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Case Report

Swyer-James-MacLeod syndrome presenting as spontaneous pneumothorax in an adult: Case report and review of literature ☆☆☆★

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ABSTRACT

Swyer-James-MacLeod Syndrome (SJMS) is an uncommon, emphysematous disease characterized by obliteration of the small bronchioles, hypoplasia/or absence of pulmonary artery and peripheral vascular bed. It is most commonly diagnosed in childhood. Patients are often asymptomatic or they could suffer from symptoms of recurrent pulmonary infections. Spontaneous pneumothorax is a rare presentation of this syndrome.

We report a case of a 42-year old female patient presented at our Emergency Department with complaints of dyspnea and pleuritic chest pain with a diagnosis of spontaneous pneumothorax in emphysematous disease.

The diagnosis of SJMS is usually based on imaging and clinical findings rather than on the results of pathologic examination; indeed, asymptomatic adult patients with SJMS are often diagnosed after a chest radiograph obtained for another reason. High-resolution computed tomography (HRCT) seems to be the most appropriate technique for the diagnosis.

SJMS can be associated with spontaneous pneumothorax which represent an emergency condition due to the underlying pathological disease. The surgical treatment of the affected lung should be considered when conservative approach is ineffective.

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Abbreviations: SJMS, Swyer-James-MacLeod syndrome.

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Introduction

Swyer-James MacLeod syndrome, first described in 1953 [1], is a rare radiologic entity characterized by hyperlucency of one or more lobes or of the entire lung, hypoplasia/or absence of pulmonary artery and peripheral vascular bed [2,3], normal bronchial system on bronchoscopy, as congenital lobar or segmental emphysema that unilaterally or bilaterally involves a lobe or segment (occasionally the entire lung). The reported incidence is 0.01% per 17,450 chest radiographs according to Gaensler et al. [4].

The most important pathologic finding implicated for its etiology was emphasized to be a developmental disorder in the cartilage of the lobar or segmental bronchus [5], the syndrome is also thought to be secondary to bronchiolitis obliterans acquired in infancy or childhood resulting in obstruction of small airways and concomitant emphysema. Hypoplasia of lung vessels is also considered to be secondary to chronic inflammation [6]. However, unilateral hyperlucency is caused by numerous and various etiologic processes. These may be categorized in 4 groups: bronchial, parenchymal, vascular, and postsurgical. Common symptoms can include chronic cough, dyspnea, chest pain, wheezing, cardiac symptoms caused by the mediastinal shift, recurrent bronchitis, and bronchopneumonia, hemoptysis; rarely do they present with symptoms of spontaneous pneumothorax due to rupture of an emphysematous bulla secondary to the inflammatory process. Many subjects are asymptomatic and the syndrome is often diagnosed during routine chest radiographs [7,5,8]. Disappearance of peripheral vascular structures on the CT-angiography and perfusion defect on the ventilation-perfusion scintigraphy, as additional investigations, would confirm the diagnosis [8]. The most distinctive feature compared with emphysema is the absence of chronic obstructive pathology in the small airways [5,9]. A differential diagnosis should consider pseudounilateral pulmonary hyperlucency such as absence or atrophy of the pectoral muscles, mastectomy, technical errors in angiography, total pneumothorax, gastrointestinal herniation, and congenital pulmonary arterial agenesis.

Patients suffering from SJMS may be asymptomatic for many years or suffer from recurrent episodes of pulmonary infections. Generally, the majority of patients with SJMS are treated conservatively. Spontaneous pneumothorax is considered an emergency condition particularly among patients with SJMS due to the underlying pathologic disease. Surgical treatment is considered only in case of increased incidence of recurrent lung infection and compression atelectasis caused by the emphysematous lung on the normal lung, and dyspnea caused by shunt. Appropriate treatment should be pneumonectomy and lobectomy if hyperinflation involves one lung or one entire lobe.

Case report

We present a case of a 42-year old patient presented in November 2019 (before the Covid-19 pandemic) at our Emergency Department with complaints of dyspnea and pleuritic

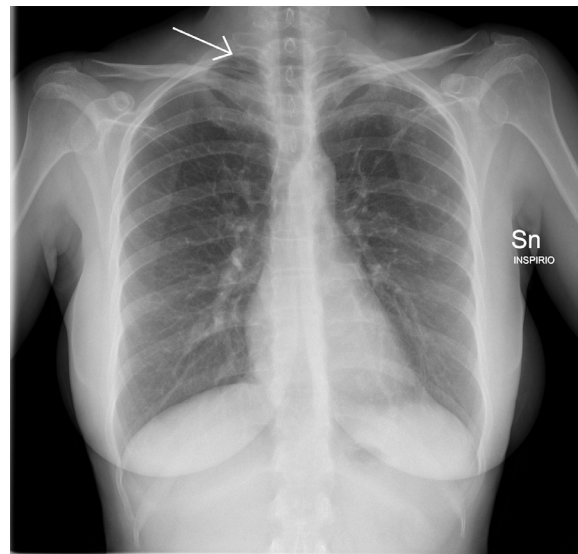


Fig. 1 – Chest X-ray at the admission shows a 2-cm right pneumothorax (arrow).

