

Swyer-James-Macleod Syndrome Presenting with Pulmonary Hypertension

Sh Hajsadeghi, M Chitsazan, HR Pouraliakbar, SR Jafarian Kerman

Tehran University of Medical Science, Tehran, Iran

Swyer-James-MacLeod Syndrome is a rare condition as a result of childhood pulmonary infection, especially bronchiolitis obliterans or viral bronchiolitis/pneumonia. It appears as increased radiolucency on chest X-ray, in the absence of obstructing lesions and can be confused for other thoracic disease processes such as a large pulmonary emboli or congenital bronchial and/or pulmonary vasculature malformations. We introduce a 46-year-old male patient presented with symptoms and signs of pulmonary hypertension which was initially misdiagnosed as chronic pulmonary emboli. This case highlights the possibility of pulmonary hypertension to be one of the cardinal manifestations of this syndrome, and outlines the significance of application of computed tomography in confirming the diagnosis of SJMS and in eliminating other diseases.

Introduction

Swyer-James-MacLeod syndrome (SJMS) is a rare phenomenon that is characterized by recurrent pulmonary infection, chronic cough, wheezing and dyspnea on exertion. It results from childhood pulmonary infection, especially bronchiolitis obliterans (BO) or viral bronchiolitis/pneumonia.¹ About 9.5% of BO cases occurred after SJMS.² In the literature, a case was reported after measles infection in a 5-year-old boy.³ This syndrome can be diagnosed by unilateral hypoplastic and hyperlucent lung, lobe or part of a lobe by Chest X-ray. It is almost always asymptomatic and diagnosed accidentally in routine examination or radiography for other respiratory disorders like asthma and may be neglected till adulthood. Some other diseases such as adenoid cystic carcinoma can also mimic this syndrome.⁴ Because of its very low incidence it is mainly misdiagnosed as other diseases that cause hyperlucency in radiographs and vascular abnormalities. Although, this syndrome has been reported in several previous studies, our patient is the second case reported with symptoms and signs of pulmonary artery hypertension. Thus, this case

highlights the possibility of pulmonary hypertension to be one of the cardinal manifestations of this syndrome, and outlines the significance of application of computed tomography in confirming the diagnosis of SJMS and in eliminating other diseases.

Case presentation

The patient was a 46 year-old Mediterranean male presented with a 2 year history of progressive exertional dyspnea, orthopnea and paroxysmal nocturnal dyspnea. He complained of productive cough with scant amounts of mucoid sputum and infrequent retrosternal chest pain within recent two years. Chest pain was unrelated to physical activity or emotional stress and lasted about 10 minutes and then relieved spontaneously with no medication. His dyspnea began limiting his daily activity since 6 months ago, which got him into class IV New York Heart Association classification. He also has been suffering from lower extremity edema evolving during this time period, more pronounced in the recent 6 months and poorly controlled with diuretics. Besides, he noted gastrointestinal complains such as dyspepsia, nausea, episodes of vomiting, constipation and abdominal distention after meals in the last 2-3 years. The patient was a cigarette smoker (25 packs/year) and had a history of inhalational

Correspondence:

Sh Hajsadeghi

Tehran University of Medical Science, Tehran, Iran

Cell: +98-9122210385

shhajsadeghi@yahoo.com

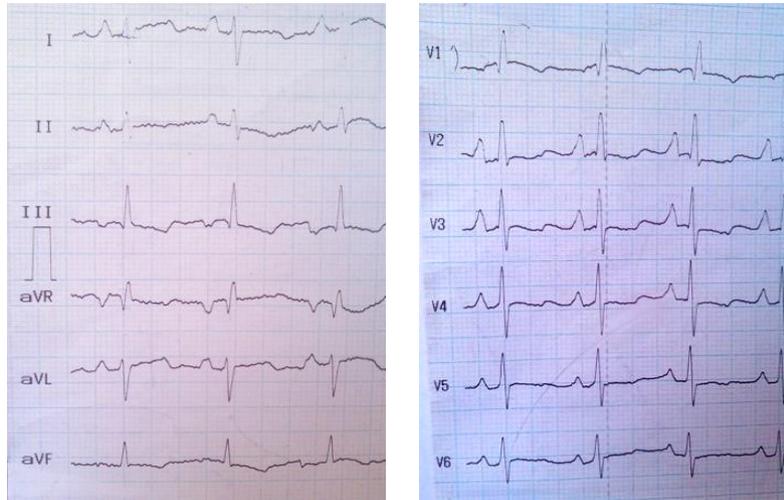


Figure 1. Electrocardiogram shows P pulmonale, right axis deviation and right ventricular hypertrophy with a qR pattern in lead V1.

