CASE REPORT

Swyer-James-MacLeod Syndrome

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Summary

Swyer-James-MacLeod syndrome is a rare complication of respiratory tract infection occurring in early childhood. We report two children with chronic cough and recurrent wheezing who fulfilled the diagnostic criteria for this disorder:
1) Unilateral loss of lung volume with hyperlucency on chest x-ray
2) Unilateral reduction in vascularity on CT scan of the chest
3) Unilateral loss of perfusion on Technetium 99c lung scan

Key Words: Unilateral, Hyperlucent lung

Introduction

A unilateral hyperlucent chest x-ray as a result of unilateral pulmonary emphysema was first described in a 6 year old boy by Swyer and James in 1953 and a year later by MacLeod in 9 adults. It is a rare pulmonary disorder that complicates respiratory tract infection in early childhood and is considered to be due to a process similar to bronchiolitis obliterans. We present two children with recurrent wheezing who were referred for possible inhaled foreign body because their chest x-ray showed a unilateral hyperlucent lung. They were subsequently diagnosed to have Swyer-James-MacLeod syndrome.

Case History 1

BEX was well until 2 years of age when she developed bronchopneumonia that required in-patient treatment and parenteral antibiotics. Her chest x-ray showed bilateral diffuse pneumonic opacities. Her tracheal secretion culture result was negative. She was discharged well but subsequently developed recurrent cough and wheezing within 6 months. She was referred to University Hospital Kuala Lumpur at the age of 3 years for possible inhaled foreign body as her chest x-ray then showed a hyperlucent left lung (Fig. 1). There was no history of choking. Examination revealed a well nourished child with decreased breath sounds on the left chest. The rest of the examination was unremarkable. Bronchoscopy revealed normal bronchial mucosa with no foreign body. High resolution CT scan of her chest showed a small hyperlucent left lung with markedly reduced vascularity (Fig. 2). There was a matched ventilation-perfusion defect of her entire left lung with Xenon-Technetium 99c lung scan. In view of these findings, she was diagnosed to have Swyer-James-MacLeod syndrome. Inhaled beclomethasone 100mcg bd and salbutamol 200mcg prn were started for her recurrent cough and wheeze and she remains well on follow-up 2 years after diagnosis with no recurrent chest infections.

Case History 2

NBH was well until 3 years of age when he developed chronic cough that occurred on most nights with occasional purulent sputum. There was no haemoptysis. He also had breathlessness on vigorous exertion and