chest pain on the right side. She hadn't history of childhood respiratory problems or any significant respiratory symptoms during her adult nor smoking habit. The patient came from the western Balkans where the population is exposed to alarming levels of air pollution, which have serious repercussions on the health conditions of citizens: these are mainly SO₂, NO_x and particulate matter. Emergency chest X-ray showed a 2 cm pneumothorax on the right apical pulmonary field (Fig. 1). Her vital signs were: blood oxygen saturation 99%, blood arterial pressure was 130/85 mmHg, heart rate 86 bpm. The routine blood tests were unremarkable. Based on vital signs and clinical presentation we decided for a conservative treatment more than pleural drainage.

During her hospital stay we observed radiological and clinical improvement so the patient was discharged after a day of observation; lung expansion on chest X-ray was achieved spontaneously.

After a month we followed up the patient with a contrast-enhanced chest CT scan (Fig. 2) that showed areas of unilateral emphysema and cystic bronchiectasis of the left upper and lower lobe: in the left apical lobe was detected an emphysematous bubble of about 6 mm with thickened walls (maximum axial thickness of about 5 mm) attached to the sub-costal pleura, at the left lower lobe CT scan showed the presence of focal cystic bronchiectasis with appearance of endoluminal secretion (Fig. 3). This radiological features strongly suggested the diagnosis of Swyer-James-MacLeod syndrome (SJMS). The radiological work up also included a pulmonary scintigraphy with ^{99m}Tc that showed no mismatch of ventilation/perfusion (V/Q) ratio (Fig. 4), a result consistent with SJMS diagnosis.

No abnormalities were detected on spirometry (Fig. 5); cardiac function on transthoracic echocardiography was physiological and didn't reveal any signs of pulmonary hypertension.

Considering the clinical presentation and negative radiological investigations for any significant functional alter-

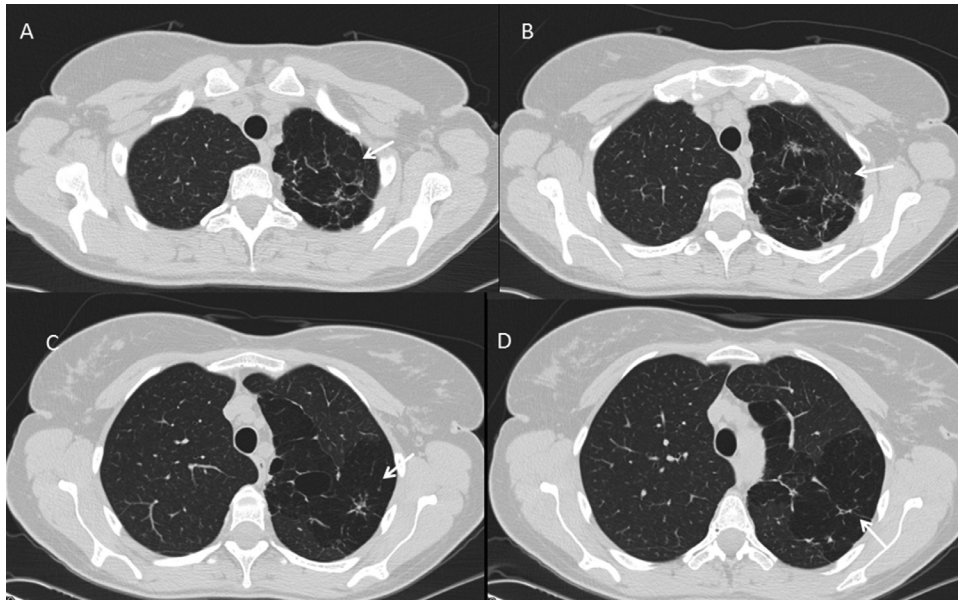


Fig. 2 – Chest CT scan: areas of unilateral emphysema and cystic bronchiectasis of the left upper (A,B) and lower lobes (C,D) (arrows).



Fig. 3 – CT scan appearance on coronal section: arrows indicate emphysematous bubbles with thickened walls.

ations, we decided for an outpatient follow up without any further indications.

