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Case Reports

Three Children with Swyer-James/Macleod Syndrome

SC ***SIT***, QU ***LEE***, MC ***CHIU***

Abstract Swyer-James/Macleod syndrome (SJMS) is an uncommon disease with the characteristic radiological feature of "unilateral hyperlucency" due to loss of pulmonary vasculature and air trapping. SJMS is considered to be a form of bronchiolitis obliterans that follows viral bronchiolitis and pneumonitis. We reported in this article three paediatric patients with a spectrum of clinical presentation of this rare syndrome. Their diagnoses were confirmed with the use of CT scan. They were treated conservatively. None of them have developed any serious complication during the relative short follow up period. We suggested that a large scale studies should be performed to investigate the long term outcome in this group of patients.

Key words Bronchiolitis obliterans; Chest X-ray; Hyperlucency; Swyer-James/Macleod syndrome

Introduction

Swyer-James/Macleod syndrome (SJMS) is a rare disease which is characterised by unilateral hyperlucent lung on the chest radiograph. This syndrome was first described by Swyer and James in 1953 in a 6 years old boy who presented with recurrent respiratory infections.[1] The chest radiograph of this patient showed relative transradiancy of the right lung with significant reduction in vascular marking. Pneumonectomy was performed and the histological examination demonstrated bronchitis, bronchiolitis, obliteration of peripheral lung capillaries and emphysema. One year later in 1954, Macleod reported similar radiological features in nine adult patients with abnormal transradiancy of one lung.[2] These patients had only mild cough or were asymptomatic. Decreased filling of the distal bronchi and alveoli was noticed on the bronchography. We described in this article 3 paediatric patients presenting with this uncommon syndrome.

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Case 1

A 7 years old girl complained of 2 months history of chronic cough. She did not have difficulty in breathing, fever, history of choking. She has enjoyed good past health all along except for a few episodes of respiratory infection requiring intramuscular injection of medication in China. She was born in China and has moved to Hong Kong for 2 years. Her immunisation status has been updated after her emigration. Her trachea was deviated to the left on examination. There was decreased movement and breath sound on her left chest. No added sound was detected on auscultation. No finger clubbing was found. Other physical examination findings were essentially normal. Reduction in left lung volume and mediastinal shift to the same side was noted on chest radiograph (Figure 1). CT scan was obtained which demonstrated small lower and lingular lobe with minimal interstital markings (Figure 2). The radiological features were compatible with the diagnosis of Swyer James/Macleod syndrome. Pulmonary function test result showed restrictive lung pattern with FVC and TLC of 70% and 80% of the predicted volume respectively.

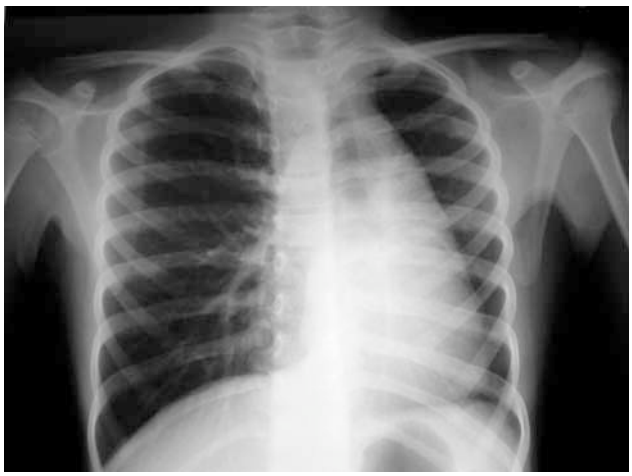

Figure 1 Chest radiograph shows reduction in perfusion and volume of left lung with mediastinal shift to the same side (Case 1).

