Case report

Swyer–James–MacLeod syndrome with an anomalous origin of coronary artery: Case report

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A B S T R A C T

Swyer–James (Macleod) syndrome was first defined in the 1950s by Swyer, James and Macleod in patients with unilateral hyperlucent lungs. Coronary artery anomalies are congenital anomalies that affect a small part of the population. They constitute about 1–2% of congenital heart diseases. The incidence of a left coronary artery arising from the right coronary sinus Valsalva has been reported as 0.017%, and 1.3% among coronary artery anomalies. We hereby present this case since the case was diagnosed in adult age and was accompanied by a rare congenital heart disease.

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1. Introduction

Swyer–James (Macleod) syndrome was first defined in the 1950s by Swyer, James and Macleod in patients with unilateral hyperlucent lungs.1–6 Swyer–James (Macleod) syndrome occurs due to bronchiolitis obliterans that may develop as a result of many causes in early childhood.1–6 Infections due to viral and atypical bacterial agents, medications, radiation therapy and foreign body aspiration may be responsible in the etiology. Coronary artery anomalies are congenital anomalies that affect a small part of the population. They constitute about 1–2% of congenital heart diseases.7,8 The incidence of a left coronary artery arising from the right coronary sinus Valsalva has been reported as 0.017%, and 1.3% among coronary artery anomalies.9,10

We hereby present this case since the case was diagnosed in adult age and was accompanied by a rare congenital heart disease.

2. Case report

A sixty-year-old female patient presented with complaints of dyspnea, cough, sputum, chest pain, swelling in the legs and reduced vision. She was a smoker of 40 packets a year, and she had remitting complaints of cough and sputum since childhood, and had been suffering from chest pain and leg swelling for a month. She had been treated for chronic obstructive pulmonary disease (COPD) for 7–8 years and in the last 2 or 3 years, she had received a diagnosis of COPD-Cor pulmonale and was being treated for it. On respiratory examination, the intensity of breath sounds was found to have decreased in the right middle and lower parts compared to the left; there were crackles in the right lower lobe and sonar rhonchi in all the regions of the lung. On posteroanterior chest radiography, a hyperlucent right lung was observed (Figure 1). On electrocardiography (ECG), sinus rhythm and non-specific ST depression and T negativity were observed on the precordial derivations. On echocardiography, the right cardiac cavities were seen to be wide and the pulmonary artery systolic pressure was measured as 65 mmHg. The D-dimer level was lower than 500 ng/ml. There were no findings related with deep venous thrombosis on the lower extremity Doppler examination. On spirometry: FEV1/FVC: 77%; With 42%, a severe restriction and obstruction in respiratory functions was found. On thoracic computerized tomography (CT) and high resolution CT, right pulmonary artery hypoplasia, reduction in the diameter of the right pulmonary artery branches, hyperlucency, right lung lower lobe bronchiectasis and atelectasis were detected (Figure 2). Since the patient had pulmonary arterial hypertension, the right
atrium was catheterized and she underwent simultaneous coronary angiography due to ECG changes. Pulmonary artery pressures were: systolic 61 mmHg, diastolic 22 mmHg, and mean: 38 mmHg, measured by the invasive catheter. Pulmonary angiographic study of the thorax displayed prominent reduction of caliber in the right pulmonary artery branches (Figure 3). The simultaneously performed coronary angiography revealed normal location and branches of the right coronary artery, while the left coronary system (left anterior descending and left circumflex arteries) was observed to have originated from the right sinus Valsalva as a single coronary artery (Figure 4).

Diagnosed as Swyer–James (Macleod) syndrome, antibiotics, bronchodilators and pulmonary hypertension treatment were begun directed toward her complaints. Cataract surgery was recommended directed to reduced vision. Following the treatment, her symptoms recovered and she was discharged with the recommendations of receiving the viral influenza vaccination every year and pneumococcal vaccination every five years and to come back for follow-up examinations.