cannabinoid usage and occupational cement powder exposure for almost 15 years. He denied any history of pulmonary infections such as measles, pertussis, recurrent pulmonary infection, bronchitis or pneumonitis during childhood. He had no family history of pulmonary diseases. On physical examination the patient was well-preserved, moderately obese, in respiratory distress and looked cyanotic with mild icteric sclera. The blood pressure was 100/80 mmHg; heart rate was 82/min and regular; respiratory rate was 32/min and morning oral temperature was 36.4°C. O₂ saturation in room air was 90%. Right ventricular heave was visible and palpable in the left lower sternal border and subxiphoid areas. Both right- and left-sided S₃ were heard at the left lower sternal border and the cardiac apex, respectively. A blowing grade III/VI holosystolic murmur was heard at the left lower sternal border, suggestive of tricuspid regurgitation. Thoracic expansion was symmetric and asymmetric resonance

to percussion was detected. Breath sounds were diminished in intensity over the left lung. Neck veins were distended to the angle of the mandible. The liver was slightly enlarged and tender; signs of ascites were evident in palpation and percussion. Additionally, a 3 plus (3+) lower extremity edema was notable. The rest of the physical examination was unremarkable. Routine laboratory results were as follows: hemoglobin level, 15.1gm/dl; hematocrit, 48.2%; red blood cell count, $5.15 \times 10^6 / \text{mm}^3$; platelet count, $1.53 \times 10^5 / \text{mm}^3$ and white blood cell count was $5.8 \times 10^4 / \text{mm}^3$; with a normal differential. A mild anisocytosis was evident (RDW=21.7), associated with MCV=93.6 fL, MCH=29.3 Pgm and MCHC=31.34%. Liver enzymes and albumin were normal with increased levels of bilirubin, both direct and total. Prothrombin time, partial prothrombin time and international normalized ratio were impaired. Additional blood tests showed increased levels of blood urea nitrogen and Creatinin. Sputum

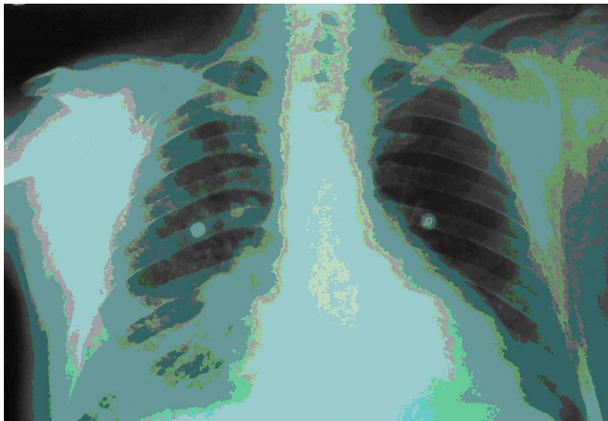


Figure 2. Chest X-ray shows unilateral hyperlucency (the left lung) with decreased volume

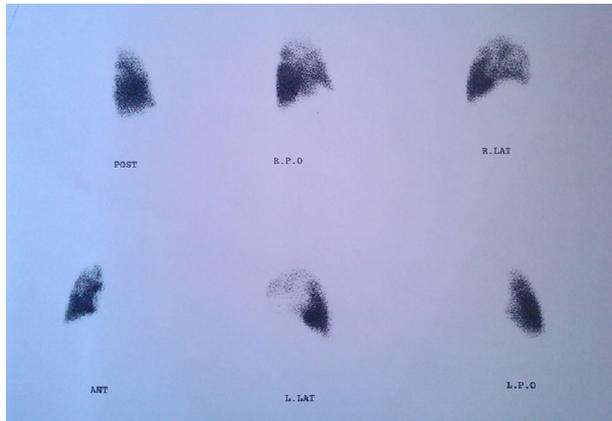


Figure 3. Lung perfusion scan. The left lung is not seen and there is nonhomogenous tracer uptake in the right lung.

Table 1. Spirometry Measurements

| Spirometric Data | %Pred | Meas1 ^a | %M1/P | Meas2 ^b | %M2/P | % M2/M1 |
|-----------------------|-------|--------------------|-------|--------------------|-------|---------|
| Vital capacity EX (L) | 4.13 | 2.67 | 64.8 | 2.84 | 68.8 | 106.2 |
| Vital capacity IN (L) | 4.13 | 2.83 | 68.5 | 2.95 | 71.5 | 101.1 |
| FEV1 (L) | 3.27 | 2.19 | 67.0 | 2.29 | 70.1 | 104.6 |
| FEV 1/ FVC (%) | | 88.82 | | 80.74 | | 90.9 |
| FEF 25-75% (L/S) | 3.92 | 1.68 | 42.8 | 1.75 | 44.5 | 103.9 |

