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Case Report/Olgu Sunumu

The Use of Lung Perfusion Scintigraphy to Diagnosis of Hepatopulmonary Syndrome

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ABSTRACT: Hepatopulmonary syndrome (HPS) is an uncommon clinical situation of unknown cause.We presented a case with chronic liver disease known for 4 years, who developed clubbing and hypoxemia. On the searching for the cause of these clinical situation, hepatopulmonary syndrome was diagnosed by contrast echocardiography and MAA lung perfusion scintigraphy. We would like to point out that in hipoxemic cirrhotic patients with intrinsic lung disease, lung perfusion scan is needed to detect the contiribution of HPS to hypoxemia.

Key words: Hepatopulmonary syndrome, lung perfusion scan

CASE REPORT

A 21-year-old patient with known cirrhosis for 4 years suffered from dyspnea. Physical examinations revealed the findings of chronic liver disease, including hepatomegaly, ascites and cuteneous spider nevi. In addition to cirrhotic findings, clubbing of the digits, cynosis of the extremities was detected. Chest X-ray demonstrated mild cardiomegaly. Contiguous arterial blood gas tests confirmed hypoxemia (PaO2< 60 mm Hg). After excluding obvious cardiac and pulmonary causes, a series of studies was planned.

Contrast echocardiography was performed by agitated saline creating microbubbles at least 15 micrometer in diameter. Saline was injected via intravenous line on the right upper extremity. Parasternal four-chamber view was used for the detection of microbubbles. After four beats, microbubbles was visualizated in the left heart chambers indicating intrapulmonary right-to-left shunting.

Lung perfusion study was performed after injection of 3 mCi Tc99m MAA

(macroaggregated albumin) via a peripheral vein. 90 % of the MAA particle size ranged between 10 and 90 micrometer. Using Elcint SPX-6 gamma camera equipped with low energy, general purpose collimator, anterior and posterior chest images were obtained. 20 % window centered on the 140 Kev photopeak of technetium-99m. For each views 500 kcount were collected. After lung views obtained, additional images from head and kidney were taken. At the end of the study a whole body scanning was performed.The uptake of radiolabelled macroaggregates over the kidney, the brain was demonstrated suggesting transit through

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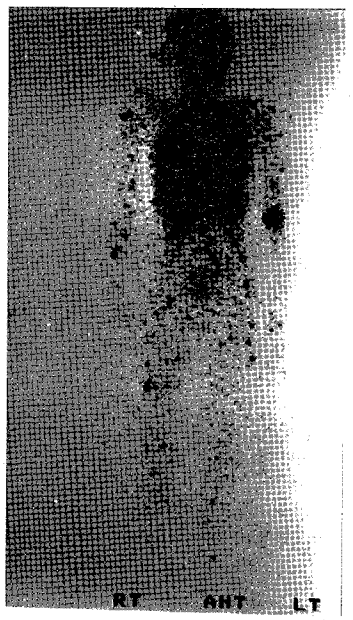

Figure 1: Whole body image of patient show distribution of radioactive particles on body.

the lung caused by either an intrapulmonary or an intracardiac shunt.

On the results of two modilities; hepatopulmonary sydrome was diagnosed.

DISCUSSION

The cause of hepatopulmonary sydrome is unknown. HPS which is characterized by arterial hypoxemia with vascular dilatations in patients with chronic liver disease who don't have significant pulmonary and cardiac disease(1,2). HPS manifest clinically with progressive dyspnea, cynosis and clubbing in patient with cirrhosis.The pathophysiology of this sydrome depends on the degree of pulmonary vascular dilatation in the absence of architectural damage. These vascular abnormalities included precapillary dilatation, arterial-venous communications and dilated pleural vessels. Several mechanism postulated to explain for pulmonary vascular dilatations. These are the faliure of clerance of circulating pulmonary vasodilatators by damaged liver, faliure of inhibition of vasoconstrictive substance by cirrhotic liver. Arterial hypoxemia results from red blood cell passing through the lungs without being enough oxygenated resulting in a functional intrapulmonary right to left shunts (3-6). The degre of the dilatations determinates the severity of the gas exchance abnormalities. Approximately 10 % patients have full criteria of HPS and 30 % patients with normal gas exchance with intrapulmonary dilatation (7).

The diagnosis of HPS based on the demonstration intrapulmonary shunting. Contrast echocardiography, lung perfusion scintigraphy and catheterization of the right side of the heart or pulmonary arteriography are used to the diagnosis of HPS (8-12).

Alhough hepatopulmonary sydrome was reported only 10 % of the cirrhotic patients, in the case of some symptoms including progressive dyspnea, clubbing, spider nevi should suggested HPS. Confirmating of the diagnosis can be made using one of three imaging modilities: contrast echocardiography, lung perfusion scintigraphy and pulmonary arteriography.

Pulmonary arteriography is less widely used because of the invasiveness.Constrast-echocardiography has been shown as a valuable tool for detecting presence of intrapulmonary vascular dilatations in patients with hypoxemia and liver disease. When normally saline injected peripherally, these bubbles trapped during the first pass in the pulmonary capilleries and reabsorbed (9,10).Bubbles visualized after four cardiac cyles after leaving right chamber evaluated as delayed. In the presence of pulmonary or intracardiac shunting, microbubbles will opacifiy the left heart chambers.The distinction between intracardiac and intrapulmonary shunts depends on the timing of the apprearance of

bubbles in left cardiac chambers. In intracardiac shunting, bubbles appear within three beats after visualization of right cardiac chambers. In the presence of intrapulmonary shunting, apperance of bubbles in left cardiac chambers delay and visualization occur four or six beats after the initial apperance of bubles in the right side of heart (13.14).

The lung perfusion study is another method of detecting intrapulmonary vascular dilatations.The majority of injected radioactive-labeled particles normally trapped in the pulmonary vascular bed and lungs pick up the particles. In the presence of the intrapulmonary or intracardiac shunting, the labelled particles is not trapped only lungs also is taken up in the brain, kidney and liver. The magnitude of shunt is estimated by the ratio of systemic to total body activity of the radionuclide (11,12,15,16). In this case we easly detected particles uptake on kidney, brain and whole body.

Contrast echocardiography is accepted to be most sensitive test in the diagnosis of hepatopulmonary syndrome in hypoxemic cirrhotic patients without intrinsic lung disease or chest radiograph abnormalities. However in cirrhotics with intrinsic lung disease or chest radiograph abnormalities, positive echocardiograms can not be considered certain. A positive lung scan with or without chest radiograph abnormalities significantly suggest the significant intrapulmonary vasodilatation is present. A negative lung scan does not exclude the presence of HPS but it may suggest that vasodilatation is relatively less important (13,17).Alhough many authors concluded that contrast echocardiography is the most useful test for intapulmonary vasodilatation, it lacks of specificity because of many cirrhotic cases with normal arterial blood gases and don't have fulfill criteria of HPS may have positive echocardiographic results. A positive MAA scan result in cirrhotic is specific for the presence of moderato to severe HPS. Abrams studied in 25 cirrhotic patients without HPS and 15 hypoxemic subjects with intrinsic lung disease.He showed that strong inverse correlation between the degree of the lung shunt farction and arterial hypoxemia (15). In conclusion, we support the following algorithm for the evaluation of HPS. A negative contrast echocardiogram rules out the diagnois of HPS. A positive contrast echocardiography in hypoxemic cirrhotic without intrinsic lung disease strongly suggests the diagnosis of HPS. In a hypoxemic cirrhotic patients with intrinsic lung disease may be supplemented by a lung perfusiin scintigraphy. MAA lung scan may be particulary useful in evaluating the contribution of HPS to the hypoxemia in cirrhotic patients with intrinsic lıng disease.

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