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SWYER-JAMES SYNDROME IN A PATIENT ADMITTED FOR INVESTIGATION OF PULMONARY THROMBOEMBOLISM

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Abstract: Unilateral hyperlucent lung syndrome, initially described by Swyer-James-Macleod (SJMS) in 1953, is considered a post-infectious complication of bronchiolitis occurring in childhood and is characterized by airflow obstruction and a decrease in the number and diameter of ipsilateral peripheral pulmonary vessels. It is a relatively uncommon and complex disease. In the literature, small groups of patients with this syndrome have been described. This article refers to a male patient, 64 years old, smoker for 80 pack-years, admitted for investigation of pulmonary thromboembolism, after presenting with dyspnea, tachypnea and desaturation. After performing a chest tomography, cylindrical cystic bronchiectasis on the left was revealed, affecting segmental and subsegmental bronchi, notably in the lower lobe and lingula, associated with volumetric reduction of the left lung, with deviation of the ipsilateral mediastinum, as well as pulmonary reticulation. Transthoracic echocardiography and arterial tomography angiography of the chest were also performed, where pulmonary thromboembolism was ruled out and an abnormal image was visualized with a hypertransparent area in his left lung, drawing the attention of the team responsible for the case. SJMS can be confused with asthma or pulmonary embolism due to similar symptoms, and because of this, it can result in inappropriate therapy. This study aimed to examine the clinical and imaging spectrum of a patient diagnosed with Swyer-James-Macleod in adulthood. Therefore, a brief review of the relevant literature was carried out to better understand the subject.
Keywords: Bronchiectasis; Syndrome, Asthma; Pulmonary Embolism; Lung; Tomography; CT angiography; Masculine; Adult; Case report.

INTRODUCTION

Unilateral hyperlucent lung syndrome had its radiological characteristics first described in February 1952, by MacLeod, at a meeting of the British Thoracic Society in London, when nine cases of adult patients aged between 18 and 41 years who presented with unilateral hyperlucent lung were reported. to chest x-ray. However, this study was only published in 1954. A year earlier, in 1953, Swyer and James published the case of a six-year-old boy who had been followed for three years for episodes of bronchospasm and who had developed pneumonia at five weeks of age. life, at two and three years of age. The chest X-ray showed hypertransparency of the right lung and a considerable reduction in vascular markings. (CHLAPOUTAKIS et al., 2021)

Swyer-James-MacLeod Syndrome (SJMS) has as its most important pathological finding implicated in its etiology a developmental disorder in the cartilage of the lobar or segmental bronchus. This disease is also considered secondary to bronchiolitis obliterans acquired in childhood, resulting in obstruction of small airways and concomitant emphysema. Thus, due to hypoplasia of the pulmonary vessels that occurs secondary to chronic inflammation associated with bronchiectasis, this syndrome can present in a symptomatic or asymptomatic manner, where, in the latter, the diagnosis ends up being made in adulthood due to eventual findings on imaging exams. Furthermore, cases of repeated infections could result in obliterative bronchiolitis, which would gradually lead to distal air entrapment, airway distension and, eventually, pan-acinar emphysema. This way, the affected areas would become hypoventilated, with hypoxic vasoconstriction. (AKSOY, et al., 2015).

Thus, the classic hyperlucency seen in imaging studies of patients with this syndrome would be secondary to a decrease in blood

supply in the affected lung areas. SJMS has different therapeutic plans from conservative treatments to invasive interventions with the aim of reducing lung damage, which will be discussed throughout this case report. Its therapy is decided according to the clinical picture and severity of the affected lung. Furthermore, for all patients with this syndrome, the importance of an updated vaccination record is reinforced in order to avoid recurrent infections, especially against influenza and pneumococcus. (AKSOY, et al., 2015).

Therefore, in this case report we describe the case of: E.D.C, 64 years old, smoker, presenting unilateral pulmonary hyperlucency on imaging tests. His clinical presentation included tachypnea, dyspnea and desaturation, which led the team to primarily rule out pulmonary thromboembolism during hospital stay. Through imaging tests, Swyer-James Syndrome was suggested and confirmed. To establish this diagnosis, other pathologies that are mentioned as differential diagnoses of the syndrome were excluded, such as: tuberculosis, with negativity of the requested geneXpert; TEP; asthma; pneumatocele and pneumothorax. In order to evaluate ventilation changes and the possibility of reducing ventilation on the affected side, spirometry was also performed, which revealed (xxxxxxxxxxxxxx), the result of which can be seen in the figure. (AKSOY, et al., 2015)

This study was carried out because it is a low prevalence clinical entity with the aim of analyzing the clinical associated with imaging findings in SSJM, in addition to the impact on the quality of life of those affected. Furthermore, there was also the intention of collaborating with future analyzes on this syndrome, which could result in the development of better-defined diagnostic criteria, and, possibly, increase the number of diagnoses made, since its condition initial clinical presentation is nonspecific. (AKSOY, et al., 2015)

REPORT OF CASE

Male patient, 64 years old, white, referred from Ituverava hospital to Santa Casa de Misericórdia de Franca with a history of chronic obstructive pulmonary disease (COPD). The patient had a significant history of smoking, 80 years/pack. Since the age of 54, the patient has reported having dyspnea associated with coughing and expectoration, which is why he sought medical help at the Ituverava Basic Health Unit and was referred to a pulmonologist. Diagnosis of chronic obstructive pulmonary disease (COPD) was made during outpatient follow-up, but due to loss of segment, treatment for COPD was not carried out.

