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Radiological Imaging in A Rare Case of Swyer-James-Macleod Syndrome: A Case Report

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Abstract

Sweyer James MacLeod syndrome (SJMS), also known as unilateral hyperlucent lung syndrome, is a rare pulmonary disorder characterized by unilateral hypoplasia or agenesis of the pulmonary vasculature, typically affecting one lung. This results in reduced perfusion and subsequent air trapping, leading to unilateral hyperlucency on chest radiographs.

Clinical presentation varies widely, often presenting asymptomatically or with recurrent respiratory infections in childhood. Diagnosis requires a combination of clinical history, imaging studies (such as chest radiography, CT scan, and ventilation-perfusion scans), and pulmonary function tests. Management focuses on symptomatic relief, including treatment of infections and pulmonary rehabilitation.

In this report, we present a case of 14-year-old girl presented with a three-day history of fever and cough with sputum with previous history of severe pulmonary infection. Diagnosing early is vital for long term survival and increasing quality of life.

Chest radiograph and computed tomography of this case showed a right hyperlucent lung with diminished vascularity.

Due to its rarity and severity, Swyer-James syndrome requires meticulous diagnosis and individualized treatment. Patients with this disease require a holistic approach to treatment, as shown in the case study. The respiratory health and quality of life of people with Swyer-James syndrome can be greatly enhanced with prompt diagnosis, efficient therapy, and continuous monitoring. We must prioritize ongoing research and raise clinical awareness to better understand and treat this uncommon lung condition. Stable pulmonary function improves with time; therefore, most patients have a good prognosis.

Keywords: SJMS, Pulmonary Function, Diminished Vascularity, Hemithorax

Case report

A 14-year-old girl presented with a three-day history of fever and cough with sputum. At age of two, she had been hospitalized due to severe pulmonary infection. Her medical history has been uneventful since then. Clinical examination of the chest revealed decreased breath sounds and asymmetrical chest expansion, predominantly on the right side. Pulmonary function tests revealed mild restrictive ventilatory defect and reduced diffusing capacity,

Figure 1: Chest radiograph shows reduced right lung volume and rightward mediastinal shift. There is relative hyperlucency of the right hemithorax

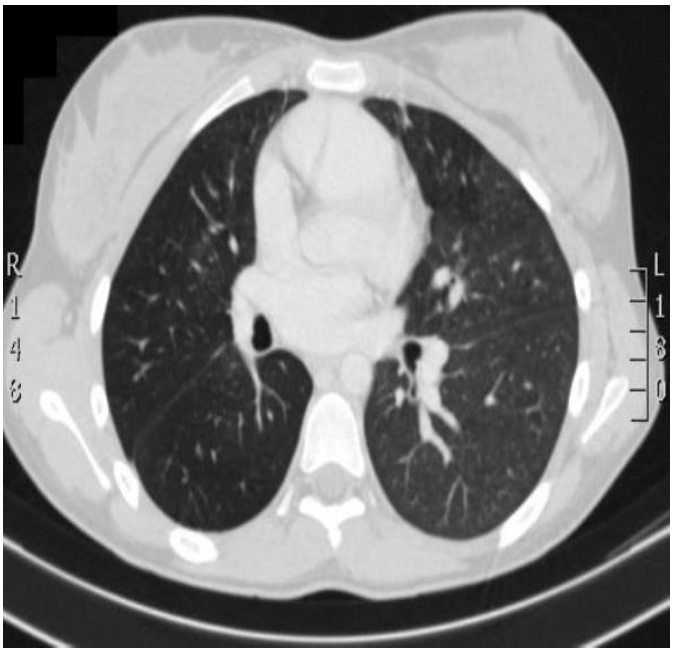

Figure 2: Axial CT image shows right hyperlucent lung with diminished vascularity on right side