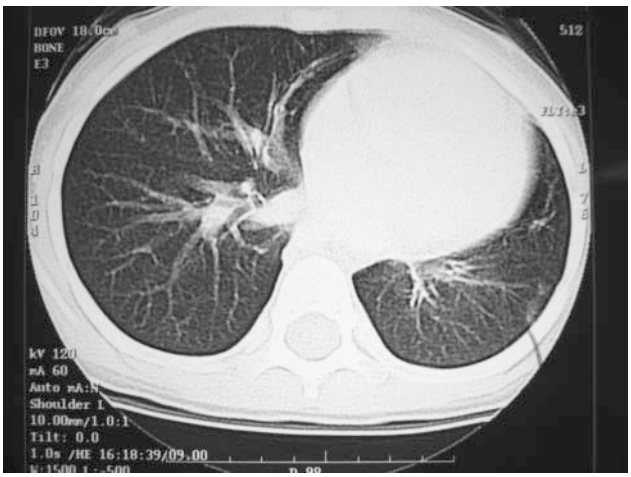

Figure 2 CT thorax reveals small left lower and linguar lobe with minimal intersitial lung markings (Case 1).

The DLCO is only 63% of the predicted value. Lung perfusion scintiscan was also performed and it revealed absence of perfusion to the left lung (Figure 3). Her cough gradually improved with symptomatic treatment. She was being followed up in our respiratory clinic and was clinically well.

She was admitted to our ward one year after her initial presentation for massive haemoptysis. There was no fever, weight loss, night sweating or contact history of tuberculosis. Blood tests including complete blood count, erythrocyte sedimentation rate, clotting profile and anti-mycoplasma IgM were normal. Mantoux test was negative. Culture of sputum and bronchoalveolar lavage (BAL) for bacteria and mycobacteria did not yield any microorganism. No abnormal cell was detected on the cytological examination of BAL fluid. CT scan showed only small left lung without other abnormality. Blood was found to be oozing out from the B6 opening of left lung on bronchoscopy. She was seen by a cardiothoracic surgeon. Her haemoptysis subsided spontaneously several days later and no surgical intervention was required. No particular cause for her haemoptysis was identified. She was subsequently discharged and there was no further recurrence of haemoptysis afterwards. We follow up this girl regularly in our respiratory clinic and she does not complain of any clinical symptom.

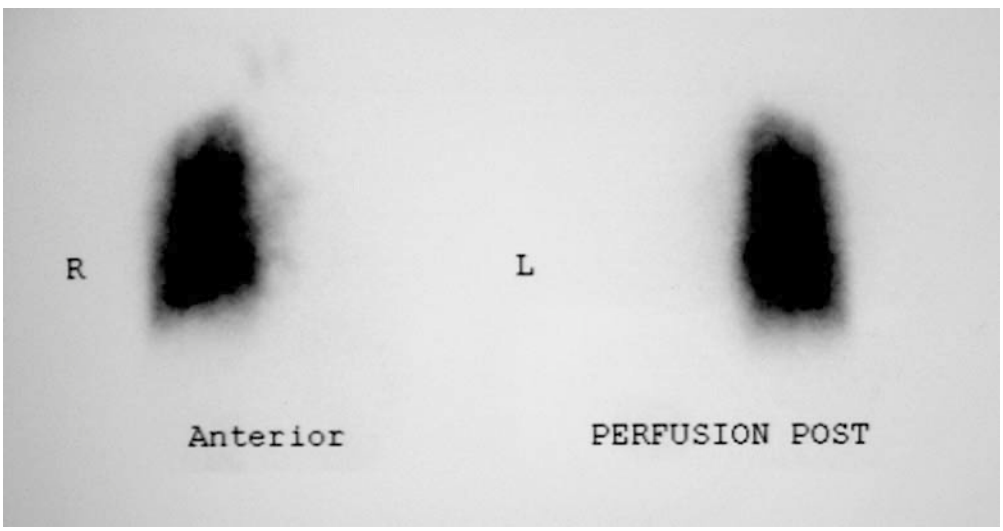

Figure 3 Lung perfusion scintiscan detects no perfusion to left lung (Case 1).