The patient states that on that occasion, around 10 years ago, he underwent spirometry, but that he does not have it and no longer remembers the result. During admission to the service, the patient presented tachypnea and dyspnea MMRC 4. Upon physical examination, he was in a regular general condition, with the presence of snoring and diffuse left-sided crepitations on lung auscultation, requiring a high-flow mask of 10l/min due to instability in the breathing pattern. and drop in saturation. Laboratory tests including blood gas analysis were carried out, all within normal limits. Aiming to exclude congenital lung malformations and evaluate central bronchial permeability, a computed tomography of the chest was performed, where cystic and cylindrical bronchiectasis on the left were visualized, affecting segmental and subsegmental bronchi, notably in the lower lobe and lingula, associated with volumetric reduction of the left lung with deviation of ipsilateral mediastinum, as well as pulmonary reticulate and pleural thickening,.probably. sequelae, findings seen below (figures 1 and 2).

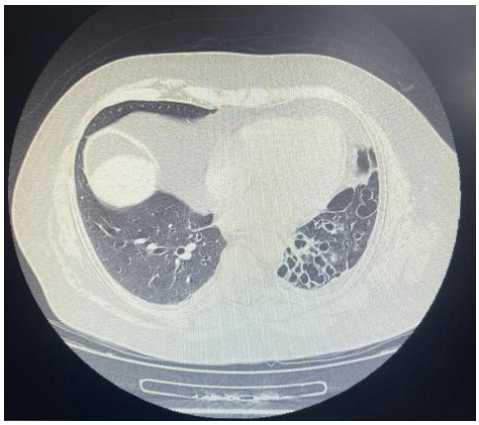

Figure 1: Chest tomography showing areas of cystic and cylindrical bronchiectasis on the left.

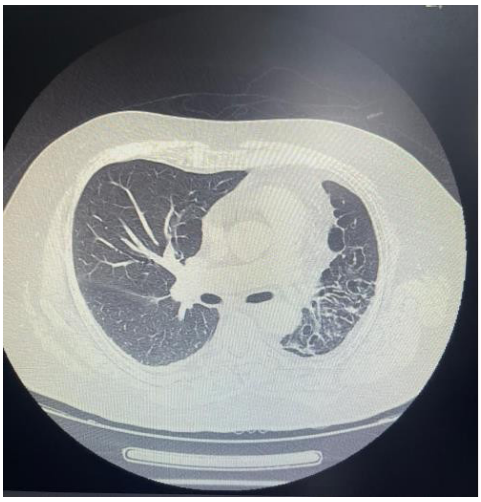

Figure 2: Chest tomography showing involvement of segmental and subsegmental bronchi and volumetric reduction of the left lung.

Chest CT angiography and transthoracic echocardiography were also performed to investigate the possibility of pulmonary venous thrombosis. In echocardiography, aorta, ascending and descending aorta had preserved dimensions, with no images suggestive of thrombi, shunt or intracavitary vegetation, thus ruling out PTE. (figure 3).

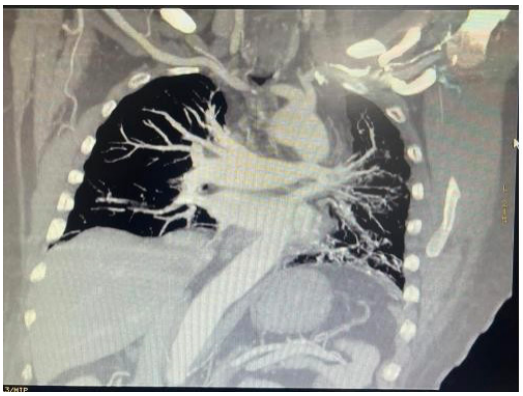

Figure 3. Thoracic CT angiography showing a marked reduction in the caliber of the pulmonary arterial vessels on the left.

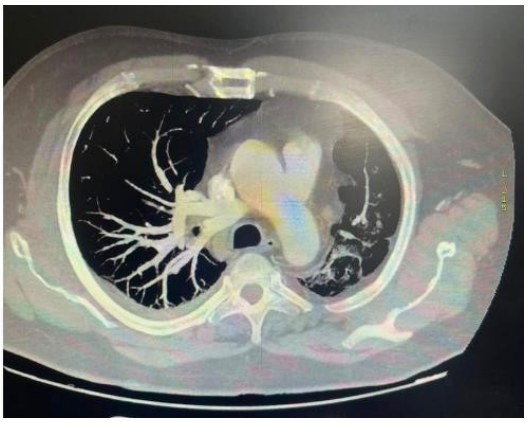

Figure 4. Thoracic CT angiography showing hyperlucency of the affected lung in relation to the contralateral one.

On thoracic tomography angiography, no filling defects were observed by the contrast medium.

in the pulmonary arteries and their main, larger proximal branches. However, an increase in the caliber of the right pulmonary artery and a marked reduction in the caliber of the pulmonary arterial vessels on the left were seen, in addition to hyperlucency of the affected lung in relation to the contralateral one. These radiological features strongly suggested the diagnosis of Swyer-James-MacLeod syndrome (figure 4).

When questioned, the patient stated that he had polio and that he had a history of repeated pneumonia during childhood. Furthermore,

he was diagnosed with COPD in 2013, but said he had never been treated for the disease. In addition, it presents as associated diseases high blood pressure and dyslipidemia. His family history was negative for asthma.

During clinical follow-up at the service, the patient's dyspnea and clinical condition improved and conservative treatment was chosen.

DISCUSSION

Swyer-James-MacLeod syndrome is defined as bronchiolitis obliterans following pneumonia in early childhood. Pulmonary infection at this time leads to the destruction of the bronchial epithelium and elastic tissue. Furthermore, the presence of submucosal fibrosis leads to luminal narrowing and irregularity in the airways and also to hypoplasia in the vascular structures and consequent hyperinflation with panacinar emphysematous changes on the distal surface of the involved bronchiole. Thus, the loss of ciliary motility increases the tendency to secondary infections, which are slightly common in these patients. (TORTAJADA et al., 2004)

Swyer-James-MacLeod syndrome is an uncommon disorder and its diagnosis is usually based on imaging and clinical examinations. This syndrome is characterized by the presence of constrictive bronchiolitis with dilation and destruction of alveolar structures, leading to significant air trapping associated with a decrease in the number and diameter of ipsilateral peripheral pulmonary vessels, which was clearly seen in the case report described. Although it classically involves an entire lung, the disorder can be lobar or segmental. (CONTI et al., 2021)

Swyer-James-MacLeod syndrome is considered a post-infectious type of bronchiolitis obliterans. Pulmonary infection with adenovirus, respiratory syncytial virus, influenza A, measles, tuberculosis, mycoplasma pneumoniae and Bordetella pertussis has been reported as possibly responsible for the development of the syndrome. As a result, updating the vaccination card is recommended for all patients. (YOLCU et al., 2018)

It has been reported that some other genetic or environmental components may be responsible for the development of this syndrome, but these are still under study. The evolution of the disease is possibly influenced by the presence or absence of bronchiectasis; patients with no or few bronchiectasis have mild respiratory symptoms and a spontaneous tendency to improve, while those with bronchiectasis have repeated episodes of pneumonia and complications such as lung abscesses and may require surgical treatment. (DIRWEESH et al., 2017).

The literature, over the years and using the most accurate means for investigating lung lesions, has made important contributions regarding unilateral or bilateral lung involvement in Swyer James-MacLeod syndrome. In most cases suffering from SJMS, therapies are mainly conservative with the use of antibiotic therapy, bronchodilators and respiratory physiotherapy with drainage posture of secretions can be useful, as was the case with the patient described. Therefore, in this case, lung re-expansion was achieved without any surgical intervention and the patient showed no signs of recurrence and was discharged from hospital. (PERARO, et al., 2014)

Surgery should be considered as the last treatment option and It is only indicated when all other therapies are ineffective or if there is the presence of associated spontaneous pneumothorax. The clinical characteristics of our case are in accordance with current literature. Radiological examinations showed probable results of previous pulmonary

infections and the patient confirmed such conditions in childhood and this reaffirms that the pathogenesis of the syndrome is attributable to airway inflammation. (SELIMOGLU, et al., 2014)

Furthermore, one must also take into consideration, the diagnosis of COPD 10 years ago and the lack of drug treatment since diagnosis due to segment loss seen by a pulmonologist. Thus, the present study describes an interesting case where the patient's clinical condition suggested pulmonary thromboembolism and, after discarding, suggesting and confirming Swyer-James MacLeod Syndrome, which should always be suspected when faced with a hyperlucent lung with the presence of bronchiectasis. in imaging exams and in patients with persistence of respiratory symptoms adequately treated but without improvement; avoiding delay in diagnosis and possible unfavorable prognosis. (PERARO, et al., 2014)

CONSENT FOR PUBLICATION

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent was given to the patient and is also available for review if necessary.

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