

Recurrent Wheeze in a Young Patient

A 22-year-old Malay lady was admitted to the respiratory service for asthma exacerbation. She had a history of asthma since childhood and was treated with a combination of inhaled corticosteroid (ICS) and long acting beta-2-agonist (LABA) with suboptimal control of her symptoms. Notably, she had 5 admissions for asthma exacerbations over a 1-year period between 2015 and 2016. She was a lifelong non-smoker and further history did not reveal any specific allergens that could have triggered her asthma. She did not complain of any nasal or gastro-oesophageal reflux symptoms. She worked as a personal assistant at an insurance company. There was no family history of chronic lung disease. There were bilateral rhonchi on examination of her chest. Vital signs and the rest of her physical examination were unremarkable. Her chest X-ray (CXR) is shown in Figure 1. Her full blood count was normal and did not reveal any eosinophilia.



Fig. 1. Chest X-ray showed hyper-inflated lungs.

A recent lung function test demonstrated evidence of obstructive airway disease with a forced expiratory volume during the 1st second/forced vital capacity (FEV₁/FVC) ratio of 45%. The FVC was normal at 2.26L (84% predicted) with a reduced FEV₁ of 1.03L (40% predicted). There was also air trapping with grossly elevated residual volume (RV) at 2.5L (241% predicted) and a residual volume/total lung capacity (RV/TLC) ratio of 53% (204% predicted). The diffusion capacity of carbon monoxide (DLCO) was normal indicating that it was predominantly an intrinsic airway abnormality.

Given her CXR and lung function findings, a high resolution computed tomography (HRCT) of the chest was performed (Figs. 2A and 2B). Further questioning revealed

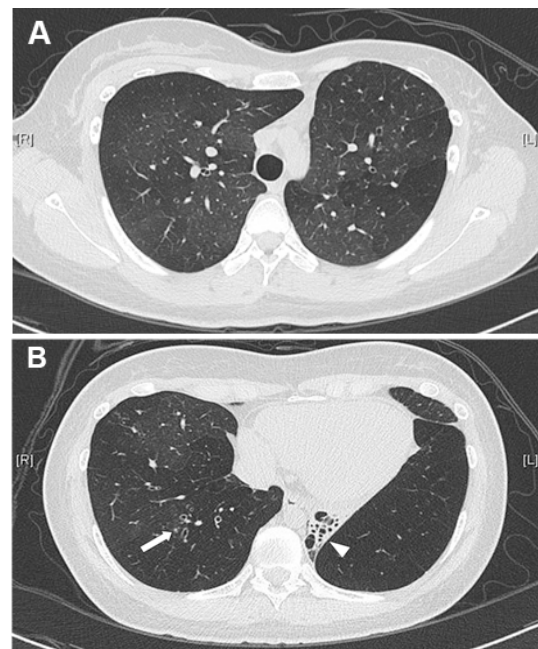


Fig. 2. High resolution computed tomography of the chest revealed: A) Mosaic attenuation in both upper lobes suggestive of small airway disease, and air-trapping was confirmed on expiratory images. B) Near-complete collapse of the left lower lobe (arrowheads) and bronchiectasis of the right lower lobe (arrows) associated with mosaic attenuation. There was reduction of vascularity and hyperlucency of the left lower zone which represents the hyper-inflated lingula.

that she was admitted to the intensive care unit at 2 years of age with severe pneumonia, and had frequent admissions for chest infections in the following year.

What is the diagnosis?

- A. Heart failure
- B. Eosinophilic granulomatosis with polyangiitis
- C. Vanishing lung syndrome
- D. Swyer-James-MacLeod syndrome
- E. Emphysema from alpha-1 antitrypsin deficiency

Discussion

The combination of clinical and imaging features is suggestive of Swyer-James-MacLeod syndrome (SJMS). SJMS, also known as unilateral hyperlucent lung syndrome, is a rare form of obliterative bronchiolitis with airflow obstruction accompanied by a decrease in the number and diameter of the ipsilateral peripheral pulmonary vessels. It is caused by injury to the developing lung before the age of 8 and usually follows viral aetiologies such as *Paramyxovirus morbillivirus*, *Influenza A*, and *Adenovirus*, and non-viral causes such as *Bordetella pertussis*, *Mycobacterium tuberculosis* and *Mycoplasma pneumoniae*.¹ The lung injury prevents the normal development of the alveolar buds² but they remain inflated due to collateral air drift.

