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Swyer-James-MacLeod Syndrome in a Patient with Eisenmenger Syndrome

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Abstract

Swyer-James-MacLeod syndrome is a condition characterized radiologically by hyperlucency in a single lung lobe associated with reduced vascularity, alveolar hyperdistention, and air trapping in the absence of bronchial airway obstruction. Ventricular septal defect is the most common congenital heart defect in childhood and causes irreversible pulmonary hypertension and Eisenmenger syndrome if not treated in a timely manner. In this case report, we present a 25-year-old patient with Swyer-James-MacLeod syndrome and Eisenmenger syndrome. Including Swyer-James-MacLeod syndrome in the differential diagnosis of patients with atypically distributed pulmonary emphysema and unilateral hyperlucency is important for early diagnosis and timely treatment.

Keywords: Swyer-James-MacLeod syndrome • Eisenmenger syndrome • Ventricular septal defect

Introduction

Swyer-James-MacLeod syndrome (SJMS) also referred to as unilateral hyperlucency of the lung, is a rare and complex lung disease. It was first described in Canada in 1953 by British pediatrician Paul Robert Swyer and Canadian radiologist George C. W. James in a six-year-old child with unilateral pulmonary emphysema and hypoplastic pulmonary artery, and one year later British pulmonologist William Mathieson MacLeod published a report of nine patients in whom he detected unilateral hyperlucency [1,2]. Viral and atypical bacterial infections, drugs, radiation therapy, and foreign body aspiration may be involved in the etiology of SJMS. Most patients are asymptomatic and are detected incidentally by lung X-ray performed in adulthood [3].

Eisenmenger syndrome (ES) is the most advanced form of pulmonary arterial hypertension resulting from congenital systemic-to-pulmonary shunts that cause a significant increase in pulmonary vascular resistance and lead to a reversed (pulmonary-to-systemic) or bidirectional shunt. Clinically, ES is a multisystemic disease associated with a large number of complications that seriously impact patients' functional capacity, quality of life, and survival [4].

In this case report, a patient being followed for ES was found to have unilateral hyperlucency on chest X-ray, leading to a diagnosis of SJMS. Our aim was to highlight this rare clinical association, as this is only the second case of concomitant SJMS and ES described in the literature.

Case Description

A 25-year-old woman presented with complaints of headache for 10 days. Her medical history included a diagnosis of ventricular septal defect (VSD) and ES at the age of 15, long-term nebulized bronchodilator therapy for recurrent bronchiolitis until the age of 2 and multiple hospitalizations for bronchopneumonia. The patient was using macitentan, tadalafil, and selexipag, and had no family history of atopy or tuberculosis.

On physical examination, her weight was 64 kg, height was 167 cm, saturation was 85-88%, blood pressure was 150/100 mmHg, heart rate was 48/min and regular, and body temperature was 36.5°C. Her general condition was fair, and she was conscious, cooperative, and oriented. Clubbing of the fingers was observed. On chest auscultation, there was increased intensity of the second heart sound and a systolic murmur at the pulmonary area. Respiratory sounds were comparable on both sides. On neurological examination, limited abduction of the right eye was noted.

Laboratory examinations indicated a white blood cell count of 12090/mm^3, hemoglobin level of 15.9 g/dL, platelet count of 277000/mm^3, and C-reactive protein level of 15.3 mg/dL. Liver and kidney function tests were normal. Electrocardiography revealed sinus rhythm, right axis deviation, P pulmonale, and right bundle branch block.

On echocardiography, a large perimembranous VSD, right-to-left shunt, and enlarged right structures were observed, and the mean pulmonary artery pressure was 50-52 mmHg. Considering the patient's findings of bradycardia and increased intracranial pressure associated with hypertension and limited abduction in the right eye, cranial imaging was performed for suspected brain abscess. Brain MRI revealed an abscess in the anterior right frontal lobe measuring 5.5 × 4 cm in the axial plane and demonstrating peripheral contrast enhancement and pronounced central diffusion restriction. After antiedema treatment, the patient underwent surgery and appropriate antibiotherapy was initiated.

