Clinical spectrum of Swyer-James-Macleod syndrome in adults

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ABSTRACT

Objective: The aim of this study is to describe the clinical and imaging features of Swyer-James-Macleod syndrome (SJMS) in 9 adults.

Methods: We reviewed the charts of 9 patients diagnosed with SJMS at the King Khalid University Hospital, Riyadh, Kingdom of Saudi Arabia over a 10 year period.

Results: The patients mean age was 38.1 years; males were more affected than females (7:2). Seven of the patients had symptoms referable to the chest and a similar number had compatible abnormalities on physical examination. The left lung was involved in all cases. Bronchiectasis was present in 7 (77.8%). Eight patients who underwent pulmonary function tests had combined defects. Two patients demonstrated significant reversibility. All patients had a stable course over at least before a 3 year follow-up period.

Conclusion: Swyer-James-Macleod syndrome has a diverse manifestations in adults and can mimic other pulmonary disorders, which may lead to incorrect diagnosis and inappropriate therapy. The course is generally a stable one.


Swyer and James first described unilateral hyperlucent lung in a child in 1953.1 One year later Macleod described 9 patients with the same syndrome.2 Now known as Swyer-James-Macleod syndrome (SJMS), this condition is characterized radiologically by a small or normal sized unilateral hyperlucent lung with decreased vascularity and air trapping on expiration. It is now believed to be a post-infective form of bronchiolitis obliterans (BO) most often following a viral infection3,4 but also described after tuberculous and mycoplasma pneumonia.5,6 Most patients are diagnosed in childhood when they present with recurrent respiratory tract infections. A few patients, however, remain asymptomatic until adulthood, when diagnosis is made either incidentally or with symptoms. This study examines the clinical and imaging spectrum of presentation of adult patients with SJMS diagnosed at the King Khalid University Hospital (KKUH), Riyadh, Kingdom of Saudi Arabia (KSA).

Methods. Nine patients presented to KKUH, over a 10-year-period (1991-2000) diagnosed with SJMS were studied retrospectively. The criteria for inclusion were (1) normal or small unilateral hyperlucent lung with air trapping on expiration, (2) presumed or documented antecedent lung infection or injury, (3) exclusion of other causes of hyperlucency. Demographic, clinical and imaging information were recorded in a data collection form. All patients had full history and physical examination in addition to plain chest x-ray and high resolution computerized tomography (HRCT) scan of the chest. Eight patients had pulmonary function tests, 5 ventilation perfusion scan, 2 bronchoscopy and one each of pulmonary angiography and aortogram.
Results. Table 1 shows the clinical, imaging and laboratory data of the patients. The mean age was 38.1 years (range 22-60 years). There were 7 males (77.8%) and 6 Saudis (66.7%). Most patients (77.7%; 7/9) presented with symptoms referable to the chest with shortness of breath on exertion being the most common. Two patients (22.3%) were discovered incidentally. The median duration of symptoms was 8 years. In all patients the left lung was involved. The right lung in addition was involved in 2 patients. Clinical signs were detectable in 8 patients with signs of air trapping being obvious in 5 (55.6%). Figure 1a & 1b show the chest radiograph of one of the patients demonstrating the typical unilateral hypertranslucency, decreased vascularity and air trapping on expiration with mediastinal swing. Bronchiectasis was present in 7 (77.8%) patients. Ventilation perfusion scans on 5 patients demonstrated the characteristic features of matched defect and marked trapping on the washout phase. Pulmonary function test universally revealed a combined defect (mild obstructive pattern with increased residual volume or total lung capacity ratio (116-167% predicted values) reflecting air trapping and reduced lung volume. All patients were treated medically and remained stable over a follow-up period of at least 3 years. One of the patient who presented with massive hemoptysis and found to have increased bronchial circulation on pulmonary angiography declined surgery but remained stable.

