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Clinical case

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Swyer James MacLeod syndrome. Case report of hyperlucid lung

Síndrome de Swyer James MacLeod. Reporte de caso de pulmón hiperlúcido

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ABSTRACT. Introduction: respiratory infections in childhood can eventually lead to rare diseases such as Swyer James MacLeod syndrome, which, although it does not have a defined etiology, has been associated with these respiratory antecedents. Reports a prevalence of 0.01% of 17,450 chest X-ray. **Clinical case:** we report the case of a young woman who consults for chest pain without irradiation and that imaging studies show greater radiolucency of the left lung, in addition to this, respiratory function tests point to a restrictive process; leading to the diagnosis of this rare syndrome. **Conclusions:** Swyer James MacLeod syndrome is a clinic-radiological entity that is usually asymptomatic and its diagnosis can be considered an incidental finding.

Keywords: James MacLeod Swyer syndrome, hyperlucid lung, case report.

RESUMEN. Introducción: las infecciones respiratorias en la infancia pueden eventualmente desarrollar enfermedades poco frecuentes como el síndrome de Swyer James MacLeod; si bien, no tiene una etiología definida, se ha asociado a estos antecedentes respiratorios. Reporta una prevalencia de 0.01% en 17,450 radiografías de tórax. **Caso clínico:** se reporta el caso de una joven mujer que consulta por dolor torácico sin irradiación y que los estudios de imagen demuestran mayor radiolucencia del pulmón izquierdo. Aunado a ello, las pruebas funcionales respiratorias orientan a un proceso restrictivo, lo que lleva al diagnóstico de este raro síndrome. **Conclusiones:** el síndrome de Swyer James MacLeod es una entidad clínico-radiológica que se presenta habitualmente asintomática y su diagnóstico puede considerarse como un hallazgo incidental.

Palabras clave: síndrome de Swyer James MacLeod, pulmón hiperlúcido, caso clínico.

INTRODUCTION

Swyer James MacLeod syndrome was described simultaneously in 1950 by three professionals, highlighting the interrelation of the different disciplines of medicine, pulmonology and radiology. The three physicians, one of English origin and two Canadians, coincided in the findings of the syndrome that today bears their names. It is related to bronchitis and bronchiolitis obliterans, acquired during childhood from a bacterial or viral infection, and its diagnosis is usually incidental in the course of a radiological evaluation in which unilateral hyperlucency of the lung is revealed.

CLINICAL CASE

A 31-year-old female patient consulted a national hospital with a history of mild retrosternal pain, insidious, not radiating and relieved by sleeping. Due to poorly defined findings in the X-ray, chest tomography was requested and she was transferred to our center. She reported a history of respiratory diseases in childhood and adolescence, such as pneumonia, bronchitis, common colds treated as outpatients, and during the pandemic she suffered from uncomplicated COVID-19.

Admitted in good general and nutritional condition, SO_2 99%, heart rate (HR) 74 beats per minute (bpm), without

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dyspnea or cyanosis. Lungs with adequate air entry and rhythmic heart without murmurs. Labs: hemoglobin 13 mg/mL; hematocrit 39%; platelets 224 × 10^9/L; D-dimer 273.59; alpha 1 antitrypsin 98.8 (82.50-320). Pulmonary function tests: spirometry; FEV1 3.09 (52%) 1.55; FVC 3.64 (59%) 2.13; FEF 25-75% 85.39 (88%) 72.89, suggestive of restriction. Body plethysmography/flow-volume uninterpretable. SB diffusion, DLCO 8.18 (80%) mild decrease. Lenin walk, walks 225 m, being 35% of his predicted, without pauses during the test. NADIR: SO_2 88% with no need for supplemental oxygen.

Chest X-ray showed left lung overdistended, displacing the mediastinum to the right and hyperlucent; computed axial tomography (CT) showed, in addition, anomalous distribution of the pulmonary vasculature with areas of hypoperfusion of variable distribution *(Figure 1)*.

Ruling out probable pulmonary thromboembolism (PTE), CT angiography was requested, which showed no evidence of PTE in the digital reconstructions, showing continuity of opacification of first and second order pulmonary arteries. Lung with left hyperclarity and hypoplasia of the ipsilateral pulmonary artery measuring 9 mm in diameter A-P compared to 20 mm on the right. Consider Swyer James MacLeod syndrome *(Figure 2)*. Echocardiogram, reports dextrocardia with EF at 55%. Low probability of PAH with pulmonary systolic pressure at 20 mmHg. Abdominal CT reports no abnormalities.

DISCUSSION

Swyer James MacLeod syndrome is a rare clinical-radiological entity that reports a prevalence of 0.01% in 17,450 chest radiographs.[1,2] We present the case of a young woman with a history of repeated respiratory disorders in childhood, which probably led her to develop structural changes in her lung. Due to the characteristics shown, this syndrome has no defined etiology, but it is related to viral infections such as influenza A virus, respiratory syncytial virus, mumps, paramyxovirus, adenovirus type 3, 7 and 21; as well as a bacterial origin, in which Bordetella pertussis, Mycobacterium tuberculosis and Mycoplasma pneumoniae are involved.[1] It is a rare emphysematous disease characterized by obliteration of the small bronchioles, hypoplasia or absence of pulmonary artery and peripheral vascular bed.[3] The functional abnormality of this syndrome is undoubtedly diffuse chronic expiratory obstruction with distal air trapping.[4] The most distinctive feature compared to emphysema is the absence of chronic obstructive pathology in the small airway,[3] which coincides with the FEF 25-75% 85.39 (88%) 72.89 data obtained from the patient.