During follow-up in the postoperative ward, the patient had cough and 75-80% oxygen saturation in room air. Posteroanterior chest X-ray revealed significant hyperlucency in the right hemithorax (Figure 1). On chest computed tomography (CT), the left main pulmonary artery and left lung vascularity appeared normal, while the right main pulmonary artery was hypoplastic with abnormally narrow branches (Figure 2). Lung perfusion scintigraphy demonstrated relatively decreased activity distribution in the right lung, and on ventilation scintigraphy, the areas of diffuse hypoperfusion on the right side were not ventilated and did not show filling (Figure 3). No pathological findings were observed on bronchoscopy to rule out the presence of lesions causing endobronchial obstruction and to investigate for additional anatomical abnormalities. Based on these findings, the patient was diagnosed with SJMS in addition to ES. The hospital medical council recommended lobectomy.

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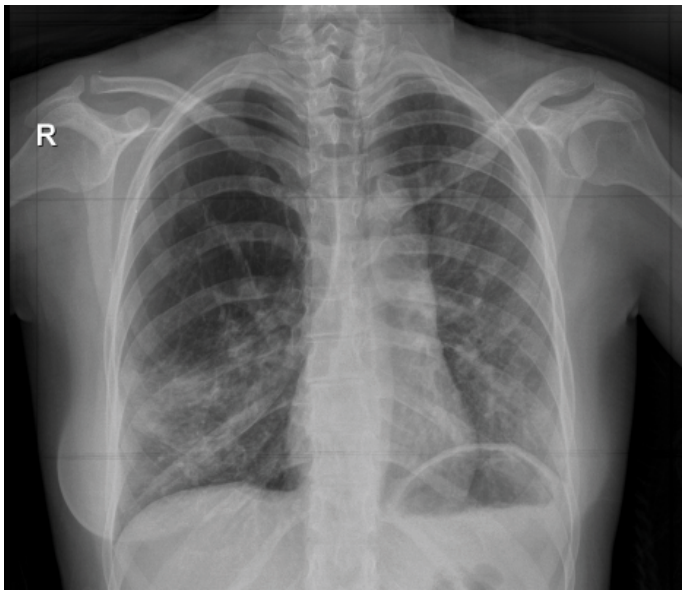

Figure 1. Chest X-ray showing hyperlucency on the right side.

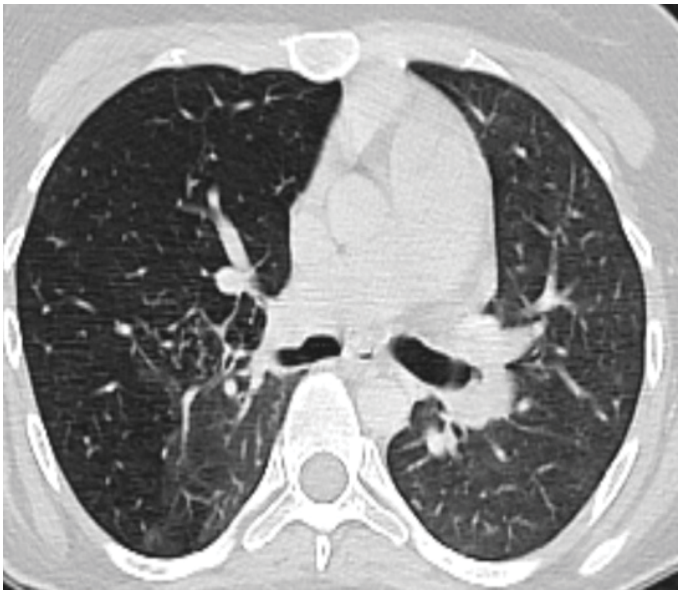

Figure 2. Chest computed tomography showing hypoplastic right main pulmonary artery.

However, the patient refused lobectomy and is currently under outpatient follow-up. With treatment for pulmonary hypertension, her oxygen saturation values in room air are 84-85%.