Discussion. The diagnosis of SJMS entails the exclusion of other causes of unilateral hypertranslucency. The nature of this study (history and full clinical examination, HRCT chest on all patients and follow-up) essentially excludes most of the more common causes. These include technical difficulties, chest wall abnormalities simulating hyperlucent lung, compensatory overinflation, emphysema, central

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex/ nationality</th>
<th>Symptoms</th>
<th>Duration</th>
<th>Signs</th>
<th>CXR / CT chest</th>
<th>PFT</th>
<th>V/Q scan</th>
<th>Others</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>43</td>
<td>Male Non-Saudi</td>
<td>Cough</td>
<td>9 years</td>
<td>Hyperresonance (L), reduced breath crepitations</td>
<td>Hyperinflation Bronchiectasis LLL</td>
<td>Mild obstructive</td>
<td>Poor perfusion and ventilation, air trapping</td>
<td>-</td>
<td>Stable</td>
</tr>
<tr>
<td>22</td>
<td>Male Saudi</td>
<td>SOBOE (cough, wheeze)</td>
<td>Childhood</td>
<td>Wheeze (L) crepitations</td>
<td>LLL cystic bronchiectasis</td>
<td>Mild obstructive</td>
<td>Matched reduced V/Q</td>
<td>Pseudomonas in sputum</td>
<td>Improved</td>
</tr>
<tr>
<td>22</td>
<td>Female Saudi</td>
<td>Hemoptysis</td>
<td>3 days</td>
<td>Reduced BS (L)</td>
<td>Focal bronchiectasis L</td>
<td>Mild obstructive</td>
<td>Matched defect</td>
<td>Aortogram, Klebsiella in sputum</td>
<td>Declined surgery, stable</td>
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<tr>
<td>40</td>
<td>Male Saudi</td>
<td>SOBOE (sputum, wheeze)</td>
<td>1 year</td>
<td>Reduced BS crepitations (L)</td>
<td>Cystic bronchiectasis (bilateral) L&gt;R</td>
<td>Mild obstructive, no reversibility</td>
<td>-</td>
<td>H.influenza in sputum</td>
<td>Stable</td>
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<td>60</td>
<td>Female Non-Saudi</td>
<td>SOBOE</td>
<td>2 years</td>
<td>Reduced BS hyperinflation</td>
<td>Focal bronchiectasis L/R</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>Stable</td>
</tr>
<tr>
<td>38</td>
<td>Male Saudi</td>
<td>Skin sarcoidosis</td>
<td>8 years</td>
<td>No obvious chest signs</td>
<td>(L) Lung reduced volume</td>
<td>Mild obstructive</td>
<td>Matched defect (L)</td>
<td>Angiography showed reduced LMFA</td>
<td>Stable</td>
</tr>
<tr>
<td>43</td>
<td>Males Saudi</td>
<td>Cough, SOB, hemoptysis</td>
<td>10 years</td>
<td>Reduced BS Wheezes, creeps (L)</td>
<td>Calcified para tracheal LAP</td>
<td>Mild obstructive</td>
<td>-</td>
<td>-</td>
<td>Stable</td>
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<tr>
<td>51</td>
<td>Male Saudi</td>
<td>Lower abdominal pain</td>
<td>-</td>
<td>Coarse (L) basal creeps</td>
<td>Cystic bronchiectasis calcified LAP</td>
<td>Mild obstructive</td>
<td>-</td>
<td>-</td>
<td>Stable</td>
</tr>
<tr>
<td>25</td>
<td>Male Saudi</td>
<td>SOBOE</td>
<td>10 years</td>
<td>Wheezes (L)</td>
<td>Basal (L) bronchiectasis</td>
<td>Mild obstructive</td>
<td>-</td>
<td>Bronchoscopy</td>
<td>Improved</td>
</tr>
</tbody>
</table>

