Swyer-James-MacLeod syndrome: an important differential diagnosis in adulthood

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DESCRIPTION

A 34-year-old man with a history of pulmonary tuberculosis in early adolescence presented to the emergency department with a 3-month history of relapsing fever, right-sided pleuritic thoracalgia and productive cough with purulent sputum. He had already received several courses of antibiotics with recurrence of symptoms. Chest examination revealed decreased breath sounds over the upper-right lung, but no crackles or wheezing; there was no peripheral oedema. Laboratory studies showed neutrophilic leucocytosis (total leucocyte count of $22.6 \times 10^9/L$ and absolute neutrophil count of $21.24 \times 10^9/L$) and elevated C reactive protein (373 mg/L). Chest radiograph revealed a hyperlucent right upper lobe with ipsilateral opacities (figure 1), and contrast CT confirmed confluent densifications associated with reduced pulmonary vasculature and increased lucency of the right upper lobe (figure 2). These particular findings (unilateral hyperlucency and corresponding decreased ventilation and perfusion) were already present on previous imaging examinations, but were disregarded. A bronchoscopy with biopsy was performed and endobronchial abnormalities were excluded. Cultures of sputum and bronchoalveolar lavage were negative, including for *Mycobacterium*. The patient was given a broad-spectrum antibiotic with a favourable response. Further pulmonary ventilation/perfusion scintigraphy (VPS) revealed a matched ventilation and perfusion defect of the affected area. Based on the clinical presentation and investigations, the diagnosis of Swyer-James-MacLeod syndrome (SJMS) was established.

SJMS is a rare complication of recurrent pulmonary infections in childhood, most commonly obliterative bronchiolitis. Various infectious agents are

Figure 2 CT ((A) coronal and (B) axial views) demonstrating decreased attenuation of the upper right lobe with signs of air trapping, marked hypoperfusion (yellow arrows) and bronchiectasis (green arrow) at the onset of symptoms (2019) and 3 months later on hospital admission (2020). Confluent opacities (blue arrows) compatible with active infection are shown in the right-sided images.

implicated, including viruses (measles, respiratory syncytial virus, influenza A) and bacteria (*Bordetella pertussis*, *Mycoplasma pneumoniae*, *Mycobacterium tuberculosis*).¹ The injury to the bronchiolar epithelium triggers an inflammatory response that leads to the development of submucosal fibrosis and destruction of the alveolar wall, causing airflow limitation and obstruction of the pulmonary capillary bed.² This functional hypoplasia of the bronchial vasculature may impair the growth of the affected lung, leading to a lung of preserved or slightly reduced size.

In most cases, diagnosis occurs in childhood, but some patients remain asymptomatic until adulthood, when SJMS has a variable spectrum of manifestations, ranging from recurrent respiratory infections to productive cough, exertional dyspnoea or haemoptysis; occasionally it is an incidental imaging finding.^{3,4} The differential diagnosis includes both congenital and acquired lung disorders (pulmonary agenesis, aplasia or hypoplasia, congenital lobar or localised interstitial emphysema, pneumothorax).^{2,3} The characteristic radiological pattern is defined by a hyperlucency associated with decreased pulmonary vascularity, expiratory air trapping and hyperinflation, which may be confined to one lung or lobe.^{2,5}

Figure 1 Posteroanterior view of chest radiograph showing right upper lobe hyperlucency (yellow arrows) at the onset of symptoms (2019) and 3 months later on hospital admission (2020), associated with ipsilateral opacities (blue arrows) compatible with active infection.

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Images in...

In some patients, bronchiectasis may be present and influence the clinical course and prognosis.³ Although a posteroanterior chest radiography is usually the first test performed, CT is the most sensitive and valuable imaging method for the evaluation of the lung involvement.⁵ A VPS may be useful to assess the extent of the disease.⁴ CT and resonance magnetic angiographies are other diagnostic modalities for SJMS.³

Conservative management based on prevention (pneumococcal and influenza vaccination) and prompt treatment of recurrent respiratory infections is the mainstay of therapy, but lung volume reduction surgery may be considered in certain cases.²

Learning points

- Swyer-James-MacLeod syndrome (SJMS) is a rare lung disease, considered a consequence of recurrent respiratory infections in childhood.
- Because of its characteristic radiological pattern, SJMS is often referred to as unilateral hyperlucent lung syndrome due to distension of the alveoli in conjunction with decreased parenchymal perfusion.
- Prompt recognition of this entity, especially in adulthood, is crucial, as it can mimic other pulmonary diseases and lead to misdiagnosis and an inappropriate approach.

Careful clinical and imaging assessment allows for a correct diagnosis, which is paramount when SJMS is suspected.

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