Case 2

A 5 years old girl was admitted for 4 days history of fever and cough. There was also associated shortness of breath. Several days of azithromycin was prescribed by a general practitioner before this admission but there was no clinical improvement. She was hospitalised twice one year before this episode of illness, first time for mycoplasma pneumonia and the second because of pneumonia caused by adenovirus and moraxella catarrhalis. She did not have any chronic symptom after these attacks of pneumonia. There was deviation of her trachea to the right side on examination. Her apex was slightly displaced to 1 centimetre medial to left mid clavicular line. The percussion note was dull on the right mid and lower zone. Diminished breath sound and crepitation was also noted over the same area. Other physical findings were essentially normal. Blood test revealed leucocytosis. The blood culture was negative and the sputum culture grew only commensals. Collapse-consolidation was shown on chest radiograph. She was treated as pneumonia with cefotaxime. She responded to the medical treatment with defervescence shortly after admission. Ten days course of antibiotics was given. However, there was persistent collapse of the right middle and lower lobe on chest X-ray (Figure 4). The bronchoscopic finding was unremarkable. CT thorax (Figure 5) demonstrated collapse of right middle lobe and consolidation of right lower lobe. There was also associated reduction in vascularity over the right lung. No feature of bronchiectasis was detected. The radiological appearances were suggestive of Swyer-James/Macleod syndrome. She was subsequently discharged and being regularly seen in our respiratory clinic. However, she defaulted our follow up appointment after coming back for once.

She presented again 4 years later with pneumonia caused by haemophilus influenzae. There was no clubbing of her fingers. Collapse of right middle and lower lobe with shifting of mediastinum to right side was detected on chest radiograph. She was successfully treated with a course of cefuroxime. Chest radiograph was repeated 3 months later which showed resolution of lung collapse but persistent decrease in right lung vasculature and volume with shifting of mediastinum to same side. She has been clinically well during her one year of follow up in our respiratory clinic.

Case 3

A 16 months old girl was referred to our clinic for incidental finding of tracheal deviation on chest X-ray obtained during an episode of bronchiolitis. She was clinically asymptomatic. On detail enquiry, she had past history of respiratory syncytial virus bronchiolitis requiring mechanical ventilation in another hospital one years ago. The parent did not attend the follow up after discharge. There was no finger clubbing on physical examination. Breath sound was slightly decreased over the left upper

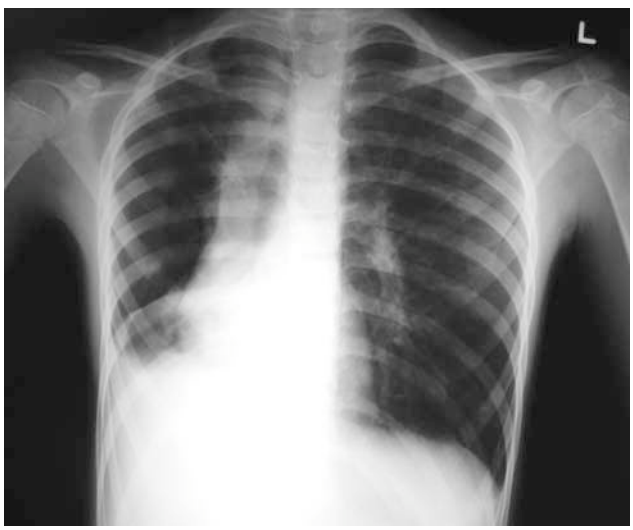

Figure 4 Chest X-ray demonstrates collapse of right middle and lower lobes with shifting of mediastinum to ipsilateral side (Case 2).

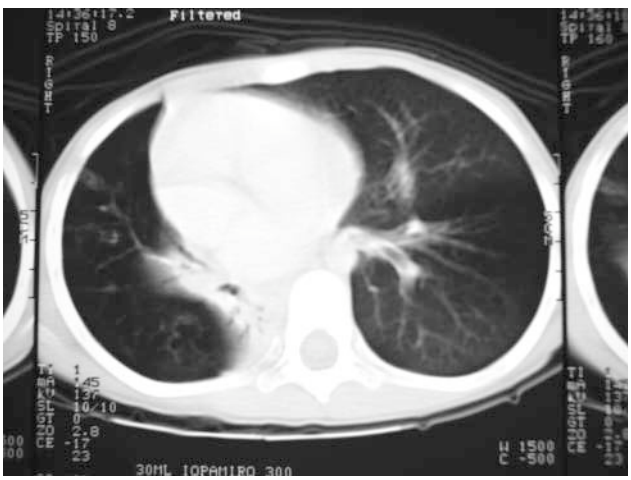

Figure 5 CT thorax shows subsegmental collapse of right lower lobe. Both volume and perfusion of the right middle and lower lobes are decreased (Case 2).

