

**ADULT DIAGNOSIS OF SWYER-JAMES-MACLEOD SYNDROME:
RETROSPECTIVE ANALYSIS OF FOUR CASES**

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Abstract

Swyer-James-MacLeod Syndrome (SJMS) is a rare constrictive bronchiolitis with airflow obstruction and decreased number and diameter of ipsilateral peripheral pulmonary vessels. This syndrome is characterized by unilateral hyperlucency on chest radiography. Computerized tomography provides useful additional information. The diagnosis is usually made in childhood, but sometimes occur in adulthood. The disease often presents with dyspnea, decreased exercise tolerance, cough, hemoptysis and recurrent pulmonary infections. Swyer-James-MacLeod Syndrome may be confused with asthma or pulmonary embolism due to similar symptoms and may result in inappropriate therapy. This case series examines the clinical and imaging spectrum of four patients who were diagnosed as SJMS in adulthood.

Key words

swyer-james, macleod, pulmonary artery hypoplasia

Introduction

In 1953, Swyer and James, in 1954 Macleod, giving much more detail, described patients with unilateral hyperlucent lung. Swyer-James-Macleod syndrome (SJMS) or unilateral hyperlucent lung syndrome is a rare entity associated with postinfectious bronchiolitis obliterans occurring in childhood (1). This syndrome is a long-term complication of bronchiolitis in children, especially after adenoviral infection occurring in infancy (2). The affected child may be asymptomatic, but more often patient has recurrent pulmonary infections and after all develops bronchiectasis. Dyspnea, hemoptysis, and chronic productive cough are visible symptoms. A basic pathologic condition is the bronchiolitis associated with the obliteration of the small airways and severe emphysematous pattern owing to related alveolar destruction and dilated lung parenchyma(3). Peripheral pulmonary vascularization is decreased as a result of inflammation. Unilateral, or bilateral involvement is possible. The chest radiograph demonstrates lobar or unilateral hyperlucent lung; normal or reduced volume of the affected lung. Airflow obstruction is generally present on pulmonary function testing (PFT), and ventilation and perfusion (V/Q) scanning often reveals markedly decreased perfusion of the affected lung (4). The bronchographic findings are striking and limited to the abnormal lung(5). The major bronchi are normal, but the smaller branches are club-like, and occasionally small buds project from the ends of the peripheral divisions. Generally, there is almost complete absence of alveolar filling, demonstrated by a well-demarcated clear zone between the bronchiectatic smaller bronchi and the chest wall (5). This disorder is diagnosed in childhood after an evaluation for recurrent respiratory infections typically, but sometimes patients who have little or no sequelae bronchiectasis have minor symptoms or are asymptomatic and may, therefore, miss their diagnosis until adulthood (6). The clinical and imaging spectrum of 4 patients who were

diagnosed as SJMS in adulthood at a university hospital presented in this case series. These patients were remarkable with involvement of the left side and accompanied by bronchiectasis all of them. It is interesting that 3 patients were asymptomatic until adulthood despite they had bronchiectasis and obstructive defect in PFT.

Cases

Case 1:

A 39 years-old male patient admitted with complaints of dyspnea, cough and hemoptysis. He was admitted to different hospitals with complaints of cough and sputum production for about 3 years. Hemoptysis was in the amount of a teaspoon and very rare. He was treated for acute bronchitis with antibiotics and bronchodilators in the last 3 years. He has no smoking history. There was no significant problem in childhood. Pulmonary auscultation demonstrated the decreased breath sound on the basis of left hemithorax. The chest radiograph on admission disclosed unilateral hyperlucency, bronchiectasis and reduced lung volume on left lung. computerized tomography (CT) scans demonstrated pulmonary artery hypoplasia, bronchiectasis, decrease in density and volume loss on left lung(Figure 1). There was a moderate obstruction on PFT.

Case 2:

A 51 years-old female patient admitted with complaints of dyspnea, cough, chest pain and hemoptysis. Her symptoms had started after a severe infection of the respiratory tract at the age of 11 years. The patient's symptoms, decreases and disappears from time to time but continues for about 40 years. There was a moderate obstruction on PFT and she was being on medication by inhaled long-acting beta-agonist and corticosteroid combination for a long time with chronic obstructive pulmonary disease(COPD) diagnosis. Her smoking history was negative. There was

no significant problem in childhood. Subcrepitan crackles on the base of left hemithorax with bilateral expiratory wheezes was positive on pulmonary auscultation. The chest radiograph on admission disclosed unilateral hyperlucency, bronchiectasis and reduced lung volume on left lung. Computerized tomography scans demonstrated pulmonary artery hypoplasia, widespread bronchiectasis, decrease in density and volume loss on left lung(Figure 2).

