservative in most cases, and recommendations include protection from infections, influenza, and pneumococcal vaccination. In smokers, quitting smoking may result in the resolution of dyspnoea. Although the clinical outcome is generally good, sputum culture should be performed in cases of frequent pulmonary infections. Surgical resection of bronchiectasis may be necessary in uncontrolled cases.

Conclusion

Although SMJS is rare and generally incidental and asymptomatic, this syndrome should not be forgotten in the differential diagnosis of unilateral hyperlucence appearance on chest radiography in patients with exertional dyspnea and a history of frequent pulmonary infections.

Conflict of interest

The authors have no conflicts of interest to disclose.

Compliance with Ethical Statement

Written informed consent was obtained from the patient for publication of this case report.

Financial Support

The authors declared that no financial support was received for this paper.

Author's Contributions

M.P., A.C.P., H.İ.K., H.İ.Y., G.A.: Study idea/Hypothesis; M.P., A.C.P., H.İ.K., H.İ.Y., G.A.: Design; M.P., A.C.P., H.İ.K., H.İ.Y., G.A.: Data Collection; A.C.P.: Analysis; M.P., A.C.P.: Literature review; A.C.P.: Writing; A.C.P.: Critical review.

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