zone. No other added sound was detected. Chest radiograph revealed mild tracheal deviation to right side, hyperlucency with reduced vascularity over the left upper and middle zones (Figure 6). The left upper and lingular lobes were hyperlucent with decrease in lung volume and attenuation of arteries on CT thorax (Figure 7). The slight right side deviation of trachea as shown on both CT and chest X-ray was normal. The radiological features were compatible with Swyer-James/Macleod syndrome. She has been seen in our respiratory clinic for a year without development of any abnormal symptom or sign.

Discussion

The prevalence of SJMS was reported to be 0.01% in 17,450 survey chest radiograph.[3] It is believed that this syndrome is the sequelae of acute bronchiolitis complicating with obliteration of small airways, leading to destruction of lung parenchyma and hypoperfusion of the involved segment or lobe. While bronchiolitis obliterans accounts for the major factor of underlying pathogenesis, reflex pulmonary vasoconstriction and hypoperfusion causing hypoplasia of pulmonary vasculature also contributes to hyperlucency of the pathologic lung.[4] This may be a probable physiological protective mechanism to lessen effect of ventilation and perfusion mismatch in the case of lung damage. Some infection including pertussis, adenovirus, measles, mycoplasma were described in previous reports to be associated with SJMS.[5-10] As SJMS is an unusual complication of bronchiolitis, it is likely that other common respiratory viral infection of bronchiolitis such as respiratory syncytial virus may also result in SJMS. In fact, all three patients in our series had history of lower respiratory tract infection. Case 2 and case 3 actually had documented mycoplasma, adenoviral and RSV infection before diagnosis of SJMS. These findings were consistent with the observations in other studies.

SJMS presents with variable clinical features. Patient can be completely asymptomatic with hyperlucent lung field being an incident finding on the chest radiograph taken for other indications. Alternatively, they may have chronic cough, haemoptysis, exertional dyspnoea, recurrent chest infection or other respiratory symptoms secondary to bronchiectasis.[3,4,10] Spontaneous pneumothorax or pneumomediastinum can be the initial manifestation of this uncommon disease as well.[11] The clinical features of our patients are similar to those described in earlier reports except that one of them (case 1) had massive haemoptysis during her course of illness. This presentation has been previously described in adult patient with SJMS. To our knowledge, it is the first time that this symptom is reported to occur in a paediatric patient. The pulmonary function test in SJMS often revealed a restrictive lung pattern with decrease in vital capacity and expiratory flow.

Although being regarded as a typical radiological appearance of SJMS, "unilateral hyperlucency" on chest X-ray can be a feature of several other diseases. The radiological abnormality may be the consequence of congenital pulmonary artery agenesis/hypoplasia or acquired stenosis or compression of main pulmonary vessels. On the other hand, parenchymal disease including

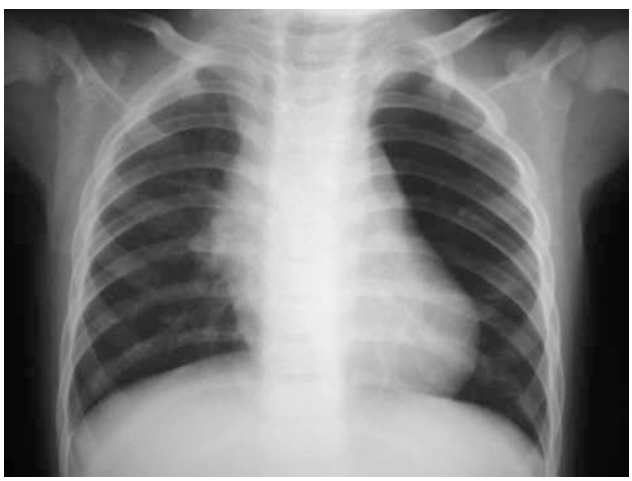

Figure 6 Chest radiograph illustrates hyperlucency with decreased vascularity over left upper and lingular lobes (Case 3).