^aBefore bronchodilator; ^bAfter bronchodilator; **Note:** EX: Expiration; IN: inspiration; FEV1: Forced expiratory volume in 1seconds; FVC: Forced vital capacity; Pred: Predicted; Meas1: Measured1; FEF 25-75%: Forced expiratory flow between 25 and 75% of vital capacity.

smear and culture demonstrated no pathogens, including acid fast bacilli. Skin test with tuberculin antigen was negative. The electrocardiogram showed conspicuous P wave consistent with marked atrial enlargement and right ventricular hypertrophy with a qR pattern in lead V1. (Fig. 1)

Transthoracic echocardiography was performed and showed severe pulmonary hypertension (systolic pulmonary artery pressure =90mmHg), severe right ventricular enlargement and systolic dysfunction, associated with severe tricuspid regurgitation (tricuspid regurgitant gradient =75mmHg). Small circumferential pericardial effusion was seen. Transesophageal echocardiography showed decreased flow in the left pulmonary veins. Inferior vena cava was increased in size; with less than 30% respiratory variation. Echocardiographic parameters were otherwise normal.

Chest x-ray (Fig. 2) showed hyperinflation, reduced lung volume and vascular markings in the left hemithorax. Pulmonary bay was prominent.

Arterial blood gas analysis showed hypoxemia and hypocapnia. Mixed restrictive and obstructive patterns were found by spirometry. (Table 1)

Doppler sonography of the lower extremity veins failed to show any evidence of prior deep vein thrombosis. Patient underwent radioisotope lung perfusion scan by Tc99m labeled MAA (figure 3) which showed nonhomogenous tracer uptake in the right lung and absence of perfusion in the left lung, and suggested high probability for left main pulmonary artery thromboembolism.

Contrast enhanced Multi-detector CT (MDCT) scan of the thorax performed on a 16 slice scanner (sensation Siemens 16 Germany) and showed no pulmonary emboli. Review of the axial images and of the multiplanar post-processed reconstructions showed lung asymmetry and increased lucency of the left lung and tubular bronchiectasis (Fig. 4). Left-sided pulmonary arteries were absent from proximal part. Air trapping was demonstrated

in both lungs, especially in the left. Ground glass opacity in the right lung was detected with mild pericardial reaction. Right pulmonary arteries showed enlargement. Two linear opacities were detected at the base of right lung and another one at the base of the left lung; which suggested scar tissue or atelectasis. Heart was enlarged and IVC was dilated.

Discussion

Swyer-James Macleod syndrome, also named unilateral hyperlucent syndrome, is a rare condition which was first described by Swyer and James in 1953 followed by Macleod in 1954.⁵⁻⁷

This syndrome is characterized radiographically by hyperlucency of a lobe, or the entire lung in the absence of obstructing mass with decreased vascular markings. It sometimes has areas of bronchiectasis. Initially, SJMS was thought to be congenital in origin and was attributed to hypoplasia of pulmonary artery. This theory was changed later because the number of bronchial generations and vascular branches in these patients are normal.⁸

Nowadays, SJMS is generally believed to be a post-infectious complication of viral bronchiolitis or pneumonitis acquired during early childhood.⁹ It also has been suggested that it may be due to an ongoing inflammatory process.¹⁰ It is presumed that bronchiolitis obliterans, the major defined pathogenic component, results in inflammation and fibrosis in the walls and contiguous tissue of the membranous and respiratory bronchioles with narrowing of their lumens.¹¹ Inter-alveolar septae fibrosis causes obliteration of the pulmonary capillary bed and secondarily diminishes blood flow to the major pulmonary artery segments which in turn results in the hypoplastic arterial development. In addition, the reduction in ventilation causes a compensatory decrease in perfusion via vasoconstriction. Hyperexpansion of the terminal air sacs secondary to bronchiolar obstruction of the peripheral airways offers additional mechanical resistance to blood flow