Obliterative bronchiolitis (OB) encompasses a spectrum of disease that is associated with small airway injury caused by certain inhalational agents, infections, drug exposures, autoimmune causes and also as a complication of lung or hematopoietic stem cell transplantation.³ It is therefore also important to exclude other causes of OB in the workup of SJMS. CXR often provides a clue as it can show hyperlucency of one lung. The preferred imaging modality involves the use of HRCT imaging of the chest with thin collimation sections taken in both inspiratory and expiratory phases. In a review of 8 computed tomography (CT) images,⁴ unilateral lucency was seen in 7 cases and bilateral lucency in 1 case. Air trapping was found in all cases where inspiratory and expiratory scans were obtained. Other imaging features include atelectasis in the ipsilateral lower lobe (4 out of 8 cases) with accompanying bronchiectasis. Small foci opacities were also seen, which most likely represent residual scarring or chronic infections from previous pneumonia. CT chest also aids in excluding other differential diagnoses such as congenital hypoplastic lung, bullae or vascular abnormalities such as proximal interruption of the pulmonary arteries or pulmonary artery hypoplasia.

Patients with SJMS are usually young and can be asymptomatic or present with productive cough,

haemoptysis, shortness of breath, exertional dyspnoea or recurrent infections. Clinically, they can mimic asthma, and hence diagnosis requires a high index of suspicion.

There is airflow obstruction seen on the lung function test, and ventilation/perfusion scanning of the lungs often shows decreased perfusion of the affected lung.⁵

There is no consensus on how best to treat SJMS. General measures such as airway clearance and postural drainage with chest physiotherapy should be performed. Timely administration of appropriate vaccinations is essential to prevent pulmonary infections. Most patients receive bronchodilators and ICS. Pulmonary infections need to be treated early and aggressively, and if recurrent infections remain a problem, long-term antibiotic prophylaxis may be considered. In rare cases, such as those with recurrent pneumonias, recurrent pneumothoraces or worsening lung function, patients with SJMS are treated by surgical lung resection (lobectomy or pneumonectomy).⁶

In our patient, she had a history of childhood pneumonia and subsequent recurrent admissions for chest infections which could account for the complete collapse of the left lower lobe and the bronchiectatic changes. The imaging findings were not suggestive of heart failure where one would see smooth interlobular septal thickening, ground glass opacities or consolidation in dependent parts of the lung and pleural effusion. In eosinophilic granulomatosis with polyangiitis, one would expect eosinophilia on the full blood count, with CT imaging commonly showing ground-glass attenuation, consolidation, nodules or masses and pleural effusion.⁷ Emphysema would result in a reduction of DLCO, and the imaging findings were not suggestive of emphysema. Vanishing lung syndrome results from giant bullae in one or both upper lobes that are at least one-third of the hemithorax and compressing the surrounding lung parenchyma, which was not seen in the imaging of our patient.

Conclusion

SJMS can be erroneously diagnosed as asthma and the presence of a unilateral hyperlucent lung should raise the suspicion of this interesting but rare condition.

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Answer: D

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Audrey CR Wee,¹ MBBS, MRCP, MMed, Gin Tsen Chai,² MBBS, MRCP, FAMS,
John Abisheganaden,² MBBS, MRCP, FRCP, Gregory JL Kaw,³ MBBS, MMed, FRCP

¹Department of Respiratory and Critical Care Medicine, Khoo Teck Puat Hospital, Singapore

²Department of Respiratory and Critical Care Medicine, Tan Tock Seng Hospital, Singapore

³Department of Diagnostic Radiology, Tan Tock Seng Hospital, Singapore

Address for Correspondence: Dr Chai Gin Tsen, Department of Respiratory and Critical Care Medicine, Tan Tock Seng Hospital, 11 Jalan Tan Tock Seng, Singapore 308433.

Email: gin_tsen_chai@ttsh.com.sg