Results and Discussion

Swyer-James-MacLeod syndrome can be summarized as the obliteration of small bronchioles, absence of peripheral vascular bed, and presence of pulmonary artery hypoplasia and emphysema. This syndrome seems to be an acquired disease that follows viral bronchiolitis and pneumonia in childhood [5]. Adenovirus, measles virus, *Mycoplasma pneumoniae*, *Bordetella pertussis*, or *Mycobacterium tuberculosis* are etiological agents implicated in the development of this syndrome [6]. The main pathological event is bronchiolitis associated with bronchiolar obliteration and subsequent alveolar destruction and dilation in the lung parenchyma. Peripheral pulmonary vascularization decreases due to inflammation. As a result, air trapping and hypoperfusion create the radiographic appearance of hyperlucency [7]. The first-line examination is usually a posteroanterior chest X-ray that reveals decreased bronchovascular signs, a minor hilar shadow, and hyperlucency in the affected segment or lobe. Chest CT is generally the preferred modality

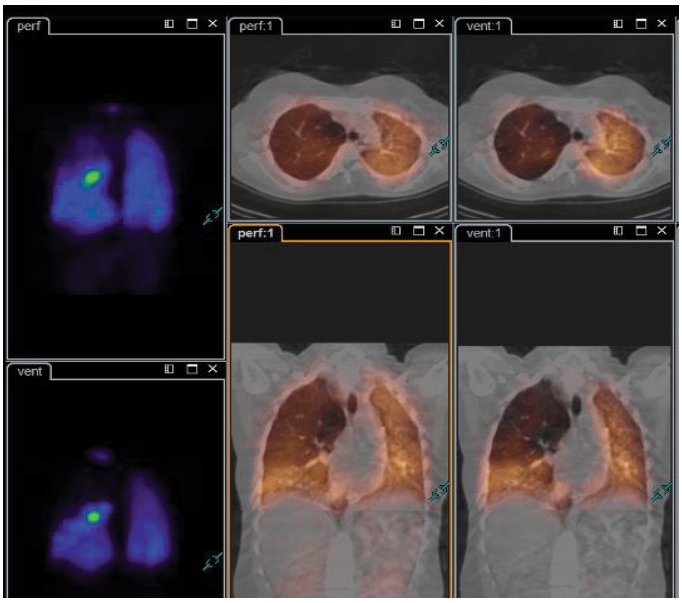

Figure 3. Scintigraphy showing diffuse hypoperfusion and hypoventilation on the right side.

for evaluating the extent and distribution of the disease [8]. CT demonstrates emphysema, bullae, bronchiectasis, and atelectasis, enables evaluation of the pulmonary arterial system and exclusion of endobronchial lesions, and is also noninvasive. In our patient, right hyperlucency and a small hilar shadow were first detected on posteroanterior lung X-ray, followed by chest CT clearly demonstrating a hyperlucent right lung and hypoplastic right pulmonary artery and bronchus.

The management of SJMS is patient-centered and based on a conservative management approach. The prevention of recurrent respiratory infections, administration of influenza and pneumococcal vaccines, and prompt treatment of infections are imperative. Mucolytics, corticosteroids, and inhaled bronchodilators are also used. Chest physiotherapy using techniques such as percussion and postural drainage may be indicated, as well as long-term oxygen therapy in cases of respiratory failure. Patients should be referred for pulmonary rehabilitation [8,9]. These patients are believed to be susceptible to postoperative respiratory problems because of decreased inspiratory capacity and impaired lung diffusion and perfusion [10]. This may explain the low postoperative saturation in our patient. Surgical intervention can be performed in SJMS patients with recurrent lung infections and whose symptoms are not adequately controlled with optimal medical treatment. Surgical options include lung volume reduction surgeries such as pneumonectomy, lobectomy, or segmentectomy [11].

A few patients presenting with pulmonary hypertension and SJMS have been described in the literature [12-15]. It is suggested that SJMS should be regarded as a clinical condition that results in pulmonary hypertension [14]. Congenital heart diseases with left-to-right shunts can cause excess pulmonary flow, leading to pulmonary hypertension and ES. Although the cause of pulmonary hypertension in our patient was a large VSD, it should be kept in mind that SJMS can also cause pulmonary hypertension. Our case is the second in the literature documenting the coexistence of congenital heart disease, pulmonary hypertension, and SJMS. We believe the reason for the small number of cases in the literature may be that SMJS was not investigated sufficiently when congenital heart disease was diagnosed and may have been overlooked during diagnosis. Including and investigating SJMS during diagnosis and providing appropriate treatment may improve the prognosis in such cases by preventing the development of pulmonary hypertension.

