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CAZURI CLINICE

Swyer James Syndrome: a case report

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REZUMAT

Sindromul Swyer James: prezentare de caz

Sindromul Swyer James este o boală rară caracterizată prin bronșiolită și pneumonie ce apar timpuriu în viață.

O fetiță de 2 ani a fost referită clinicii noastre acuzând wheezing, tuse cronică și febră prezente încă de la naștere. Diagnosticul de sindrom Swyer James a fost confirmat pe baza semnelor clinice, radiografiei și tomografiei computerizate toracice. Copilul primește tratament suportiv și simptomele respiratorii s-au ameliorat.

Cuvinte cheie: *Sindrom Swyer James, hipertransparență pulmonară, bronșiolită, pneumonie*

ABSTRACT

Swyer James syndrome is a rare condition clinically characterized by bronchiolitis and pneumonitis early in life.

A 2 year old girl referred to us with complaints of wheezing, chronic cough and fever since birth time. Based to her clinical symptoms and radiograph and CT scan findings, diagnosis of Swyer James syndrome was confirmed. She is under supportive treatment and her respiratory symptoms have been improved.

Key words: *Swyer James syndrome, hyperlucency of the lung, bronchiolitis, pneumonitis*

Introduction

Swyer James syndrome, also called Swyer James Macleod (SJMs) is a rare condition clinically characterized by bronchiolitis and pneumonitis early in life[1].

As the lung is expected to grow by alveolar development in the first 2-8 years of life, the post infectious obliterative bronchiolitis causes hypoplasia as a result of diminished vascularisation and lack of progressive growth and alveolar development of the lung[2].

The characteristic radiographic feature is pulmonary hyperlucency.

Swyer James syndrome presents as a complication of infection. According to previous reports, organisms causing infection resulting in SJMs are mostly Morbilivirus (measles virus), Bordetella pertussis, and adenovirus[3,4].

Following these infections SJMs produces some effects as: areas of lung hyperlucency, air trapping, and bronchial/bronchiolar disease with wheezing.

Case Report

A 2 year old female child presented to us with wheezing and recurrent pneumonia from birth time. Her weight and height were under 3rd percentile (Weight: 10 kg Height: 81cm). On physical examination generalized wheezing was detected in both lungs while the rest of physical examinations were unremarkable.

Chest radiograph revealed hyperlucency of left lung in comparison to right lung (Figure 1), feature that was also present on the CT scan of the chest (Figure 2, 3).

Bronchoscopy was performed to detect any foreign body aspiration in one of the main bronchi, but the result was normal.

Due to the patients symptoms (failure to thrive, nausea, vomiting and wheezing), a sonography was carried out to rule out an abdominal problem causing the clinical features.

The sonography result was normal, so subsequently Barium swallow was performed and the results showed a severe gastro esophageal reflux.

She received antibiotics for her pneumonia and, she also received bronchodilator. Protein Pomp Inhibitor (PPI) was prescribed for her gastro esophagial reflux. The patient is on antibiotic prophylaxis and she is under regular follow up.

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Figure 1.
Chest X-ray: Trachea deviation to the left is seen and left lung is more lucent in comparison to right lung.

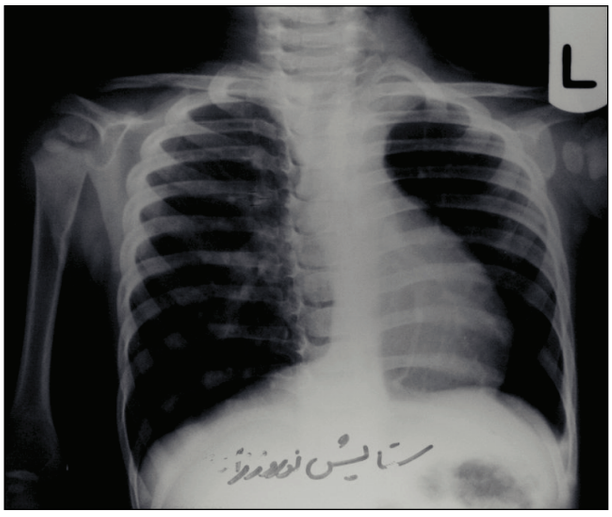


Figure 2.
Thoracic CT scan: Left lung has a smaller volume in comparison to right lung.