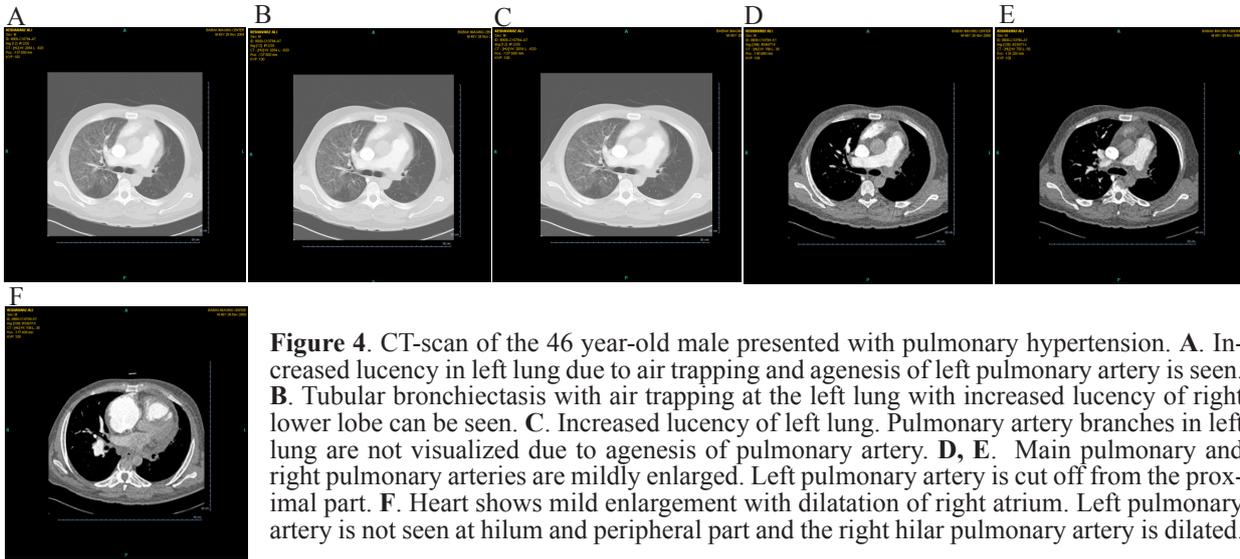


Figure 4. CT-scan of the 46 year-old male presented with pulmonary hypertension. **A.** Increased lucency in left lung due to air trapping and agenesis of left pulmonary artery is seen. **B.** Tubular bronchiectasis with air trapping at the left lung with increased lucency of right lower lobe can be seen. **C.** Increased lucency of left lung. Pulmonary artery branches in left lung are not visualized due to agenesis of pulmonary artery. **D, E.** Main pulmonary and right pulmonary arteries are mildly enlarged. Left pulmonary artery is cut off from the proximal part. **F.** Heart shows mild enlargement with dilatation of right atrium. Left pulmonary artery is not seen at hilum and peripheral part and the right hilar pulmonary artery is dilated.

through the alveolar capillaries and contributes to atrophy of the vascular bed.⁸ Hypoplasia of the pulmonary arteries is a reflection of this decrease in blood flow.

Clinically, most patients are asymptomatic and the disease is often an incidental finding on chest x-ray. Moreover, patients with this syndrome have been described with dyspnea, cough, hemoptysis, recurrent pulmonary infection, failure to thrive.^{10,12} However, most cases present during adulthood.

Diagnosis in the past relied on typical roentgenographic findings, described above, but radiography seldom fails to make an accurate diagnosis. For example, the unilateral hyperlucency of the affected hemithorax on the chest x-ray can be mimicked by an oligemia due to a large chronic pulmonary embolism (known as Westermark's sign) and distinguishing these two conditions cannot be made accurately by radiography. Additionally, this condition should be distinguished from other causes of unilateral hyperlucency of the lung such as congenital vascular and/or bronchial malformations.

With increasing capabilities of CT-imaging this modality can be used for a more accurate diagnosis, especially CT-scans performing during inspiration and after expiration. Although, SJMS is conventionally considered as unilateral disease, the availability of cross-sectional imaging has challenged this concept. It is now increasingly obvious that this condition may have bilateral heterogeneous involvement.¹³ In these circumstances, air trapping in the contralateral lung, defined as a mosaic pattern of attenuation, can be demonstrated by CT-imaging and confirmed by lack of change in volume on expiratory CT-scans and/or presence of perfusion scan defects. In other words, this syndrome should be

more correctly defined as⁹ a disease with a spectrum of involvement rather than a unilateral condition.

Additionally, CT-scan may be able to show ipsilateral or contralateral bronchiectasis associated with some cases of SJMS. Although, there is evidence suggesting that bronchiectasis is not a necessary component of SJMS.¹⁴ As a result, despite characteristic findings by chest radiography, CT-scan is the imaging modality of choice in establishing the diagnosis of SJMS.¹⁵

Our patient is the second case of SJMS diagnosed during evaluation for right-sided heart failure and severe pulmonary hypertension.¹⁶ In the initial steps for evaluating the patient presented with pulmonary hypertension a diagnosis of chronic pulmonary thromboembolism was suggested due to unilateral lung oligemia, seen on chest x-ray and consistent with Westermark's sign. However, when the patient underwent pulmonary CT-angiography pulmonary emboli was excluded and congenital absence of left pulmonary artery was suggested by the radiologist, but due to absence of mediastinal shift and significant lung volume loss, this diagnosis became questionable and CT-scan findings, mentioned above, made the diagnosis of SJMS more possible. Eventually, SJMS was confirmed by chest CT-scanning; which was performed during inspiration and expiration.

Acknowledgement

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