Conclusion

Considering SMJS in the differential diagnosis of patients with atypically distributed pulmonary emphysema and unilateral hyperlucency accompanying

congenital heart disease with left-to-right shunt is important for the early diagnosis and timely treatment of this rare condition.

Ethical Approval

Not applicable.

Informed consent

Written informed consent was obtained from the patient for publication of this abstract and any accompanying images.

Conflict of Interest

The authors declare no conflict of interest, financial or otherwise.

Authors' Contributions

Dolunay Gürses: Data curation, supervision, writing – original draft, writing – review and editing.

Merve Oğuz: Conceptualization, data curation, writing – original draft.

Doğangün Yüksel: Clinical and imaging data curation, writing – review and editing.

Furkan Ufuk: Clinical and imaging data curation, writing – review and editing.

Münevver Yılmaz: Clinical data curation, writing – review and editing.

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Availability of Data and Materials

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

References

1. Macleod, WM. "Abnormal transradiancy of one lung." *Thorax* 9 (1954): 147.
2. Swyer, Paul R and GCW James. "A case of unilateral pulmonary emphysema." *Thorax* 8 (1953): 133.
3. Konietzko, N. "Clinical features of chronic obstructive pulmonary disease." *Resp Med* 1171 (2003): 83.
4. Hou, Yulin, Li Wen, Tingting Shu and Rong Dai, et al. "Efficacy and safety of pulmonary vasodilators in the patients with Eisenmenger syndrome: A meta-analysis of randomized controlled trials." *Pulmonary Circul* 11 (2021): 20458940211015823.
5. Fregonese, Laura, Donata Girosi, Elena Battistini and Bruno Fregonese, et al. "Clinical, physiologic, and roentgenographic changes after pneumonectomy in a boy with Macleod/Swyer-James syndrome and bronchiectasis." *Pediatric Pulmonol* 34, (2002): 412-416.
6. Da Silva, Paulo Sérgio Lucas, Renato Lopes and Henrique Monteiro Neto. "Swyer-James-MacLeod syndrome in a surgically treated child: A case report and brief literature review." *J Pediatric Surg* 47 (2012): e17-e22.
7. Behrendt, Anna and Yi Lee. "Swyer-James-MacLeod Syndrome." StatPearls Publishing, 2021.
8. Dirweesh A, Alvarez C, Khan M and Shah N. A unilateral hyperlucent lung - Swyer-James syndrome: A case report and literature review. *Respir Med Case Rep* 20 (2017): 104-106
9. Omar, Muhammad, Muhammad Ahsan Saeed and Amol Patil. "Swyer-James-Macleod Syndrome: Case Report and Brief Literature Review." *South Dakota Med* 72 (2019).
10. https://tgkdc.dergisi.org/uploads/pdf/pdf_TGKDC_631.pdf
11. Yagi, Yuriko, Masato Minami, Yoko Yamamoto and Ryu Kanzaki, e al. "Pneumonectomy in an older patient with swyer-james-macleod syndrome with a giant bulla." *Ann Thoracic Surg* 109 (2020): e263-e265.
12. Demirkan, Burcu, Yesim Guray, Umit Guray and Sule Korkmaz. "Ventricular septal defect and Swyer–James (Macleod's) syndrome together: A case report." *Int J Cardiol* 113 (2006): E4-E6.
13. Chen, I-Chen, Hsiu-Lin Chen, Yi-Ching Liu and Yen-Hsien Wu, et al. "Unique Pulmonary Hypertension in Young Children: A Case Series Study." *Children* 9 (2022): 1064.
14. Yuce, Murat, Nazan Bayram, Orhan Ozer and Vedat Davutoglu. "An unusual cause of pulmonary hypertension: Swyer–James–Macleod syndrome." *Int J Cardiol* 223 (2016): 212-214.
15. Capela, Carlos, Paulo Gouveia, Marco Sousa and Maria J. Regadas. "Adult diagnosis of Swyer-James-MacLeod syndrome: a case report." *J Med Case Rep* 5 (2011): 1-4.

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