간폐증후군

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The Hepatopulmonary Syndrome

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간폐증후군은 간질환, 폐환기의 이상으로 야기되는 동맥 저산소혈증, 그리고 광범위한 폐혈관 확장의 3징으로 정의된다. 특히 폐모세혈관의 비정상적 확장에 의한 폐동맥과 폐정맥의 직접적인 교통은 우-좌 단락을 야기하며 이로 인한 동맥 저산소증은 호흡곤란의 원인이 된다. 조영 심초음파는 비침습적으로 이러한 우-좌 단락을 확인 및 진단할 수 있는 유일한 방법으로 사료된다. 본 증례는 호흡 곤란을 호소하는 63세의 간경변 환자에서 조영 심초음파를 이용하여 간폐증후군을 진단한 증례이다.

중심 단어 : 간폐증후군 · 조영 심초음파 · 우-좌 단락.

Introduction

The hepatopulmonary syndrome (HPS) is defined as the triad of liver disease, pulmonary gas exchange abnormalities leading to arterial deoxygenation, and widespread pulmonary vascular dilatation.[1] Although both acute and chronic liver diseases have been associated with HPS, most commonly it is associated with cirrhosis. Portal hypertension seems to be the predominant factor related to this syndrome.[2] The hallmark of pulmonary vascular changes in HPS is dilated vessels at the precapillary and capillary level and direct arteriovenous communications. This causes right-to-left shunting of blood flow, mismatch between ventilation and perfusion, and diffusion limitation. Contrast transthoracic echocardiography may be the only diagnostic test to demonstrate right-to-left shunt in these patients.[3] It is performed by intravenous injection of agitated saline as an echocardiographic contrast, while visualizing the atria. In the case of HPS, contrast appears in the left atrium 2 to 5 s after it is seen in the right atrium.

논문접수일 : 2005년 11월 21일
심사완료일 : 2005년 12월 21일
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Case Report

A 63-year-old woman was evaluated for progressively worsening exertional dyspnea. Her past medical history was unremarkable except liver cirrhosis for 10 years. The electrocardiogram revealed sinus rhythmwith early transition in V1. Pulmonary function test was normal. Transthoracic echocardiography showed borderline enlarged left atrium but normal sized other cardiac chambers with normal left ventricular systolic function (ejection fraction 68%). HPS was suspected because of a long history of liver cirrhosis. