ARCHIVES OF THE TURKISH SOCIETY OF CARDIOLOGY

Official journal of the TURKISH SOCIETY OF CARDIOLOGY

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

Swyer-James-MacLeod Sendromu ve Eisenmenger Sendromu Birlikteliği

ABSTRACT

Swyer-James-MacLeod syndrome is characterized radiologically by hyperlucency in a single lung lobe, accompanied by reduced vascularity, alveolar hyperdistention, and air trapping, without bronchial airway obstruction. The most common congenital heart defect in childhood, ventricular septal defect, leads to irreversible pulmonary hypertension and Eisenmenger syndrome if not treated promptly. This case report presents a 25-year-old patient with Swyer-James-MacLeod syndrome and Eisenmenger syndrome. It is crucial to include Swyer-James-MacLeod syndrome in the differential diagnosis of patients with atypically distributed pulmonary emphysema and unilateral hyperlucency for early diagnosis and timely intervention.

Keywords: Eisenmenger syndrome, Swyer-James-MacLeod syndrome, ventricular septal defect

ÖZET

Swyer-James-Macleod Sendromu, bronşiyal hava yolu tıkanması olmadan tek akciğer lobunun havalanma fazlalığı ile hiperlüsen görünüm ve azalmış vaskularite ile karakterize radyolojik bir durumdur. Ventriküler septal defekt çocuklukta en sık görülen konjenital kalp kusuru olup zamanında tedavi edilmezse geri dönüşü olmayan pulmoner hipertansiyona ve Eisenmenger sendromuna neden olur. Bu vaka raporunda Swyer-James-Macleod Sendromu ve Eisenmenger sendromu birlikteliği olan 25 yaş hasta sunuldu. Atipik dağılımlı pulmoner amfizemi, tek taraflı hiperlüsensisi olan olgularda Swyer-James-Macleod Sendromu'nun ayırıcı tanıda düşünülmesi erken tanı ve uygunsuz tedavilerin engellenmesi açısından önemlidir.

Anahtar Kelimeler: Eisenmenger Sendromu, Swyer-James Macleod Sendromu, ventriküler septal defekt

CASE REPORT
OLGU SUNUMU

Dolunay Gürses[1]
Merve Oğuz[1]
Doğangün Yüksel[2]
Furkan Ufuk[3]
Münevver Yılmaz[1]

[1]*Department of Pediatric Cardiology, Pamukkale University Medical School, Denizli, Türkiye*
[2]*Department of Nuclear Medicine, Pamukkale University Medical School, Denizli, Türkiye*
[3]*Department of Radiology, Pamukkale University Medical School, Denizli, Türkiye*

Corresponding author:
Merve Oğuz
✉ mrvkorkutoguz@gmail.com

Received: May 05, 2023
Accepted: November 02, 2023

Cite this article as: Gürses D, Oğuz M, Yüksel D, Ufuk F, Yılmaz M. Swyer-James-MacLeod syndrome in a patient with Eisenmenger syndrome. *Turk Kardiyol Dern Ars.* 2024;52(6):460-463.

DOI:10.5543/tkda.2023.59829

Swyer-James-MacLeod syndrome (SJMS), also known as unilateral hyperlucency of the lung, is a rare and complex pulmonary condition. It was first described in 1953 in Canada by British pediatrician Paul Robert Swyer and Canadian radiologist George C. W. James[1], who observed a six-year-old child with unilateral pulmonary emphysema and a hypoplastic pulmonary artery. The following year, British pulmonologist William Mathieson MacLeod[2] reported on nine patients exhibiting unilateral hyperlucency. The etiology of SJMS may involve viral and atypical bacterial infections, exposure to drugs, radiation therapy, and foreign body aspiration. Most patients are asymptomatic, with the condition often discovered incidentally during adulthood lung X-rays.[3]

Eisenmenger syndrome (ES) represents the most advanced form of pulmonary arterial hypertension, arising from congenital systemic-to-pulmonary shunts that significantly increase pulmonary vascular resistance, resulting in a reversed (pulmonary-to-systemic) or bidirectional shunt. Clinically, ES is a multisystemic disease associated with numerous complications that severely affect patients' functional capacity, quality of life, and survival.[4]

In this case report, we discuss a patient monitored for ES who displayed unilateral hyperlucency on a chest X-ray, leading to a diagnosis of SJMS. Our aim is to underscore this rare clinical association, marking only the second instance of concurrent SJMS and ES documented in medical literature.

Case Report

A 25-year-old woman presented with a 10-day history of headaches. Her medical history included a diagnosis of ventricular septal defect (VSD) and ES at age 15, ongoing nebulized bronchodilator therapy for recurrent bronchiolitis since infancy, and multiple hospital admissions for bronchopneumonia. The patient was prescribed macitentan, tadalafil, and selexipag, with no family history of atopy or tuberculosis.

During the physical examination, she weighed 64 kg, stood at 167 cm, and showed an oxygen saturation of 85-88%. Her blood pressure was 150/100 mmHg, heart rate was slow at 48 beats per minute and regular, and body temperature was 36.5°C. Her general condition was stable, and she was conscious, cooperative, and oriented. Clubbing of the fingers was observed. Chest auscultation revealed an intensified second heart sound and a systolic murmur at the pulmonary area, with comparable respiratory sounds on both sides. A neurological examination noted limited abduction of her right eye.

Laboratory tests showed a white blood cell count of 12,090/mm^3, hemoglobin level of 15.9 g/dL, platelet count of 277,000/mm^3, and C-reactive protein level of 15.3 mg/dL. Liver and kidney function tests were within normal ranges. Electrocardiography indicated sinus rhythm, right axis deviation, P pulmonale, and right bundle branch block.

Echocardiography identified a large perimembranous VSD, a right-to-left shunt, and enlargement of the right structures. Her mean pulmonary artery pressure was recorded at 50-52 mmHg. Considering the patient's findings of bradycardia, increased intracranial pressure associated with hypertension, and limited abduction of the right eye, cranial imaging was conducted to investigate a suspected brain abscess. Brain magnetic resonance imaging (MRI) revealed a 5.5 × 4 cm abscess in the anterior right frontal lobe on the axial plane, showing peripheral contrast enhancement and pronounced central diffusion restriction. After receiving anti-edema treatment, the patient underwent surgery, and appropriate antibiotic therapy was initiated.

During her postoperative recovery in the ward, the patient experienced coughing and an oxygen saturation of 75-80% in room air. A posteroanterior chest X-ray revealed significant hyperlucency in the right hemithorax (Figure 1). Chest computed tomography (CT) showed normal vascularity in the left lung and main pulmonary artery, while the right main pulmonary artery appeared hypoplastic with abnormally narrow branches (Figure 2). Lung perfusion scintigraphy indicated relatively decreased activity distribution in the right lung. Ventilation scintigraphy revealed areas of diffuse hypoperfusion on the right side that were not ventilated and did not exhibit filling (Figure 3). Bronchoscopy was performed to rule out lesions causing endobronchial obstruction and to investigate additional anatomical abnormalities; however, no pathological findings were observed. Based on these results, the patient was diagnosed with SJMS in addition to ES. The hospital medical council recommended a lobectomy, but the patient declined and is currently under outpatient follow-up. With ongoing treatment for pulmonary hypertension, her oxygen saturation levels in room air range from 84-85%.

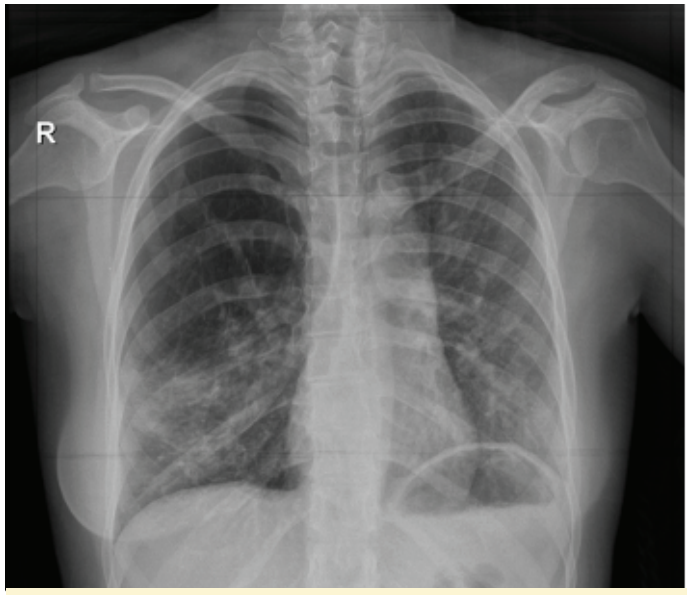

Figure 1. Chest X-ray showed significant hyperlucency in the right hemithorax.

