J of Nuclear Medicine Technology, first published online May 24, 2022 as doi:10.2967/jnmt.122.264190

Evaluation of Hepatopulmonary Syndrome with Technetium-99m Macroaggregated Albumin Scintigraphy

Short running title: Hepatopulmonary syndrome on MAA scan

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Disclosures: None

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Word count of the manuscript: Previously 750, now after revision and adding more references: 822 (Including title, abstract, keywords, introduction, case report, discussion, conclusion, disclosure, references, figure legends)

Financial support for the work: None

Abstract

Hepatopulmonary syndrome (HPS) is characterized by intrapulmonary vascular dilatation causing hypoxemia in patients with liver disease. Technetium-99m macroaggregated albumin (99mTc-MAA) scintigraphy have diagnostic value in suspected HPS by detecting clinically significant right to left shunt. In presence of cirrhosis, MAA scan with extra-pulmonary organs visualization is specific of intrapulmonary shunting. MAA scintigraphy also provides added value of quantification of the shunt.

Keywords

Hepatopulmonary syndrome, 99mTc-MAA scintigraphy, intrapulmonary shunt

Introduction

We report a case of hepatopulmonary syndrome (HPS) confirmed with Technetium-99m macroaggregated albumin (99mTc-MAA) scintigraphy. HPS is an uncommon condition where there is hypoxemia due to intrapulmonary vascular dilatation (IPVD) in the context of liver disease.

Case Report

A 38-year-old male presented with dyspnea on exertion and thrombocytopenia. The patient was extensively evaluated including MRI and liver biopsy, which demonstrated cirrhosis. Patient had consistent low partial pressure of oxygen (PO2) ≈55. Alveolar arterial oxygen gradient ($P(A\text{-}a)O_2$) was 25 mm Hg. A transthoracic echocardiogram with saline showed microbubbles in left heart chambers after four cardiac cycles

following contrast appearance in the right heart suggestive of extracardiac shunt likely pulmonary arteriovenous malformation. 99mTc-MAA lung scintigraphy after injection of 144.3 MBq (3.9 mCi) of radiotracer showed increased tracer uptake in the brain, kidneys, spleen and subcutaneous tissues indicating right to left shunt, likely intrapulmonary shunt in setting of cirrhosis (Fig. 1). On quantification, brain shunt fraction was 20.3% (Fig. 2). Elevated alveolar-arterial gradient with pO2 <60, echocardiographic and scintigraphic evidence of intrapulmonary shunting with no known chronic lung disease in setting of cirrhosis were consistent with hepatopulmonary syndrome in our patient. The patient is currently undergoing evaluation for a liver transplant.

Discussion

The pathognomonic intrapulmonary vascular dilatations in HPS causes impaired oxygen transfer from alveoli to red blood cells, inducing intrapulmonary right-to-left shunt. In HPS, hypoxemia and dyspnea may increase in the upright position due to preferential perfusion of dilated vessels in lung bases (*1*). Severity of HPS based on degree of hypoxemia is described as mild, moderate, severe and very severe if PaO_2 is ≥80, 60-79, 50-59, <50mm Hg respectively. HPS is frequently underdiagnosed (*2*). Currently, only effective treatment for HPS is liver transplantation. It is important to diagnose HPS at the earliest to expedite the treatment for better outcome. Bubble echocardiography with arrival of bubbles in left heart after ≥3 cardiac cycles following contrast appearance in the right heart or 99mTc-MAA scintigraphy showing uptake in the brain can distinguish hypoxemia from HPS and other etiology (*3*). MAA scintigraphy is more specific than echocardiogram and also can quantify and measure the degree of shunt.

Under good quality control measures and absence of shunt, no extrapulmonary organs should be visualized as the injected MAA particles are trapped in the pulmonary microvasculature. However, when there is intrapulmonary shunt, fraction of the particles enter systemic circulation leading to visualization of other organs and systems. The standard technique of pulmonary-brain-shunt percent calculation is done by drawing regions of interest around the brain and lungs and determining the geometric mean (GM) of brain and lung counts (*4,5*). HPS is suggested if $P(A\text{-}a)O_2$ is ≥15 mm Hg or ≥20 mm Hg for patients >64 years and quantitative index MAA ≥6%. A whole body 99mTc-MAA uptake calculation is another method for detecting IPVD (*6*).

Conclusion

Patients with history of chronic liver disease along with dyspnea should be further evaluated for possible hepatopulmonary syndrome. In patients with elevated alveolar arterial gradient, bubble echocardiography or 99mTc scintigraphy aids in diagnosis of intrapulmonary shunt. Though echocardiography is sensitive, it lacks specificity by providing false positive results in patients with concomitant lung diseases. A positive MAA scan with tracer uptake in extrapulmonary organs in a cirrhotic patient is specific for HPS. MAA scintigraphy also quantifies the extent of shunt.

Disclosure

No conflict of interest

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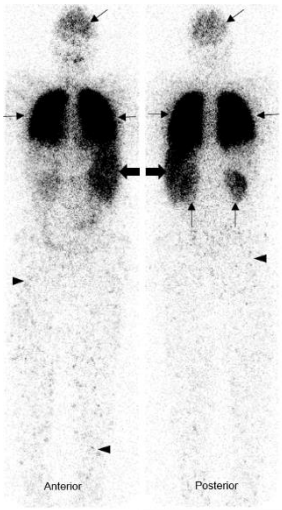

Figure 1: 99mTc-MAA planar whole body image in anterior and posterior projections show intense radiotracer uptake in the lungs (thin transverse arrows) and shunted activity in brain (oblique arrows), spleen (thick arrows), kidneys (vertical arrows) and subcutaneous tissues (arrowheads).

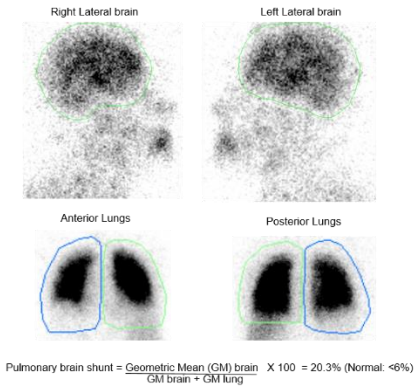

Figure 2: Planar images of the brain in right and left lateral projections and of lungs in anterior and posterior projections show areas of interest drawn to calculate pulmonary-brain-shunt.