Discussion

SJMS is an uncommon disorder. The diagnosis is usually based on imaging and clinical findings. SJMS is characterized by the presence of constrictive bronchiolitis with dilatation and destruction of alveolar structures, leading to signif-



Fig. 4 – Pulmonary scintigraphy shows no mismatch of ventilation/perfusion of the lungs.

icant air trapping associated with decreased number and diameter of ipsilateral peripheral pulmonary vessels. Although classically involving an entire lung, the disorder can be lobar or segmental. Lung oligoemia in SJMS is a combination of diminished pulmonary capillary beds secondary to inter-alveolar septal fibrosis, mechanical resistance by overinflated terminal airspaces and reflex vasoconstriction to minimize the ventilation-perfusion mismatch [10]. The combination of air trapping and oligoemia leads to the translucent appearance of the affected lung on chest radiograph [11]. The bronchial system is unobstructed. Usually patients present with productive cough, shortness of breath, and dyspnea. Patients with little or no bronchiectasis have minor symp-

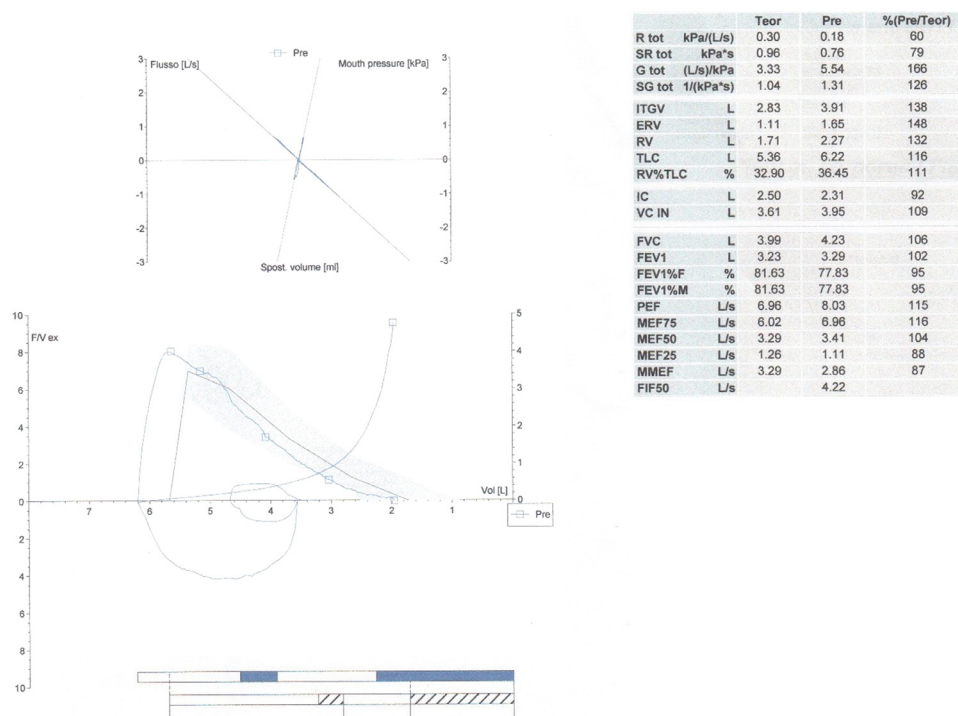


Fig. 5 – Normal spirometry.

toms or are asymptomatic and may remain undiagnosed until adulthood. Moreover, adult patients with SJMS are often diagnosed after a chest radiograph obtained for another reason [12]. The diagnosis of SJMS requires the exclusion of other causes of unilateral hypertranslucency such as those related to congenital bronchial and/or vascular abnormalities. Ventilation/perfusion (V/Q) ratio is very helpful in determining the extent of the disease and correlates well with high-resolution computed tomography (HRCT) which seems to be the most appropriate technique. In the majority of cases suffering from SJMS, therapies are primarily conservative. Antibiotic therapy, use of bronchodilators and chest physiotherapy with postural drainage of the secretions may be useful. Surgery should be considered as the last treatment option and it is only indicated when all other therapies are ineffective [14]. According to symptoms, patients with SJMS can be classified into the following categories regarding surgical indications: patients presenting with recurrent infections not improving with conservative support [13–16]; patients with clinical deterioration and resulting failure to thrive [16–19]; and finally those presenting with spontaneous or recurrent pneumothorax [19,20].

The clinical features of our case is in accordance with the current literature although a diagnosis of spontaneous pneumothorax caused by SJMS in adults is infrequent.

Although radiological exams had showed probable outcomes of previous pulmonary infections, the patient has always denied such conditions in childhood; this confirms that the pathogenesis of the syndrome is attributable to airways inflammation.

The clinical course and prognosis are influenced by the presence of underlying bronchiectasis and treatment is gen-

erally conservative; when associated with spontaneous pneumothorax, surgical therapy should be considered only when there is no clinical or radiologic improvement after conservative treatment; in this case the pulmonary re-expansion was achieved without any intervention and the patient showed no signs of recurrence to date.

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 is available for review by the Editor-in-Chief of this journal.

Availability of Data and Materials

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

Authors' contributions

LC conceived the case report. LC and GP acquired the data, drafted and wrote the article. CG, RD, MDD and PC critically revised the article. All authors read and approved the final manuscript.

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