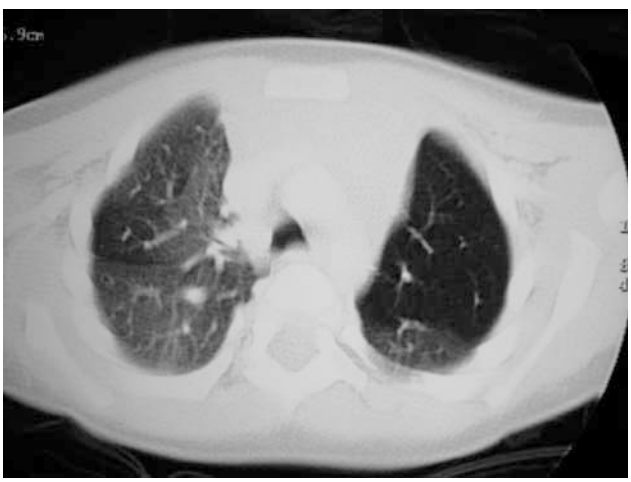

Figure 7 CT scan reveals hyperlucent left upper and lingular lobes with reduction in lung volume and attenuation of arteries (Case 3).

congenital lobar emphysema, bronchogenic cyst, bronchiectasis with air trapping, emphysematous bulla, emphysema resulting from bronchial stenosis and bronchospasm may give rise to similar radiological appearance.[3]

The initial diagnostic tool for SJMS should be plain chest radiograph obtained during both inspiration and expiration with radiolucency of affected lung being the pathognomonic feature of this disease. There may be diminished lung volume on involved side and shift of mediastinum during expiration to the normal lung. Lobar collapse was noticed on the chest X-ray and CT scan in case 2. This radiological feature has been observed in previous published case series of SJMS and bronchiolitis obliterans.[10,12] The collapse of lung is caused by fibrous obliteration of bronchi secondary to inflammation of the disease process. High resolution CT has now largely replaced the more sophisticated and invasive diagnostic procedures such as pulmonary angiography, radionuclide lung ventilation/perfusion scintigraphy and broncography as the choice of investigation for confirming the presence of SJMS. With the use of HRCT, hyperlucency, anatomy of lung parenchyma and pulmonary vessels can be better defined especially after intravenous contrast injection. HRCT also has the additional advantage of ruling out endobronchial tumor partially obstructing the lumen of a lobe or main bronchus in adult patients with SJMS.

The aim of management of SJMS is to prevent and treat intercurrent respiratory infection. Surgical intervention namely lobectomy, pneumonectomy is seldom required except when there is uncontrolled infection of the diseased lung segment.[3,10] There are only a few published literatures documenting use of lung resection in patients with SJMS.[3,10,13] Fregonese et al and his colleagues reported successful treatment of a 11 years old boy who suffered from SJMS complicated with bronchiectasis and fulminant pneumonia by pneumonectomy.[3]

The prognosis of SJMS is generally favourable. Bronchiectasis is a possible complication that affects the outcome of SJMS.[10] Therefore, patients with SJMS should be regularly followed up to look out for this complication and all respiratory tract infection must be adequately treated to prevent its occurrence. There is also risk of predisposition to pneumothorax and pneumomediastinum though it is rare. In case 3, parents defaulted follow up appointment of their child after an episode of severe bronchiolitis. This girl was subsequently diagnosed to have SJMS 2 years later. While it may be impractical to see all patients with lower respiratory tract infections after their discharge, follow up of patients with severe chest infection requiring PICU admission should be ascertained to monitor for development of long term pulmonary consequences namely SJMS, bronchiectasis and bronchiolitis obliterans. The three cases in our report ran a relatively benign clinical course. All of them were managed conservatively and none have had any complication as mentioned during the relative short follow up period. A large scale study should be performed in future to investigate the long term outcome particularly lung function and complication rate in this group of patients.

In summary, we reported three children with Swyer James/Macleod syndrome. They presented with a different spectrum of clinical symptoms. All of them have characteristic "unilateral hyperlucency" on the chest radiographs and their diagnoses were established on CT scan. We managed these patients conservatively and none of them have developed any serious complication.

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