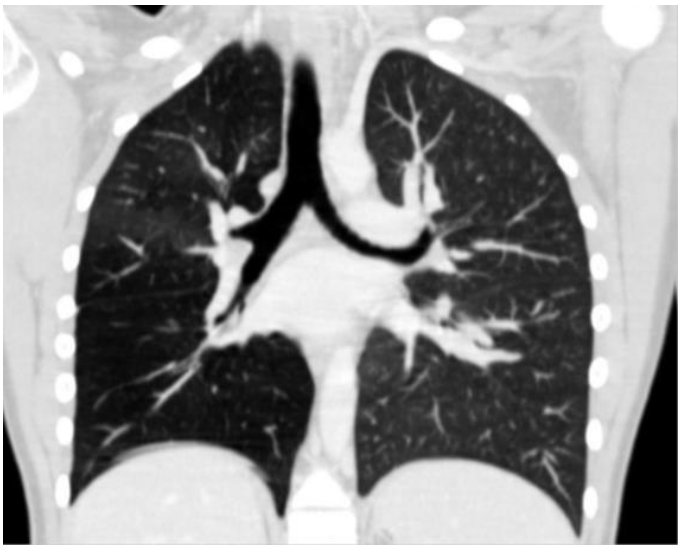

Figure 3: Coronal CT image with maximum intensity projection (MIP) shows right hyperlucent lung with diminished vascularity

Discussion

Swyer-James-MacLeod syndrome (SJMS) is an uncommon pulmonary condition distinguished by the presence of abnormally transparent lung tissue on one side of the chest. This disorder is acquired when there is a disruption in the development of blood vessels and lung tissue in an area affected by bronchiolitis obliterans during childhood. This leads to the formation of underdeveloped blood vessels and portions of the lungs that seem abnormally transparent [1]. Typically, this condition is identified during childhood and is characterized by recurring chest infections. Nevertheless, certain people may have a reduced number of symptoms, leading to the possibility of overlooking the diagnosis until adolescence or maturity [2]. The diagnosis of SJMS is determined by identifying certain imaging features. The characteristic features observed on radiographs in individuals with this illness are unilateral lung or lobar hyperlucency accompanied by a decrease in lung capacity. The diagnosis is confirmed by the demonstration of expiratory air trapping [3]. CT scans show reduced density of lung tissue and

diminished vascular patterns in the afflicted areas. The whole of the lung can be impacted, however there may also be involvement in certain lobes, segments, or subsegments in a scattered distribution. Regions of reduced tissue density can be observed amidst regions of unaffected tissue in both lungs [4]. Observation of bronchiectasis, mild subpleural parenchymal scarring, atelectasis, and pulmonary artery hypoplasia can be made on the afflicted side [1]. The primary distinction of SJMS is in its differentiation from an endobronchial lesion that only partially obstructs the lumen of a lobar or main bronchus, such as a foreign body in infants or a bronchial tumor in adults. Additional differentials encompass pulmonary bullae and pulmonary artery agenesis [3]. Computed tomography (CT) is crucial in distinguishing between different diagnoses, as it may show if the bronchial tree is open and provide a more detailed understanding of the lung tissue and blood vessels.

CT is more effective in displaying the size and spread of the illness and can provide a clearer picture of associated disorders such bronchiectasis [4].

The primary method of therapy is a cautious strategy that focuses on controlling existing lung infections and avoiding new ones. The long-term consequences that can occur in adulthood include pulmonary hypertension. However, the exact extent of the morbidity and mortality associated with this disease is not yet fully defined [1, 2].

Conclusion

Swyer-James syndrome is a rare and distinctive pulmonary condition characterized by the underdevelopment of one lung due to prior severe respiratory infections, often in childhood. Its clinical presentation can range from asymptomatic to significant respiratory distress, and its management primarily focuses on alleviating symptoms and preventing complications.

The diagnosis relies on a combination of clinical history, imaging studies, and pulmonary function tests, which help differentiate it from other similar conditions. Although the syndrome can lead to varying degrees of respiratory impairment, many individuals manage well with appropriate care and intervention. Regular monitoring and a tailored treatment approach are crucial to maintaining quality of life and preventing long-term complications.

As a condition with a relatively low prevalence, continued research and awareness are essential for advancing our understanding of Swyer-James syndrome. By improving diagnostic techniques and treatment options, healthcare providers can better support individuals affected by this rare but impactful disorder.

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