Adult diagnosis of Swyer-James-Macleod syndrome

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SUMMARY

Swyer-James-Macleod syndrome (SJMS) is a rare, complex disease characterised by unilateral hyperlucent lung or lobe owing to loss of pulmonary vasculature and alveolar hyperdistention. Patients with SJMS may be asymptomatic at diagnosis, but its differential diagnosis with asthma is difficult. Almost all the reported cases of SJMS were managed by conservative therapy, comprising prevention and treatment of recurrent pulmonary infections.

BACKGROUND

Swyer-James-Macleod syndrome (SJMS) is a rare condition characterised by unilateral hyperlucency of a part of or the entire lung. It was first described in 1953 in a 6-year-old child,¹ and a year later, nine adult cases were reported by MacLeod.² The syndrome is thought to be secondary to bronchiolitis obliterans acquired in infancy, which results in obstruction of small airways and secondary emphysema, hypoplasia and/or agenesis of the pulmonary arteries resulting in pulmonary parenchyma hypoperfusion.

Patients are usually asymptomatic but typically present with a history of recurrent episodes of pulmonary infection with onset early in life. Symptomatic patients have dyspnoea on exertion, haemoptysis and/or chronic productive cough. It is identified most commonly by chest x-ray study revealing an increased density in the lung fields of the affected side with coexistent decreased lung volumes and is frequently an incidental finding. This radiological finding, first described by MacLeod as chest x-ray 'transradiancy', has been termed as 'hyperlucency' by radiologists in the intervening years, but is not specific for the syndrome.³

CASE PRESENTATION

A 40-year-old man was admitted to the hospital with dyspnoea, especially on exercise, for the past

6 months. He had a history of upper airway infections before the beginning of symptoms. He was treated for acute bronchitis with antibiotics bronchodilators and chest physiotherapy, in the last year. He has a 10 pack-year history of smoking. There was no significant problem in childhood. Chest auscultation demonstrated the decreased breath sound on the basis of left hemithorax.

INVESTIGATIONS

The chest radiograph on admission disclosed unilateral hyperlucency of right lower lung and decreased vascularity. CT scans revealed hyperlucency and diminished vascularity in the left lower lobe with hyperinflation of the pulmonary parenchyma (figure 1). CT scans demonstrated tubular bronchiectasis and minimal thickening of the bronchial walls in the left-affected lower lobes (figure 2). CT also showed similar tubular bronchiectasis in the left lung apicoposterior segment (figure 3).

DISCUSSION

SJMS is an uncommon clinic and pathological syndrome. The disease starts as an obliterative bronchiolitis with concomitant vasculitis commonly following infections with organisms such as adenovirus, measles or pertussis. The damage to the terminal or respiratory bronchioles in early childhood possibly prevents normal development of their alveolar buds.⁴ The patients of SJMS may be asymptomatic, but usually present with chronic and repeated pulmonary infections, dyspnoea on exertion, haemoptysis and even severe respiratory impairment.⁵⁻⁶ Clinically, the disease is often an incidental finding on chest x-ray study, although patients with this syndrome have been described with dyspnoea, decreased exercise tolerance, cough, haemoptysis, failure to thrive and recurrent pulmonary infections, as well as lung abscess.⁷⁻⁸ Clinical findings are non-specific and may include decreased chest expansion, rales or hyper-

Figure 1 Axial (A) and coronal (B) CT scans demonstrating hyperinflation (arrows) and diminished vascularity (arrowhead) in the pulmonary parenchyma.

To cite: Tutar O, Tekcan DE, Samanci C, et al. *BMJ Case Reports* Published online: 10 December 2012 doi:10.1136/bcr-2012-007349

Figure 2 Axial (A) and coronal (B) computed tomography scans demonstrating tubular bronchiectasis in the left lower lobes.

Figure 3 Axial CT scan demonstrating tubular bronchiectasis in the left lung apicoposterior segment.

resonance.⁹ Diagnosis in the past relied on typical chest x-ray findings in the absence of obstructing airway mass, pulmonary embolism or other cause for the radiological findings examined via bronchoscopy, ventilation-perfusion scans, angiography or scintigraphy. Pulmonary function test showed obstructive defecation in varied degrees.⁵ CT scan has been shown to have a higher sensitivity for the detection of hypoplastic lung lobes, bronchiectasis and pulmonary bronchus size when compared with standard end expiratory frontal chest x-ray studies.¹⁰ Treatment of SJMS is typically conservative and supportive, including close follow-up and management of recurrent pulmonary infections, and rarely with lung resections for recalcitrant disease.

Rarely, patients with SJMS are treated by lung resection for recalcitrant disease managed with chest physiotherapy for left bronchial drainage, low-dose inhaled steroid and inhaled bronchodilators.

Learning points

- Swyer-James-Macleod syndrome (SJMS) is a rare disease characterised by unilateral hyperlucent lung or lobe owing to loss of pulmonary vasculature and alveolar hyperdistention.
- The syndrome is thought to be secondary to bronchiolitis obliterans acquired in infancy, which results in obstruction of small airways and secondary emphysema.
- The patients of SJMS are usually asymptomatic, but present with chronic and repeated pulmonary infections, dyspnoea on exertion, haemoptysis and even severe respiratory impairment.
- The treatment is typically conservative and supportive.

Competing interests None.

Patient consent Obtained.

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