Case Report

Swyer-James-MacLeod Syndrome Misdiagnosed as COPD: A Case Report

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ABSTRACT

Swyer-James-MacLeod Syndrome (SJMS) is a rare disease characterized with unilateral hyperlucency and hypo-vascularity in pulmonary radiology. The basic pathology is the constrictive bronchiolitis developing depending on recurrent respiratory tract infections in babyhood. The majority of the patients receive diagnoses of recurrent respiratory tract infections in childhood or in young adulthood periods. SJMS may be easily skipped in discriminative diagnosis if the radiology is not inspected carefully in patients who have obstructive air flow in respiration function tests and who have past smoking history. High-resolution computer tomography (HRCT) of the lungs is the basic imaging method used today for diagnosis. We are presenting a 57-year-old female patient who was misdiagnosed with chronic obstructive pulmonary disease (COPD) for many years.

Key words: computed tomography, swyer-james-macleod syndrome, unilateral hyperlucent lung

INTRODUCTION

Swyer-James-MacLeod Syndrome (SJMS) is a complex and rare disease characterized by post-infectious constrictive bronchiolitis histopathology, classically, unilateral hyperlucency in lung radiology and small hilus depending on the hypo-vascularity in the affected side. The syndrome received its full name with MacLeod’s diagnosis of 9 patients, whose ages varied between 18 and 41, after they were presented with recurrent broncho-pneumonia attacks and a 6-year-old patient by Swyer and James in 1953[1-2]. The majority of the patients receive diagnoses of recurrent respiratory tract infections in childhood or young adulthood. SJMS may be easily skipped in discriminative diagnosis of the patients of older age group if the radiology is not inspected carefully in patients who have obstructive air flow in respiration function test and who have past smoking history. High-Resolution Computer Tomography (HRCT) of the lungs is the basic imaging method used today for diagnosis. We are presenting a 57-year-old female patient who was followed with the misdiagnosis of chronic obstructive pulmonary disease (COPD) for many years together with literature information.

CASE REPORT

A 57-year-old female patient who was misdiagnosed with COPD for 11 years was referred to our clinic due to increasing difficulty in breathing for the past 3 months. The patient had smoked cigarettes for 20 years, and had quit smoking for the past 5 years. The patient was hospitalized 3 times with the diagnosis of pneumonia presented with hemoptysis in the past 2 years. The respiratory sounds were decreased in the left lung basal, and there were crepitant rales in bilateral basals. In the respiratory function test of the patient, the forced vital capacity (FVC) was 1.45 lt, and the expected value was 64%; the FEV₁ was 0.67 lt, and the expected value was 35%; FEV₁/FVC was 46%. In the diffusion test, the diffusing capacity of the lungs for carbon monoxide (DLCO) was 18.8 ml/mmhg/min, 92% of the expected value. The DLCO/alveolar volume

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(VA) was 4.62 mL/mHg/min/L, 93% of the expected value. In the lung volume measurement, the vital capacity (VC) was 1.51 lt, 66% of the expected value, the residual volume (RV) was 7.25 lt, 454% of the expected value and total lung capacity (TLC) was 8.76 lt, 213% of the expected value. The RV/TLC rate was increased (83%). The patient was using the salmeterol xinafoate and fluticasone propionate combination, tiotropium bromide monohydrate and, when necessary, salbutamol sulphate + ipratropium bromide monohydrate inhaler medications regularly. The full blood, C-reactive protein (CRP) and biochemical tests were normal, no infection was detected. In the pulmonary graphics, the left lung was hyperlucent when compared with the right lung, the lower left lobe was hypoplasic, and the left hilus was not followed (Fig 1). In pulmonary HRCT, the lower left lung lobe was hypoplasic, the left lung upper lobe and lingular lobe were increased. The volume of the left lung was decreased, the right lung was hypertrophic and mosaic attenuation was observed in the bilateral parenchyma. There was clear bronchiectasis in both lungs especially in the left lower lobe (Fig 2). In the 3D reformation sections of the HRCT, the right pulmonary artery diameter (25 mm) was increased and the left lung lower lobe pulmonary artery diameter (7.5 mm) was hypoplastic (Fig 3). Bronchoscopy was applied to the patient and the area until the right side system sub-segments were observed as being clear; there was increase in the secretion and it was clear in the lower left lobe superior segment. In addition, the lower left lobe sub-segments were narrow and hyperemic. In the biopsy, bronchial wall showing inflammation findings was determined. In the lung ventilation/perfusion (V/Q) scintigraphy, the total left lung perfusion was 19.8%, the right lung was 80.2% (Fig 1). There was
matched V/Q defect. Conservative treatment was recommended to the patient including infection control (influenza and pneumococcal vaccines), pulmonary rehabilitation and bronchodilator therapy. Patient was also informed that surgery could be a choice in situations of persistent infections and recurrent hemoptysis attacks.