Case 3

A 40 year old female was referred to us with complaints of cough, sputum, chest pain continued for 3 years. The recurrent pulmonary infections for about 3 years and an unstudied chronic productive cough were notable in her medical history. Bilateral expiratory wheeze was positive on physical examination. Chest radiographs showed hyperlucency of the left lung, with air-trapping during expiration. Computerized tomography scans demonstrated pulmonary artery hypoplasia, bronchiectasis, decrease in density and volume loss on left lung and mosaic perfusion defect on right lung(Figure 3). Pulmonary function tests revealed mild airway obstruction with no response to bronchodilators.

Case 4:

An 33 year old female patient was referred to our hospital with the pre-diagnosis of difficult asthma. She used bronchodilator and corticosteroid therapy about 6 months but did not benefit from the treatment. Although prominent symptom was dyspnea, a small amount of cough, sputum production and chest pain were positive. Bilateral expiratory wheeze and crepitant rales on the basis of left hemithorax were positive on pulmonary auscultation. Chest radiographs showed hyperlucency of the left lung. CT scans demonstrated pulmonary artery hypoplasia, bronchiectasis, decrease in density and volume loss on left side and mosaic perfusion defect on

both left and right side(Figure 4). Pulmonary function tests revealed moderate obstruction with response to bronchodilators and mild restriction

Discussion

Unilateral bronchiolitis obliterans with hyperinflation is quite rare, with a prevalence of 0.01% in 17,450 surveyed chest radiographs (7). It is presently considered to be an acquired disease secondary to viral bronchiolitis and pneumonitis in childhood etiological associated with *Paramyxovirus morbillivirus*, *Bordetella pertussis*, *Mycobacterium tuberculosis*, *Mycoplasma pneumoniae*, *influenza A* and *adenovirus types 3, 7 and 21*.(1). Interestingly, 3 of our patients, did not have any history of significant respiratory infections in childhood. Clinically, the disease is often presents with dyspnea, decreased exercise tolerance, cough, haemoptysis and recurrent pulmonary infections (2). Dyspnea on exertion was the most frequent symptom in a series with 8 patients in contrast to an earlier study of 9 patients in which dyspnea was not a prominent feature (8, 9). Our cases had one or more of these symptoms. Dyspnea and cough were most frequent symptoms. Physical examination findings are non-specific and may include decreased chest expansion, rales or hyper-resonance. Abba's study showed that most adults with SJMS are symptomatic, often for a prolonged period of time at presentation (9). Three of our patients are asymptomatic, for many years although one of them had symptoms ongoing for nearly 40 years at intervals. Complications of unilateral hyperlucent lung syndrome include recurrent infection in areas of bronchiectasis, lung abscess, and spontaneous pneumothorax (10, 11)

Swyer-James-Macleod syndrome is characterized by unilateral hyperlucency in radiographs and air-trapping. Chest radiography shows a normal or small lung with increased translucency owing to small hilar vessels that arborize sparsely and are attenuated. This typical chest radiography view was present in our cases and it was a stimulus for further examination. in

these cases. Thorax CT and high resolution CT provides useful additional information, such as patchy bilateral regions of hyperlucency or bronchiectasis. Computerized tomography showed the typical findings of hyperlucent areas with diffuse oligemia and air-trapping during expiration in two cases. These findings are known as a mosaic perfusion. Bronchiectasis is not necessary for diagnosis but present in some cases and there are several explanations for this. First, bronchiolitis obliterans can cause atelectasis or scarring that in turn leads to bronchial dilatation. There were left side bronchiectasis in our 4 cases. Pulmonary function test showed obstructive defect in varied degrees(12). In our cases, there was an obstruction in 4 patients and restriction in one patient at spirometric examination. Hypoxemia was not present in none of them in pulse oximeter. 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(13). Only symptomatic treatment needed in our patients. SJMS, can be confused with many chronic lung disease due to similar symptoms and may result in inappropriate therapy. One of our patients had been treated for long years with a COPD diagnosis. The other case was referred to us with the pre-diagnosis of difficult asthma .

Conclusion

In conclusion the main reason for reporting this case series is the rarity of cases who have been diagnosed with SJMS in adulthood. Although respiratory tract infection is considered to be a very important factor, many patients as 3 of our patients had no history of airway infection in childhood. Therefore, some other genetic or environmental factors may contribute to the development of this syndrome.

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Figure Legends

Figure 1. A. Right main pulmonary artery in normal diameter on thorax computed tomography in Case 1.**B.** Left pulmonary artery hypoplasia on thorax computed tomography in Case 1.

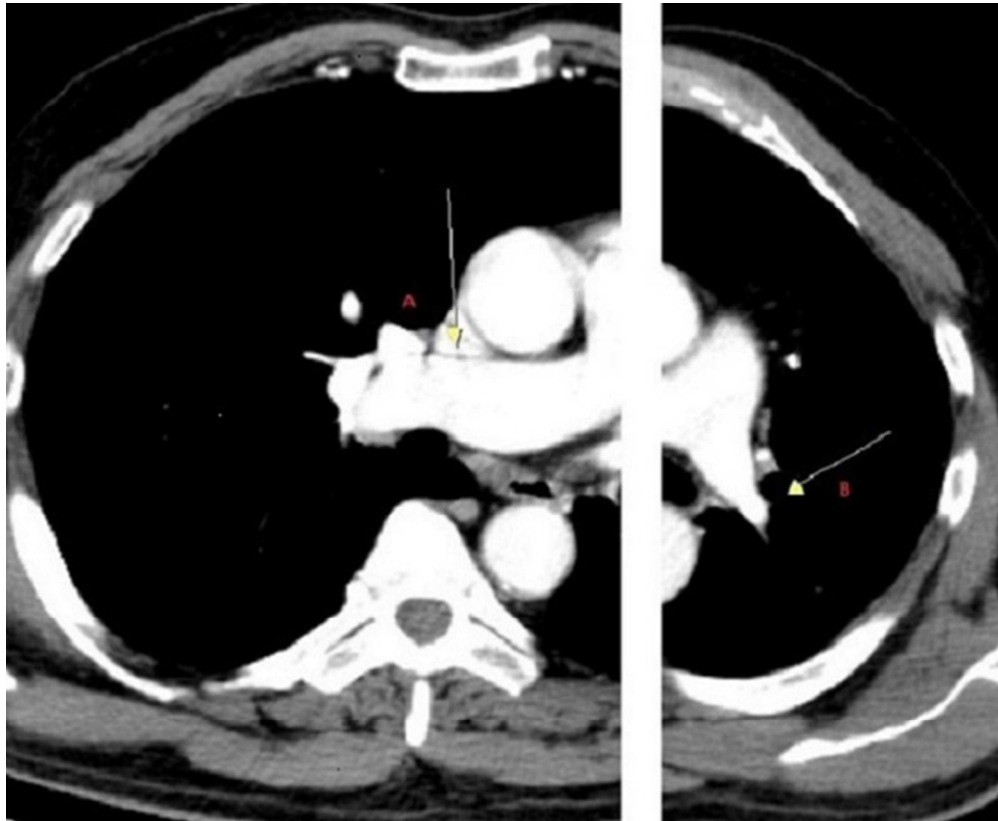
mAs:250, kV: 120.000000, SL:1.5

Figure 2. Widespread bronchiectasis on left lung on thorax computed tomography in Case 2.

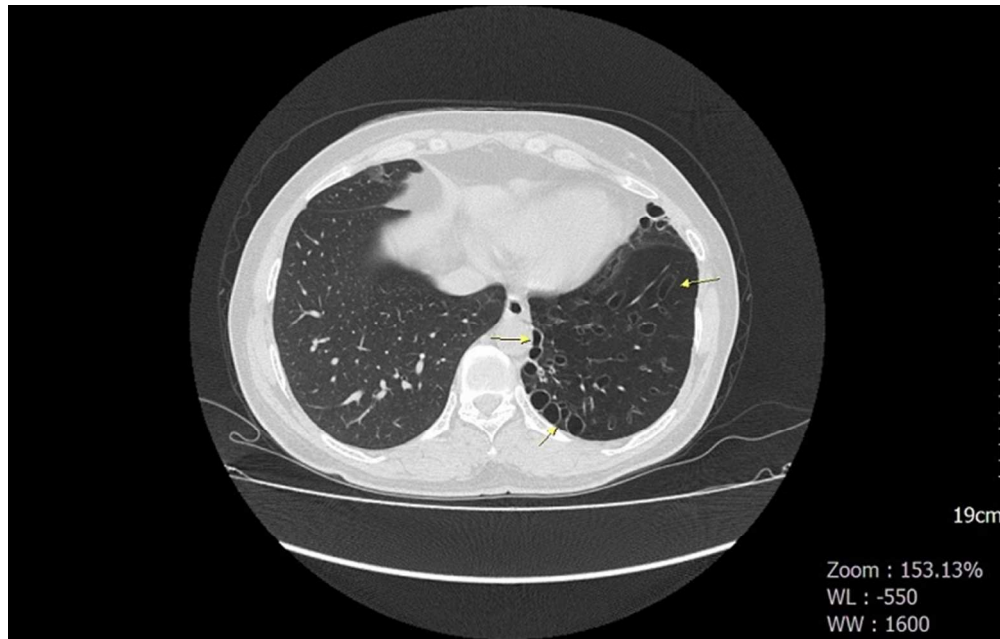
mAs:150, kV: 120.000000, SL:1.00

Figure 3. Mosaic perfusion areas on left lung in Case 3. mAs:300, kV:120.000000, SL:1.0

Figure 4. Low density areas on left lung in Case 4. mAs: 126, kV:120.000000, SL:1.0



158x129mm (96 x 96 DPI)



198x126mm (96 x 96 DPI)



198x122mm (96 x 96 DPI)



198x126mm (96 x 96 DPI)