Case Report

Swyer-James-Macleod Syndrome: A Rare Entity

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Abstract

Swyer-James-MacLeod syndrome is a radiological entity characterised by hyperlucency of one or more lobes or of the entire lung associated with decreased number and diameter of ipsilateral peripheral pulmonary vessels resulting in difficult visibility of the arterial network, and unobstructed bronchial system.

We report the case of a 12-year-old girl who presented with chief complaint of pain in the chest. Chest radiograph (postero-anterior view) showed hyperlucent left lung field with increased lung volume and shift of the mediastinum to the right side. Contrast-enhanced computed tomography (CECT) of the chest showed hyperlucency and diminished vascularity in the upper lobe and lingula of left lung with hyperinflation of the pulmonary parenchyma. On the basis of clinical and radiological findings, the patient was diagnosed to have Swyer-James-MacLeod syndrome.

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Key words: Swyer-James-MacLeod syndrome, Unilateral hyperlucency

Introduction

Swyer-James-Macleod syndrome or unilateral hyperlucent lung syndrome is an uncommon disease associated with post-infectious bronchiolitis obliterans that follows viral bronchiolitis and pneumonitis occurring in childhood.[1]

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

Typically, this disorder is diagnosed in childhood after an evaluation for recurrent respiratory infections but sometimes patients who have little or no sequelae bronchiectasis have minor symptoms or are asymptomatic and the diagnosis may be missed until adulthood.[3] In this case, the child presented with the unusual complaint of pain in the chest and was diagnosed as a case of Swyer-James-Macleod syndrome.

Case Report

A 12-year-old girl presented with a complaint of continuous, diffuse, non-radiating, dull pain in the left side of the chest for the past three months. There was no history of cough or expectoration or trauma. There was a history of occurrence of repeated episodes of chest infections since the past 3-4 years. On general physical examination, the child was of average built with no chest wall or spinal defects and no limitation of shoulder movements on either side; pulse 76 beats per minute, respirations 20 breaths per minute and oxygen saturation of 98% at room air on pulse oximetry. Respiratory system examination revealed hyper-resonant left chest with decreased breath sounds.

Chest radiograph (postero-anterior view) showed hyperlucent left lung field with increased lung volume and shift of the mediastinum to the right side (Figure 1). Pulmonary function testing revealed forced vital capacity (FVC) 72%, forced expiratory volume in first second (FEV_1) 66% and ratio of FEV_1/FVC 95%.

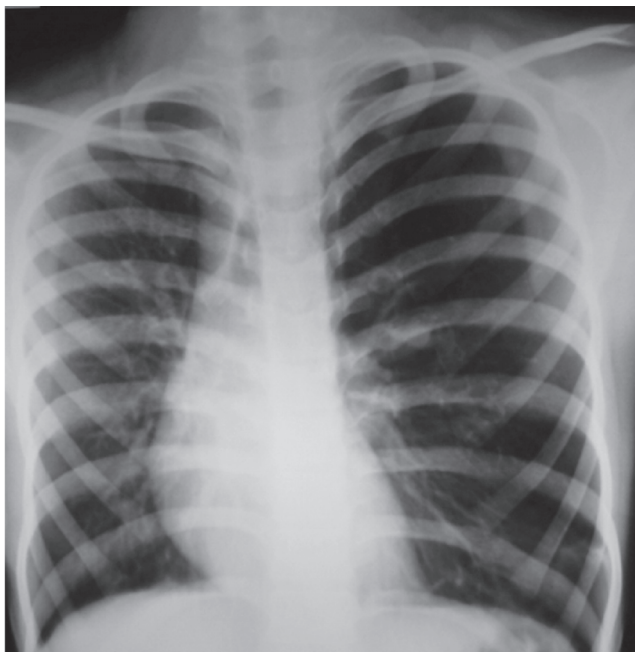

Figure1. Chest radiograph (postero-anterior view) showing hyperlucent left lung field with increased lung volume and shift of the mediastinum to the right side.

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Echocardiography was normal. Contrast-enhanced computed tomography (CECT) of the chest (Figure 2) showed hyperlucency and diminished vascularity in the upper lobe and lingula of the left lung with hyperinflation of the pulmonary parenchyma. Typical mosaic pattern was seen in sections taken in prone position (Figure 3). No endobronchial lesion, pulmonary artery aberrations or bronchiectasis were detected.

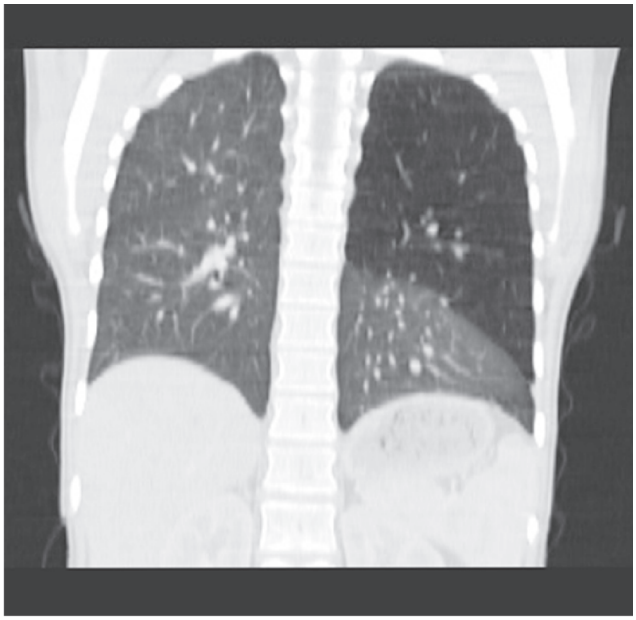

Figure 2. Computed tomography of chest (lung window, coronal section) showing hyperlucency and diminished vascularity in left upper lobe and lingula with pulmonary parenchymal hyperinflation.