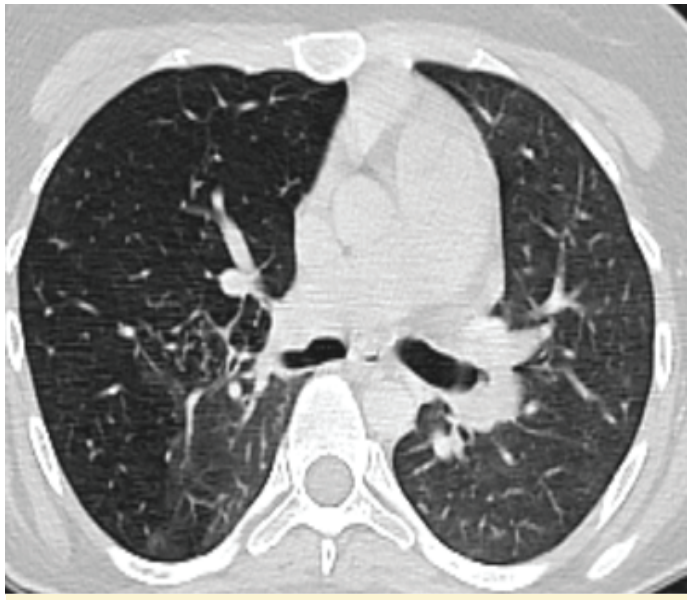

Figure 2. Chest computed tomography (CT) revealed a hypoplastic right main pulmonary artery and abnormally narrow branches.

ABBREVIATIONS

CT	Computed tomography
ES	Eisenmenger syndrome
MRI	Magnetic resonance imaging
SJMS	Swyer-James-MacLeod syndrome
VSD	Ventricular septal defect

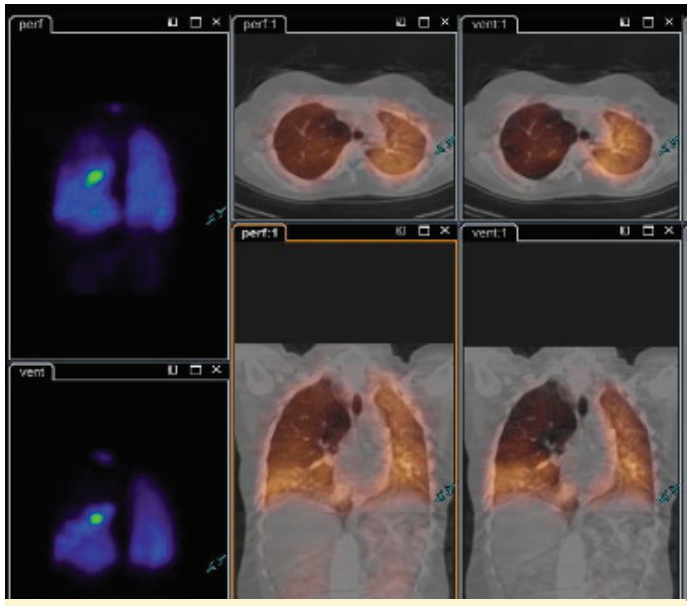

Figure 3. Lung perfusion scintigraphy indicated diffuse hypoperfusion and hypoventilation on the right side.

Discussion

Swyer-James-MacLeod syndrome is characterized by the obliteration of small bronchioles, absence of a peripheral vascular bed, and the presence of pulmonary artery hypoplasia and emphysema. It appears to be an acquired condition resulting from viral bronchiolitis and pneumonia in childhood.[5] Adenovirus, measles virus, *Mycoplasma pneumoniae*, *Bordetella pertussis*, or *Mycobacterium tuberculosis* have been implicated as etiological agents in the development of this syndrome.[6] The primary pathological event is bronchiolitis, which leads to bronchiolar obliteration, subsequent alveolar destruction, and dilation of the lung parenchyma. Inflammation reduces peripheral pulmonary vascularization, resulting in air trapping and hypoperfusion that create the radiographic appearance of hyperlucency.[7] The first-line examination typically involves a posteroanterior chest X-ray, which reveals decreased bronchovascular markings, a diminished hilar shadow, and hyperlucency in the affected segment or lobe. Chest CT, generally the preferred method, evaluates the extent and distribution of the disease comprehensively.[8] As a noninvasive modality, CT displays features such as emphysema, bullae, bronchiectasis, and atelectasis, and facilitates the evaluation of the pulmonary arterial system while ruling out endobronchial lesions. In our patient, initial posteroanterior lung X-ray showed right hyperlucency and a small hilar shadow, with subsequent chest CT clearly demonstrating a hyperlucent right lung along with a hypoplastic right pulmonary artery and bronchus. The significance of CT in differentiating pulmonary hypertension should be emphasized.