A mild chest pain prompted consultation and radiography showed a hyperlucent left lung. It is most commonly diagnosed in infancy, typically occurring in children under eight years of age, before the lung has completed lung development and maturation.[5] It may run asymptomatically into adulthood, present with usually asymptomatic spontaneous pneumothorax,[2] or as a rare emergency condition,[3] as calcified bullae[6] and with placental transfiguration of the lung parenchyma.[7] The patient's chest X-ray and CT scan showed hyperlucent segmental lung with altered vascularity, which is usually described by other authors as unilateral or lobar pulmonary hyperclarity, associated with air trapping of the hyperlucent lung during expiration.[2,5,8] In order to rule out the differential diagnosis of pulmonary thromboembolism, angiotomography was requested, which showed anatomical abnormalities of the left pulmonary artery with a smaller caliber and decreased flow. The spirometric pattern showed restrictive process. Pulmonary scintigraphy was not performed that could show decreased ventilation of the pathologic lung secondary to emphysematous changes and a marked decrease in perfusion, as a consequence of the reduced caliber of the

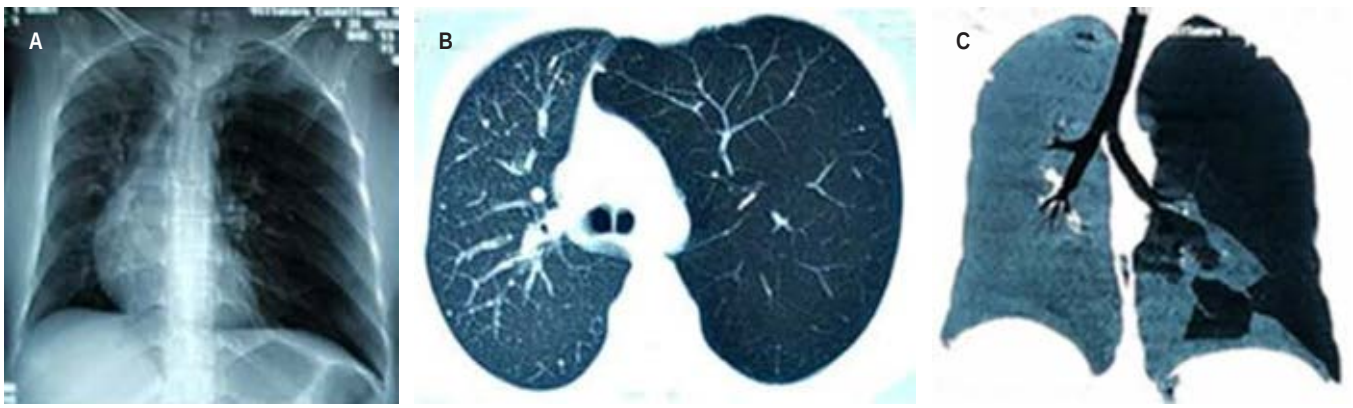

Figure 1: A) Chest X-ray. Left lung. Hyperlucent distended and mediastinal shift. **B)** Computed axial tomography. Axial section. Hyperlucent left lung, over distended. **C)** Digital reconstruction shows hypoperfusion.

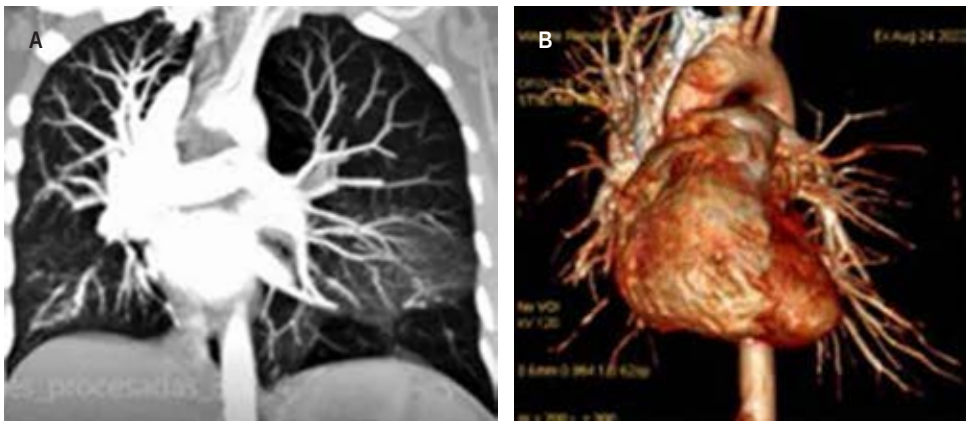

Figure 2:

A) CT angiography. Coronal section, non-uniform vascular distribution. **B)** Coronal section. Digital reconstruction. Superior lobar artery hypoplasia. Redistribution of flow.

pulmonary artery.[5,8] Therefore, diagnostic criteria for this syndrome require one of the following findings: a) unilateral loss of lung volume with hyperlucency demonstrated by chest radiography; b) unilateral reduction of vascularity on chest CT scan; and c) unilateral loss of perfusion on technetium-99m lung scan.[1,2]

Treatment is established according to symptoms and may vary from conservative management to surgical indications. Surgery is reserved for patients with recurrent lung infections that do not respond to conservative treatment or whose symptoms are not adequately controlled with optimal medical treatment. Surgical options include pneumonectomy, volume reduction surgery, lobectomy, or anatomic or non-anatomic segmentectomies,[1] performed by thoracotomy, videothoracoscopy or robotics according to preference, experience and availability of resources. Due to their clinically stable evolution, conservative treatment with outpatient follow-up was chosen.

CONCLUSIONS

Swyer James MacLeod syndrome is a rare entity that usually presents asymptomatic; its diagnosis is usually an incidental finding and treatment is conservative in most patients, with definite surgical indications.

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Conflict of interests: the authors declare that they have no conflict of interests.