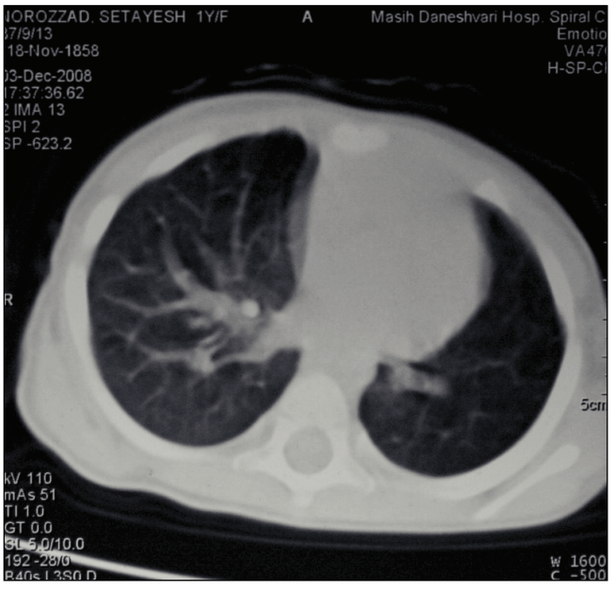
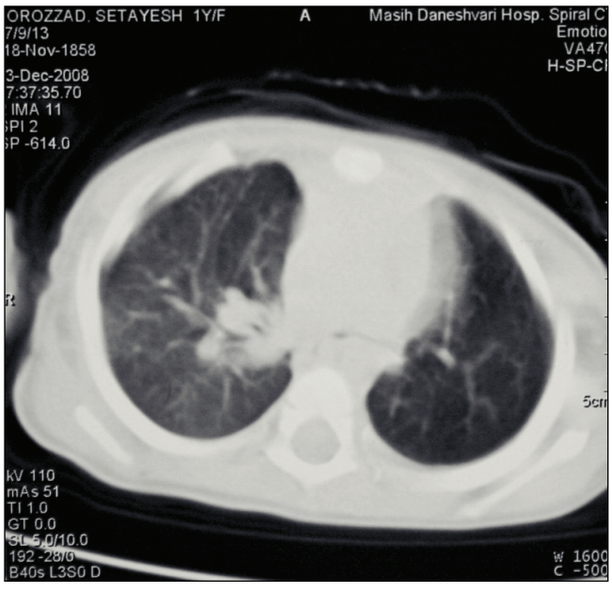


Figure 3.
Thoracic CT scan: left lung is more lucent in comparison to right lung.



Discussions

Swyer James is an uncommon syndrome which was first described in 1953 by Swyer and James and followed by Macleod in 1954[5, 6, 7].

According to a previous report the prevalence of SJMs is 0.01% in a survey on 17,450 chest radiographs[6].

SJMs is an acquired disease secondary to viral bronchiolitis and pneumonitis in early childhood[1].

The responsible organisms for respiratory infections in these cases are mostly measles, pertussis and adenovirus.

Normal development of the alveolar buds is affected by the damages of terminal bronchioles in early childhood caused by these organisms[8].

Onset of symptoms usually occurs during early childhood, although patients who have little or no bronchiectasis may be asymptomatic and might be miss diagnosed until adulthood or may be diagnosed incidentally by a chest radiograph taken for other indications, showing localised hyperlucency of the lung.

Our case developed pneumonitis since birth time. Productive cough, shortness of breath, dyspnea and wheezing, recurrent respiratory infections and other respiratory symptoms may be the manifestations of this syndrome secondary to bronchiectasis[3,6,9,10]. Our patient had similar clinical feature to those described.

Diagnosis of Swyer James syndrome in our patient was based on clinical manifestations and radiological signs (hyperlucent lung).

SJMs should be differentiated from congenital anomalies of airway or the lungs, foreign body aspiration, bronchopul-

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monary dysplasia as the radiological feature of these anomalies may be similar to radiological appearance of SJMs[6,8]. Bronchoscopy was performed for our patient to detect foreign body aspiration or abnormal division of the bronchi, and the result was normal.

Although chest radiograph is the first diagnostic tool for SJMs, CT scan remains the preferred tool for establishing the diagnosis of SJMs[11].

High resolution CT scan has largely replaced the invasive diagnostic procedures such as pulmonary angiography, radionuclide ventilation/perfusion scintigraphy and bronchography.

Following up the patient for preventing the respiratory infections is necessary in the management of this syndrome[12,13]. Our patient is on antibiotic prophylaxis to prevent the infections and she is under regular follow up.

Surgical interventions such as lobectomy are uncommonly indicated, unless there is uncontrolled infection in a segment of the lungs.

Lung resection in patient with SJMs is reported in a few published literatures[3,6].

Consequently, regular follow up for detecting and preventing bronchiectasis and treatment of respiratory infections are the main aim of SJMs management.

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