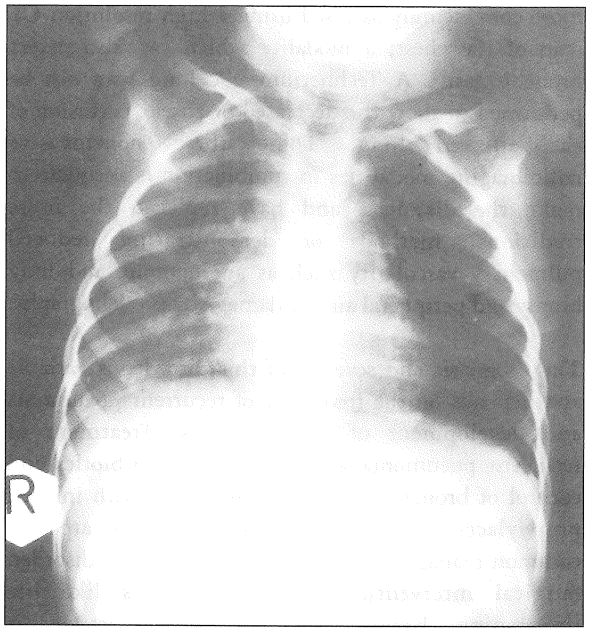

Fig. 1: Chest radiograph showing a hyperlucent left hemithorax

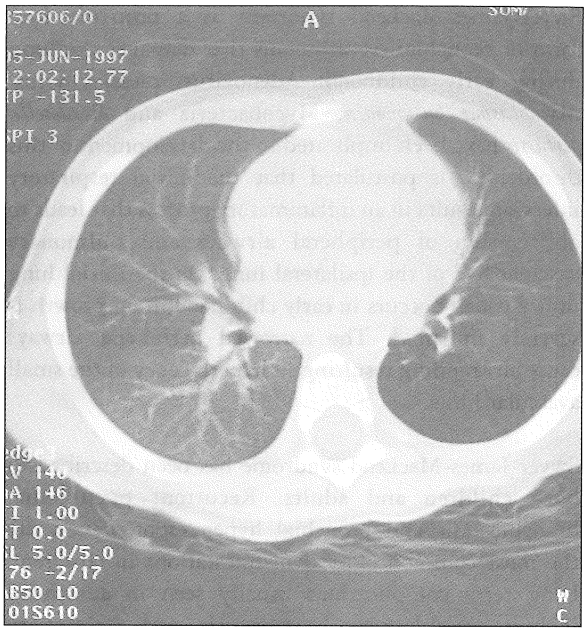

Fig. 2: CT scan of the chest showing reduced vascularity of the left lung

weight loss. He was diagnosed to have chronic asthma at 5 years of age and was started on inhaled sodium cromoglycate 20mg qid and salbutamol 200mcg prn after which his cough and wheeze improved. He was referred to University Hospital Kuala Lumpur at the age of 8 years for possible inhaled foreign body as his chest x ray showed a hyperlucent right lung. There was no history of contact with pulmonary tuberculosis. Examination revealed a thin Chinese boy (height 3rd centile, weight 10th centile). There was no clubbing. Chest expansion was reduced on the right. Auscultation revealed decreased breath sounds on the right side. The rest of the examination was normal. Investigations revealed haemoglobin 117g/L, platelet count 608 X 10^9/L, white cell count 11.9 X 10^9/L (N60% E5% L28% M6%) and ESR of 6mm/hour. Bronchoscopy showed a right main bronchus that was smaller than normal but no foreign body. Pulmonary function test showed significant restrictive lung disease with an obstructive component. {FEV_1 0.69 L (36% predicted), FVC 1.01 (33% predicted), $FEF_{25-75\%}$ 0.70 (31% predicted)}. High resolution CT scan of his chest showed that the right lung was hyperlucent and small with decreased vascularity. Technetium 99c lung scan showed marked reduction in right lung perfusion as it contributed only 12% of total lung vascularity by split function measurement. A diagnosis of Swyer-James-MacLeod syndrome was made based on these findings. NBH remains well on follow-up with infrequent cough and wheeze and markedly improved effort tolerance. His height and weight have improved to the 50th and 25th centile respectively over the next two years.

Discussion

An inhaled foreign body, extrinsic compression of the small airways and congenital lung malformations like localised emphysema should be considered in a child presenting with a hyperlucent lung on chest x-ray. However, the hyperlucent lung in these conditions are normal in size or hyperinflated. In the two patients described, the ipsilateral hyperlucent lung was instead small and avascular; a finding typical of Swyer-James-MacLeod syndrome.

Swyer-James-MacLeod syndrome is a post-infectious form of bronchiolitis obliterans that damages the lung during early childhood. Adenovirus, measles virus, *Mycoplasma pneumonia*, Mycobacteria and *Bordetella pertussis* have been implicated in the development of this disorder. It is postulated that the initial respiratory infection results in an inflammatory process that leads to obliteration of peripheral airways and pulmonary vascular bed of the ipsilateral lung. As the initial lung injury usually occurs in early childhood, lung growth is severely impaired. The narrowed peripheral airways cause air trapping resulting in hyperlucency of the small avascular lung.

Swyer-James-MacLeod syndrome has been described in both children and adults. Recurrent pneumonia, bronchiectasis and bronchial hyper-responsiveness are the most common clinical manifestations in children[1]. Long term complications usually seen in adulthood include troublesome haemoptysis and pulmonary hypertension leading to cor pulmonale.

The chest x-ray finding of an ipsilateral small hyperlucant lung is highly suggestive of Swyer-James-MacLeod syndrome. The air trapping and attenuation of the pulmonary vascularity of this condition can then be more convincingly assessed using a high resolution CT scan of the chest; a modality which can also detect bronchiectasis[2]. A Technetium 99c lung scan can be performed to show the decreased or absent perfusion of the affected lung. These three non-invasive radioimaging modalities in combination is adequate to make the diagnosis and have replaced the more traditional methods of demonstrating reduced pulmonary vascularity such as angiography and lung biopsy and peripheral airway damage by bronchography.

The prognosis and severity of this disorder is variable and depends on the frequency of recurrent pneumonia and development of bronchiectasis. Treatment of recurrent pneumonia with appropriate antibiotics and control of bronchial hyper-responsiveness with inhaled prophylactic agents and bronchodilators are the common management options needed for this disorder. Surgical intervention for complications like life threatening haemoptysis and bronchiectasis is sometimes required in adulthood[3].

In conclusion, Swyer-James-MacLeod syndrome should be considered when the unilateral hyperlucent lung found on chest x-ray is also small.

References

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