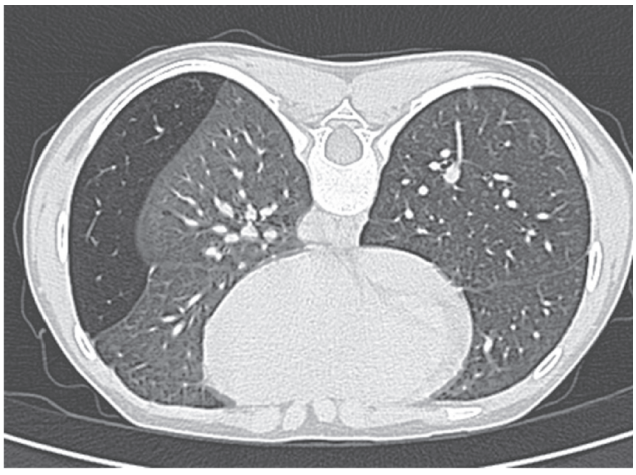

Figure 3. Computed tomography of chest (lung window) showing a typical mosaic pattern seen in prone position.

On the basis of clinical and radiological findings, the patient was diagnosed to have a case of Swyer-James-Macleod syndrome. The patient is symptom free on conservative management and is doing well on follow-up.

Discussion

Swyer-James-Macleod syndrome is considered to be a rare and complex disease characterised by unilateral hyperlucency of a part or the entire lung which was first described in 1953 by Swyer and James and by Macleod in 1954.[4] In a study involving 17,450 chest radiographs, the prevalence of this rarely encountered entity was found to be 0.001%.[5] Due to unknown factors, it usually involves the left lung.[6] It is a form of obliterative bronchiolitis with concomitant vasculitis following injury to immature lungs during the first 8 years of life. It usually follows infections with organisms like adenovirus, measles or pertussis. Both small bronchi and bronchioles are affected and the lung with abnormal airways remains inflated by collateral airdrift.[7] This damage during the early childhood prevents normal development of the alveolar ducts. Airways develop submucosal fibrosis leading to luminal irregularity and occlusion. Pulmonary vasculature is hypoplastic while the lung distal to diseased bronchioles become hyperinflated and sometimes panacinar emphysematous changes develop.

Swyer-James-Macleod syndrome presents with variable clinical features. Patients can be completely asymptomatic with hyperlucent lung-field, being an incidental finding on the chest radiograph taken for other indications. Alternatively, these may have chronic cough, haemoptysis, exertional dyspnoea, recurrent chest infections or other respiratory symptoms secondary to bronchiectasis.[8] In this case, the patient presented with second episode of an unusual and not earlier reported complaint of pain in the left side of the chest. The characteristic radiographic findings in these patients include unilateral hyperlucent lung along with decreased bronchovascular markings, a small hilar shadow and slight displacement of the mediastinum to the affected side. Rarely, the mediastinum is shifted to the contralateral side because of the long-standing nature of the illness and severe emphysematous changes in the left lung[9] as appreciated in our case. High resolution computed tomography (HRCT) has now largely replaced the more sophisticated and invasive diagnostic procedures, such as pulmonary angiography, radionuclide lung ventilation-perfusion scintigraphy and bronchography as the choice of investigation for confirming the presence of Swyer-James-Macleod syndrome.[8] With the use of HRCT, hyperlucency, anatomy of lung parenchyma and pulmonary vessels can be better defined. The HRCT findings of Swyer-James-Macleod syndrome include patchy areas of low attenuation and hypovascular areas interspersed with areas of normal attenuation. This uneven perfusion due to hypovascularity gives a typical mosaic pattern on HRCT which is more evident when section taken in prone position. Other changes on HRCT may include bronchiectasis, atelectasis and scarring[10,11] which were not evident in our case on HRCT chest and this may explain her apparent lack of symptoms for a long time till she complained of pain in

the chest, an unusual complaint in relation to this syndrome.

Bronchoscopy to visualise the airways for any foreign body or stricture was not performed in the present case as patient did not give consent for the same. Other than Swyer-James-Macleod syndrome, "unilateral hyperlucency" on the chest radiograph can be a feature of several other diseases, such as congenital pulmonary artery agenesis / hypoplasia or acquired stenosis or compression of main pulmonary vessels and certain parenchymal disorders including congenital lobar emphysema, bronchogenic cyst, bronchiectasis with air trapping, emphysematous bulla and emphysema.[11] The HRCT also helps in ruling out other causes of unilateral hyperlucent lung.[12] The aim of management of Swyer-James-Macleod syndrome is to prevent and treat intercurrent respiratory infections. Surgical interventions, namely lobectomy, pneumonectomy is seldom required except when there is an uncontrolled infection of the diseased lung segment.[12] Our patient is under regular follow-up and without any symptoms. Swyer-James-Macleod syndrome is a rare disorder; however, increased awareness is necessary for recognising and making prompt diagnosis and management of this disorder.

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