Classical Imaging Triad in a Very Young Child with Swyer-James Syndrome

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Abstract Swyer-James syndrome (SJS) or Swyer-James-MacLeod syndrome is a rare disorder characterized by roentgenographical hyperlucency of one lung, lobe, or part of a lobe. As originally described, it shows a unilateral small lung with hyperlucency and air trapping on radiographs. It is postinfectious obliterative bronchiolitis due to childhood respiratory infection. In SJS, the involved lung or portion of the lung does not grow normally. The characteristic radiologic appearance is that of pulmonary hyperlucency on chest X-ray/CT caused by over-distention of the alveoli in conjunction with diminished arterial flow detected on Tc-99 m MAA pulmonary perfusion scan. Radionuclide pulmonary ventilation studies are considered difficult to perform in very young children as patient co-operation is crucial to achieve adequate tracer entry into the lungs. We present here the case of a female child aged 2 years, who presented with chronic productive cough and fever off and on for 1.5 years. She had classical features on imaging and markedly diminished ventilation on Tc-99 m DTPA aerosol study. Our case highlights the typical findings on ventilation-perfusion scan (V/Q scan) and suggests that it is possible to

conduct a satisfactory ventilation study with Tc-99 m DTPA aerosol even in very young children.

Keywords V/Q scan · Swyer-James syndrome · Very young child

Introduction

Radionuclide pulmonary ventilation studies are considered difficult to perform in very young children as patient co-operation is crucial to achieve adequate tracer entry into the lungs. Swyer-James syndrome (SJS) or Swyer-James-MacLeod syndrome is a rare disorder characterized by roentgenographical hyperlucency of one lung, lobe, or part of a lobe. As originally described, it shows a unilateral small lung with hyperlucency and air trapping on radiographs. It is postinfectious obliterative bronchiolitis due to childhood respiratory infection. SJS seems to be an acquired disease that develops after viral bronchiolitis and/or viral pneumonia in early childhood and is one of the differential diagnoses of a hyperlucent hemithorax [1–4]. We present here the case of a female child aged 2 years, who was suspected to have SJS, which was confirmed on a ventilation-perfusion (V/Q) scan.

Case Report

A 2-year-old female child was referred to the pediatrician with history of chronic productive cough, and fever off and on for the past 1.5 years. There was no history of foreign body inhalation. On examination her vitals were stable, but she had tachypnoea. Chest examination showed bilateral inspiratory and expiratory wheeze and marginally

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diminished air entry on the left side. The chest X-ray revealed a hyperlucent left hemithorax with reduced bronchovascular markings (Fig. 1a). A contrast-enhanced chest tomography (CECT) corroborated these findings in a hyperinflated left lung. There was no evidence of bronchiectasis (Fig. 1b). Fiberoptic flexible bronchoscopy was performed to rule out the presence of any foreign body/intraluminal growth or extrinsic airway compression, and it was normal. With a strong suspicion of SJS, Tc-99 m MAA pulmonary perfusion scan was performed with 74 MBq (2 mCi) activity. It showed diffusely decreased radiotracer concentration in the entire left lung without any focal defect.

Tc-99 m DTPA aerosol lung ventilation scan was performed after 48 h and showed markedly reduced aerosol

deposition in the left lung while that in the right lung was normal. Some tracer deposition was noted in the region of the bilateral main bronchi (Fig. 2).

Discussion

SJS is considered to be a relatively uncommon and complex disease characterized by unilateral hyperlucency of a part of or the entire lung; it was first described in 1953 by Swyer and James [2]. It is considered to be an acquired disease usually secondary to viral but sometimes bacterial bronchiolitis and pneumonitis in childhood. It is associated with paramyxovirus,

Fig. 1 Chest radiograph (a) and CT (b) show hyperlucent left lung field with reduced bronchovascular markings

Fig. 2 V/Q scan showing markedly decreased ventilation and perfusion to the entire left lung

morbillivirus, *Bordetella pertussis*, *Mycobacterium tuberculosis*, *Mycoplasma pneumoniae*, influenza A, and adenovirus types 3, 7, and 21 [1]. In SJS, the involved lung or portion of the lung does not grow normally. The fibrosis and obstruction of the terminal and respiratory bronchioles, likely caused by respiratory infections in early childhood, prevent normal development of the alveolar bud [3]. The characteristic radiologic appearance is that of pulmonary hyperlucency on chest X-ray/CT caused by over-distension of the alveoli in conjunction with diminished arterial flow detected on Tc-99 m MAA pulmonary perfusion scan/CT angiography. On ventilation perfusion lung scanning, diminished activity of the affected lung is seen with perfusion scanning, and decreased gas exchange is seen during the ventilatory phase. The lung perfusion deficit seen in patients with SJS occurs because the peripheral branches of the pulmonary vessels have not developed normally, and vasculature is arrested at the stage at which the causative infection occurred [4–7]. Any disorder involving distal airway obstruction (bronchiolitis obliterans, foreign body inhalation, congenital lobar emphysema) is a differential diagnosis for SJS.

Our patient had a classical triad of hyperlucent/hyperinflated lung on X-ray/CT chest, diffusely decreased perfusion, and diffusely decreased ventilation in the entire left lung. She had recurrent episodes of pulmonary infections in her childhood. Clinically, patients usually have productive cough, shortness of breath, and dyspnea on exertion, sometimes with hemoptysis. Some patients, who have little or no associated sequelae are not diagnosed until they are adults. SJS treatment includes the early control of lung infections as well as influenza and pneumococcal vaccinations [8, 9].

We have provided images of ventilation perfusion scan, chest radiograph, and CT in this report. V/Q scan is considered difficult to do in very young children in view of the patient co-operation required for the ventilation study. Many attempts at the ventilation scan in children of even 5 years of age result in central deposition of radiotracer and poor concentration in the lungs.

Our case highlights the typical findings on ventilation-perfusion (V/Q) scan and suggest that it is possible to conduct a satisfactory ventilation study with Tc-99 m DTPA aerosol even in very young children.

Conflict of Interest No conflict of interest exists with respect to this manuscript for all the authors.

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