SOB - shortness of breath, SOBOE - shortness of breath on exertion, LAP - lymphadenopathy, LMFA - left main pulmonary artery, L - left lung, R - right lung, V/Q - ventilation perfusion, BS - breath sounds, CXR - chest x-ray, CT - computerized tomography, PFT - pulmonary function test, LLL - left lower lobe.
obstruction and other vascular anomalies related to the pulmonary vessels. Although SJMS is a rare syndrome, a study of 40 consecutive patients with chronic unilateral hyperlucent lung showed that it was the most common cause accounting for 18 patients (45%) followed by localized emphysema (20%) and congenital hypoplastic pulmonary artery (10%). It is a post infectious form of bronchiolitis obliterans (BO) characterized by the presence of submucosal and peribronchiolar fibrosis with destruction and obliterator scarring of the small airways. It is uncommon occurring in 2 of 52 (3.8%) patients with BO in one study and 3 of 69 (4.3%) in another. This study shows that most adults with SJMS are asymptomatic, often for a prolonged period of time at presentation. One of our patients was treated for a long time with steroids as bronchial asthma. Another patient presenting acutely with massive hemoptysis was initially suspected to have pulmonary embolism due to pulmonary oligemia and treated with anticoagulants. The latter state may pose problems, as anticoagulation may prove deleterious in the setting of hemorrhage from a bronchial vessel. Shortness of breath on exertion was the most frequent symptom in this series in contrast to an earlier study of 9 patients in which shortness of breath was not a prominent feature. The left lung was involved in all cases. In Lucaya et al series only 2 patients, out of 13, had unilateral involvement of the right lung. The predilection of the left lung to obstruction and subsequent recurrent infections may be related to anatomical and hemodynamic factors. These include greater length, narrower diameter, and more horizontal angulation of the left main bronchus. It may be related to its course in the sub-aortic tunnel and hilum. High resolution computerized tomography scan of the chest is essential not only in excluding other causes of unilateral hypertranslucency but also in determining the presence and type of bronchiectasis. Computerized tomography helps to exclude central bronchial obstruction (making bronchoscopy unnecessary in some cases), cysts and vascular diseases as causes of hyperlucency. This study, which is in agreement with earlier studies, demonstrated that bronchiectasis is not a universal finding in SJMS. Patients without bronchiectasis or with cylindrical bronchiectasis had a lower incidence of pneumonia episodes than those with saccular bronchiectasis. The presence of bronchiectasis may be explained by one or more of these mechanisms: 1) bronchiolitis obliterans (BO) causing atelectasis or scarring which in turn leads to bronchial dilatation, 2) bronchiectasis itself being the primary event with distal spread to peripheral small airways leading to obliteration, 3) the initiating viral infection might damage both bronchioles and bronchi simultaneously. Two of our patients had calcified lymphadenopathy in the mediastinum suggesting an antecedent tuberculous infection. In an endemic area for tuberculosis, it is expected that this organism may play a more important causative role. Swyer-James-Macleod syndrome is an obstructive pulmonary disorder. This was confirmed by our findings in 8 patients who had a mild obstructive pattern on lung function testing. An earlier study reviewed 75 cases in the literature and demonstrated the obstructive nature of pulmonary function test, the individual variation in severity and the stable nature of the disease on a long-term basis. Ventilation perfusion (V/Q) scanning of the lungs in this syndrome characteristically shows a matched V/Q defect and a marked trapping on the washout phase on 133Xe.
scintigraphy. This investigation was carried out in 5 of our patients and demonstrated characteristic features. A study by Arslan et al\textsuperscript{16} showed that this modality of investigation was able to define the extent of disease better than plain radiographs and HRCT scan. It was able to identify areas of air trapping that appeared normal on plain radiographs and HRCT scan. Ventilation perfusion scan should therefore, be an integral part of the evaluation of the extent of disease. Management of SJMS is essentially medical therapy of the concomitant bronchiectasis. All patients remained stable or improved during the follow-up period. Surgery may however, be resorted to in patients with persistent disabling symptoms or those with life threatening hemorrhage.\textsuperscript{11,17} These patients often have succular, rather than cylindrical, bronchiectasis. Swyer-James-Macleod syndrome has a diverse clinical presentation in adults ranging from the asymptomatic to the catastrophic, with characteristic imaging signs; combined defects on pulmonary function test and generally a benign course. This syndrome should be recognized to avoid confusion with asthma and pulmonary embolism, which may result in inappropriate therapy.

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References