Regional ventilation/perfusion mismatch pattern in patient with Swyer James (MacLeod’s) syndrome

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Swyer James (McLeod’s) syndrome (SJMS) is an uncommon disease, which occurs as a result of childhood bronchiolitis obliterans. Patients may not be diagnosed until later in their life. A 46-year-old man underwent ventilation/perfusion scintigraphy for acute onset of dyspnea. The scan showed markedly diminished ventilation and perfusion unilaterally on the right middle and inferior lobes. However, mismatched ventilation-perfusion pattern was shown on the upper right lobe, which was consistent with pulmonary embolism. Unilaterally matched ventilation/perfusion defect can see in SJMS in lung scintigraphy; however, when pulmonary embolism may accompany, scintigraphy should be carefully examined.

Key words: Mcleod’s syndrome, pulmonary embolism, scintigraphy, Swyer James

INTRODUCTION

Swyer James (McLeod’s) syndrome (SJMS) is a rare condition (was seen 0.01% in 17459 cases) characterized by unilateral hyperlucency of a part of or the entire lung, which was first described by Swyer and James than MacLeod.[1,2] First, this syndrome was thought to be congenital in origin and was attributed to hypoplasia of the pulmonary artery. SJMS is a variant of postinfectious bronchiolitis obliterans that is most commonly the sequela of a viral (adenovirus or RVS) infection during infancy or early childhood. Mycoplasma infection has also been implicated.[3] SJMS may be seen following measles, pertussis, tuberculosis, and mycoplasma infections.[4]

CASE REPORT

A 46-year-old male patient was admitted to Istanbul University, Cerrahpasa Medical Faculty, Department of Chest Disease, Istanbul in 2009 with dyspnea and chest pain. While examination of the patient it is learned that he had a history of multiple respiratory infections in childhood. Chest examination showed diminished breath sounds over the right hemithorax. Chest X-ray (CXR) showed hyperlucency on the lower part of the right hemithorax [Figure 1].

Thorax computed tomography (CT) revealed bronchiectatic changes, hyperlucency of the lower part of the right hemithorax due to hypoplastic left pulmonary artery [Figure 2]. Thus, diagnosis of SJMS associated with right lung middle lobe hypoplasia was made.

Patient referred to the nuclear medicine department for ventilation-perfusion scintigraphy to rule out pulmonary thromboembolism. Ventilation/perfusion scintigraphy (VQ) was performed in the same day. Technetium-99m-labeled macroaggregated human albumin (99mTc-MAA) perfusion scan was performed after the injection of 185-200 MBq of 99mTc-MAA into an anterocubital vein, and Tc-99m labeled carbon particles (Technegas) was performed after inhalation of technegas with tidal breathing in upright position for ventilation scintigraphy. Planar images and single photon emission computed tomography images of each scan were obtained by a double-headed gamma camera. VQ images showed severe unilaterally matched VQ defects on the right middle and inferior lobes. However, mismatched ventilation-perfusion pattern was shown on the upper right lobe, which was consistent with pulmonary embolism [Figure 3].

DISCUSSION

Swyer–James–MacLeod syndrome is a rare lung disorder, which occurs as a result of childhood bronchiolitis obliterans. The onset of symptoms typically occurs during infancy or early childhood.
involved lung or portion of the lung does not grow normally and is slightly smaller than the opposite lung. The characteristic radiographic appearance is that of pulmonary hyperlucency, caused by over distention of the alveoli in conjunction with diminished arterial flow. In general, this disorder is diagnosed in childhood after evaluations for recurrent respiratory infections.[5] After the initial infective insult, radiographic findings appear after months to years. Usual investigations performed are chest radiograph, high-resolution CT, magnetic resonance imaging, angiography, ventilation-perfusion scanning.

Swyer–James–MacLeod syndrome is usually diagnosed with chest radiography which shows hyperlucency of a lung lobe and a small hemithorax on the affected side. However, CXR can be insensitive and may appear normal. Despite characteristic findings by chest radiography, CT is the most valuable imaging technique in the diagnosis of SJMS. When using CT scan peribronchial thickening and the diagnosis of bullae and subsequent air trapping can be seen.[3]

Swyer–James–MacLeod syndrome is often referred to as hyper- or trans-lucent unilateral lung because of the decreased parenchymal perfusion.[6] Although one lung or one lobe can be affected, CT has made it apparent that the disease may be more heterogeneous in distribution and contralateral parenchymal lesion may be present.[7]

Ventilation/perfusion scintigraphy is a diagnostic modality that performed to rule out pulmonary embolism and shows a decrease in ventilation to the affected lung, secondary to emphysematous changes and a matched decrease in perfusion.[9] In our patient, VQ scintigraphy showed matched defect in the affected right middle and lower lobe.

Pulmonary angiography is extremely valuable in showing narrowed and withered tree-like pulmonary arteries in the affected side. Magnetic resonance (MR) angiography demonstrates smaller pulmonary artery and its branches on the affected side. Serdengecti et al. compared MR angiography with ventilation-perfusion scintigraphy in 11 patients and concluded that the magnetic resonance angiography is a fast, accurate, without radiation, and noninvasive technique for supporting the diagnosis of Stevens–Johnson syndrome.[9]

The important differential diagnosis of the SJMS are congenital lobar emphysema, congenital hypoplasia of the lung, compensatory unilateral emphysema secondary to lobectomy, pulmonary embolic disease, pneumothorax, and foreign body in air way.
AUTHORS’ CONTRIBUTIONS

All authors have contributed in designing and conducting the study. All authors have assisted in preparation of the first draft of the manuscript or revising it critically for important intellectual content. All authors have read and approved the content of the manuscript and confirmed the accuracy or integrity of any part of the work.

REFERENCES


Source of Support: Nil, Conflict of Interest: None declared.