3. Discussion

Although rarely seen, this syndrome was detected at a rate of 0.01% on 17,450 chest X-rays. Our case is the only case of Swyer–James (Macleod) syndrome diagnosed in our clinic in the last two years. Our case is diagnosed at an adult age. Exertional dyspnea, hemoptysis and chronic productive cough are the most prominent
symptoms. The complaints of cough and sputum recurred since childhood age in our case, and in the last month, chest pain and leg swelling were observed. The patient had been under follow-up treatment for COPD for up to 7 or 8 years before and for COPD-cor pulmonale in the last 2 or 3 years. The symptoms, history and radiological findings of our case were consistent with that of the literature. The main point to be emphasized is that the case was followed-up and managed for up to the age of 60 for chronic bronchitis depending only on her symptoms and neglecting the right hyperlucent lung appearance, and in the last 6 or 7 years, followed-up and treated for COPD-cor pulmonale since the diagnosis of Swyer–James (Macleod) syndrome may be made by routine radiographic investigation.

In order to rule out malignancy, a computerized tomography was performed. Computerized tomography findings revealed no mass or findings that would make us think about pulmonary embolism. The presence of a small ipsilateral hilus renders it to be differentiated from pulmonary artery agenesis. The primary pathology is diffuse obstruction in the peripheral airways. Vascular changes occur secondarily. On pulmonary angiography, pulmonary artery branches were few in number and had narrow diameters. On anteroposterior chest X-ray, there may be an observation of a decrease in hilar and pulmonary vascular shadows in addition to an increase in lucency. Owing to underdevelopment of the lung on the affected side, the lung volume may be found to be decreased or normal. Chest X-ray and thoracic CT demonstrated no difference between the affected and normal lung volumes. On thoracic CT, bronchiectasis and atelectasis were found. Furthermore, there was a decrease in the number of right lower pulmonary vascular structures. No lesion that may have caused increased local ventilation by obstructing the bronchi was observed on thoracic CT.

Respiratory function tests in cases with Swyer–James (Macleod) syndrome have been reported to show mild syndrome have been reported to show mild increase in lucency. In this case, a severe syndrome have been reported to show mild decrease in hilar and pulmonary vascular shadows in addition to an increase in lucency. Owing to underdevelopment of the lung on the affected side, the lung volume may be found to be decreased or normal. Chest X-ray and thoracic CT demonstrated no difference between the affected and normal lung volumes. On thoracic CT, bronchiectasis and atelectasis were found. Furthermore, there was a decrease in the number of right lower pulmonary vascular structures. No lesion that may have caused increased local ventilation by obstructing the bronchi was observed on thoracic CT.

Coronary angiography, which was performed because of prevailing ST and T changes revealed single ostium coronary artery anomaly. Cardiovascular system anomalies concomitant with Swyer–James (Macleod) syndrome are rare. Two cases with muscular bridge and ventricular septal defect have been reported. As far as we know, there have been no reports regarding a coronary artery origin anomaly accompanying this syndrome.

According to coronary angiography data, the incidence of coronary artery anomaly in adult age has been reported to be about 1%. The most unusual form of coronary artery anomalies is a single coronary artery (8.8%). In this case, the left coronary artery was seen to have originated from the right aortic sinus. In this case, the blood supply of the heart would be through a coronary artery with a single ostium. This anomaly is quite important clinically as it may result in myocardial ischemia, life-threatening ventricular arrhythmia and even sudden deaths depending on the anatomic anomalies of the coronary artery.

In conclusion, cases reported with unilateral pulmonary hyperlucency on radiography should remind the clinician of a rare disease called Swyer–James (Macleod) syndrome. Coronary arteries originating from a single ostium may accompany this as a life-threatening congenital anomaly.

Conflict of interest statement

We declare that we have no affiliation with or financial involvement in any organization or entity with a direct financial interest in the subject matter or materials discussed in the manuscript.

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