Management of SJMS is patient-centered, focusing on conservative strategies. Preventing recurrent respiratory infections, administering influenza and pneumococcal vaccines, and prompt treatment of infections are crucial. Therapeutic measures also include mucolytics, corticosteroids, and inhaled bronchodilators. Chest physiotherapy techniques such as percussion and postural drainage may be indicated, along with long-term oxygen therapy for respiratory failure. Patients should also be referred for pulmonary rehabilitation.[8,9] These patients are at an increased risk of postoperative respiratory complications due to decreased inspiratory capacity and impaired lung diffusion and perfusion,[10] which may explain the low postoperative saturation in our patient. Surgical interventions can be performed in patients with SJMS who experience recurrent lung infections and whose symptoms are not adequately controlled by optimal medical treatment. Surgical options include lung volume reduction surgeries such as pneumonectomy, lobectomy, or segmentectomy.[11]

A few cases of patients with pulmonary hypertension and SJMS have been reported in the literature.[12-15] It has been suggested that SJMS should be considered a clinical condition that leads to pulmonary hypertension.[14] Although we believe that SJMS contributed to the pulmonary hypertension in our patient, the coexistence of SJMS and ES may be coincidental. This case represents the second instance in the literature of concurrent congenital heart disease, pulmonary hypertension, and SJMS. The limited number of cases reported in the literature may result from insufficient investigation of SMJS during the diagnosis of congenital heart disease, which could lead to its oversight. As the number of case studies grows, the understanding of this relationship will become clearer.

Informed Consent: Written informed consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – D.G., D.Y.; Design – M.O., F.U., M.Y.; Supervision – D.G.; Materials – D.Y., F.U.; Data Collection and/or Processing – M.O., M.Y..; Analysis and/or Interpretation – D.G., M.O.; Literature Review –D.G., M.O., D.Y., F.U., M.Y.; Writing – D.G., M.O.; Critical Review – D.G., M.Y.

Use of AI for Writing Assistance: No artificial intelligence assisted technologies were used in this article.

Conflict of Interest: The authors have no conflicts of interest to declare.

Funding: The authors declared that this study received no financial support.

References

1. Swyer PR, James GC. A case of unilateral pulmonary emphysema. *Thorax.* 1953;8(2):133–136. [CrossRef]
2. Macleod WM. Abnormal transradiancy of one lung. *Thorax.* 1954;9(2):147–153. [CrossRef]
3. Konietzko N. Clinical Feartures of Chronic Obstructive Pulmonary Disease. In: Gibson GJ, Geddes DM, Costabel U, Sterk PJ, Corrin B, editors. *Respiratory medicine*. 3rd ed. London: Saunders; 2003:1171–1183.
4. Hou Y, Wen L, Shu T, Dai R, Huang W. Efficacy and safety of pulmonary vasodilators in the patients with Eisenmenger syndrome: A meta-analysis of randomized controlled trials. *Pulm Circ.* 2021;11(2):20458940211015823. [CrossRef]
5. Fregonese L, Girosi D, Battistini E, et al. Clinical, physiologic, and roentgenographic changes after pneumonectomy in a boy with Macleod/Swyer-James syndrome and bronchiectasis. *Pediatr Pulmonol.* 2002;34(5):412–416. [CrossRef]
6. da Silva PS, Lopes R, Neto HM. Swyer-James-MacLeod syndrome in a surgically treated child: A case report and brief literature review. *J Pediatr Surg.* 2012;47:e17–e22. [CrossRef]

7. Behrendt A, Lee Y. Swyer-James-MacLeod Syndrome. Accessed May 22, 2024. https://www.statpearls.com/point-of-care/29791.
8. Dirweesh A, Alvarez C, Khan M, Shah N. A unilateral hyperlucent lung - Swyer-James syndrome: A case report and literature review. *Respir Med Case Rep.* 2017;20:104–106. [CrossRef]
9. Omar M, Saeed MA, Patil A. Swyer-James-Macleod Syndrome: Case report and brief literature review. *S D Med.* 2019;72(11):518–520.
10. Oto Ö, Açıkel Ü, Tüzün E, Uğurlu ŞB, Çatalyürek HH. Swyer-James MacLeod (Unilateral hiperlüsent akciğer) Sendromlu bir hastada çift kapak replasmanı. *Turk J Thorac Cardiovasc Surg.* 1995;3:27–29.
11. Yagi Y, Minami M, Yamamoto Y, et al. Pneumonectomy in an older patient with Swyer-James-MacLeod Syndrome with a giant bulla. *Ann Thorac Surg.* 2020;109(4):e263–e265. [CrossRef]
12. Demirkan B, Guray Y, Guray U, Korkmaz S. Ventricular septal defect and Swyer-James (Macleod's) syndrome together: A case report. *Int J Cardiol.* 2006;113(1):E4–E6. [CrossRef]
13. Chen IC, Chen HL, Liu YC, et al. Unique pulmonary hypertension in young children: A case series study. *Children (Basel).* 2022;9(7):1064. [CrossRef]
14. Yuce M, Bayram N, Ozer O, Davutoglu V. An unusual cause of pulmonary hypertension: Swyer-James-Macleod syndrome. *Int J Cardiol.* 2016;223:212–214. [CrossRef]
15. Capela C, Gouveia P, Sousa M, Regadas MJ. Adult diagnosis of Swyer-James-MacLeod syndrome: A case report. *J Med Case Rep.* 2011;5:2. [CrossRef]