IMAGING GAMUT

Utility of ventilation-perfusion scintigraphy in Swyer-James syndrome

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Key words: Perfusion lung scan, Swyer-James Syndrome, Ventilation lung scan

Background A 36-year-old woman with repeated history of respiratory tract infection, presented with acute shortness of breath and chest pain. Chest X-ray showed a hyperlucent left lung with diminished vascular markings. The patient was referred for lung ventilation-perfusion (VQ) scanning to rule out pulmonary embolism.

Procedure Perfusion lung scanning was performed after intravenous administration of 185 MBq of ^{99m}Tc-macroaggregated albumin, ^{99m}Tc-MAA, and images obtained in multiple projections (anterior, posterior, right and left laterals and both posterior-obliques). Aerosol ventilation scan images were obtained in the same projections following inhalation of ^{99m}Tc-Technegas.

Findings The perfusion lung scan showed diminished perfusion to the left lung. The ventilation scan showed a symmetrical reduction of ventilation in the left lung together with interspersed hot areas most likely representing air trapping (Figure 1).

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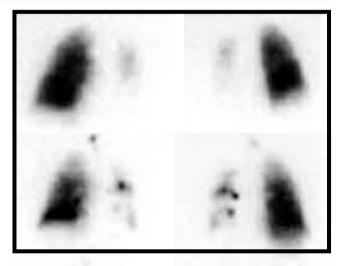


Figure 1 Perfusion scan images (top row) and ventilation scan images (bottom row) in anterior (left) and posterior (right) projections. There is generalised matching reduction of perfusion and, to a lesser extent, ventilation in the left lung with deposition of aerosol in the airways noted on the ventilation scan images

Radiographs and CT scan of the chest showed hyperlucent areas in left lung with diminished vascular markings (Figure 2 & 3).

Conclusion The combination of scintigraphic and radiographic findings suggested a diagnosis of Swyer-James Syndrome. This case highlights the utility of the VQ lung scan as a noninvasive diagnostic tool, which together with the anatomical imaging modalities, plays an essential and complementary role in establishing the diagnosis of Swyer-James Syndrome.



Figure 2 Chest X-ray shows the hyperlucent left lung with reduced vascular markings.

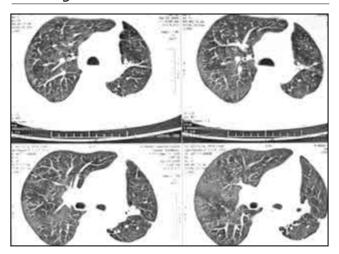




Figure 3 CT scan showing hyperlucent areas and decreased pulmonary vascular markings in the left lung without an anteroposterior gradient attenuation

Comments Swyer-James Syndrome (SJS) is a rare condition and appears to be a consequence of bronchiolitis obliterans associated with bacterial or viral (adenovirus) infection during childhood, which results in subsequent underdevelopment of the lung or a portion of the lung affected [1, 2]. The involved lung or portion of the lung does not grow normally and is slightly smaller than the opposite lung. Typically, it is unilateral and a whole lung is affected, but changes may be confined to a lobe or segment. Most patients are asymptomatic, but some may experience chronic cough, repeated chest infections, haemoptysis and other respiratory symptoms related to associated bronchiectasis [3,4]. This syndrome should be recognized to avoid confusion with asthma and pulmonary embolism, which may result in inappropriate therapy.

The characteristic radiographic appearance is that of pulmonary hyperlucency, caused by overdistention of the alveoli in conjunction with diminished arterial flow [5, 6]. CT scan shows hyperlucent areas due to decreased pulmonary perfusion of the lung without an anteroposterior gradient attenuation [7]. VQ lung scanning is not only essential in establishing the diagnosis in patients with SJS by showing matched ventilation and perfusion defects [8], but has the advantage of demonstrating additional areas of involvement that appear normal on a chest radiograph. Further, scintigraphy is considered to be more sensitive than CT in the identification of the affected lung regions and in delineating the extent and distribution of the involved regions [9]. An awareness of SJS by the reporting physician is important in establishing the correct diagnosis on a VQ lung scan in patients with matching pulmonary perfusion and ventilation defects.

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