**DISCUSSION**

The diagnosis of SJMS is clinical-radiological and is performed by excluding the reasons of the unilateral hyperlucent images in lung x-ray. The prevalence of this rare disease was reported as 0.01% in 17,450 pulmonary radiographies\[^3\]. Mistaken imaging technique (high contrast and low kilo-voltage, the rotation of the patient, the existence of scoliosis), lack of asymmetrical soft tissue (mastectomy, Poland syndrome), extra-pulmonary air collections (pneumothorax, mediastinal emphysema, bullous emphysema, congenital lobar emphysema), air way obstruction (endobronchial foreign object or tumor, bronchial compression), pulmonary vascular diseases (pulmonary emboli, pulmonary artery hypoplasia) are among the major reasons that have to be excluded\[^4\]. Opacity in normal pulmonary radiology is formed with pulmonary vascularity. Since the pulmonary vessels that are surrounded with air contain blood in soft tissue opacity-density, the decrease in their size and number result in hyperlucency in radiological terms. The hyperlucency in SJMS is the pulmonary hypo-vascularity and the decreased blood flow developing as depending on the fibrosis of the interalveolar area\[^5\]. With recurrent infections before the age of eight, narrowing and occlusion occurs in the lumen depending on the submucosal fibrosis in the small air ways located in the distal of the terminal bronchioles. Measles, bordetella pertusis, mycoplasma pneumonia, influenza A,

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**Fig 2:** In pulmonary HRCT, in coronal sections, especially bronchiectasis areas in the left lung, decrease in the volume of the left lung, increase in the aeration in the right lung, hypoplastic left pulmonary artery and decreased peripheral vascularity are observed. In the sagittal section, compensator hypertrophy in left lung upper lobe and lingula, and hypoplastic left lower lobe are observed.

**Fig 3:** Hypoplastic left pulmonary artery and its branches that are decreased in number and in diameter are observed in 3D pulmonary HRCT reformat images.
adenovirus (Type 3, 7 and 21), mycobacteria tuberculosis and paramyxovirus morbillivirus are the microorganisms most commonly held responsible[6]. Thickening in the walls of the bronchioles (peribronchial inflammation), fibrosis, and bronchial dilatation prevent the normal development of the alveolar bag. These acquired bronchiolar decrease the obliteration blood flow and lead to hypoplastic pulmonary vascularity. Emphysema and bronchiectasis develop as secondary to the recurrent infections. The type and the severity of the bronchiectasis determine the appearance age and the severity of the symptoms[5-6]. The size of the lungs changes according to the period in which the bronchiolitis develops. The size of the lungs stay smaller with the damage developing in babyhood, and bronchiectasis becomes clearer and the patient gives symptoms in early childhood period. Since lung development is influenced less in SJMS pathology that occurs in late childhood period, the size does not change and the bronchiectasis is minimal. There are exertional dyspnea, chronic productive cough, hemoptysis and recurrent respiratory tract infections in patients[4]. Our case had dyspnea which did not heal despite the treatment. She had productive cough depending on the bronchiectasis, and seldom had hemoptysis attacks. When we questioned her history, we learnt that she had frequent infections in her childhood. We consider that the bronchiectasis in pulmonary HRCT is secondary to the viral bronchiolitis she had. We also consider that medical history and physical examination must extend to childhood in adults who have frequent respiratory tract infections and whose symptoms do not heal despite treatments, and evaluate the condition in detail with pulmonary HRCT. Our patient had partial hyperlucency in X-ray and this was not beneficial in detecting SJMS.

There are obstructions in air flow at various levels in pulmonary function test. The pulmonary function tests of the 75 cases in the literature were examined and it was observed that although the severity changed from patient to patient, the obstructive type was observed more frequently, and it progresses in a stable manner in the long run[7]. We determined the obstructive type air flow pattern in our case, which complies with the literature.

Pulmonary graphics taken in inspiratory and expiratory phase is the first imaging method for diagnosis. In expiratory film, rather than the inspiratory one, the hyperlucency depending on air imprisonment, mediastina swift to the healthy side and the opacity not increasing are diagnostic signs. There is an over-air in the pulmonary parenchyma distal to the obliterated terminal bronchial due to collateral air-flow. Bronchography was used more for diagnosis in the past before tomography was used frequently. Classical SJMS sign was the common bronchiectasis, and together with it, the terminal bronchiolar showing the filling defect, which is expressed as “pruned tree”[1-2]. Pulmonary HRCT firstly eliminates the single-sided hyperlucency- causing reasons. Air trap, attenuation in mosaic pattern, and no change in volume are diagnostic signs in the sections taken in expiration. The disease and symptom relation, which shows the existence and prevalence of bronchiectasis, gives us a clue. In addition, bronchial dilatation shows hypovascularity and atelectasis. In pulmonary angiography, there is reduction in the pulmonary artery diameter, and decrease in peripheral area vascularization. In pulmonary scintigraphy, there is single-sided decrease in perfusion and matched ventilation-perfusion (V/Q) defect. Arslan et al conducted a study and reported that the V/Q scan was more beneficial in showing the air trap areas, which cannot be observed in pulmonary graphics and in HRCT, in determining the level of the disease[8]. The perfusion defect in our patient was found to be depending on hypo-vascularity in left lung lower lobe. We reached the diagnosis in our patient with pulmonary HRCT and supported it with V/Q scanning.

Abba et al reported 9 patients with SJMS involving the left lung and whose average age values were 38.1. Seven of the patients were male and exertional dyspnea was the most frequent symptom. They reported that most of the adult patients were symptomatic for a long time before the diagnosis, bronchiectasis was not a rule for SJMS, and ventilation perfusion scanning was important in determining the level of the disease[6]. The treatment of SJMS is conservative. Inhaled broncho-dilator medication to control the symptoms and respiratory tract infections, antibiotics, and vaccinations of influenza and pneumonia are used. Lobectomy or pneumonectomy are applied in persistent infections and recurrent hemoptysis attacks.

CONCLUSION

As a result, if detailed medical history with physical examination and radiological tests are not conducted, it is difficult to discriminate SJMS from other obstructive pathology reasons in old age. Especially in the presence of hyperlucency in partial areas in the lungs, the SJMS diagnosis may be omitted if supplementary imaging is not conducted. By presenting this case, we wanted to be the models in the literature by showing that SJMS may be confused with COPD in older patients.
REFERENCES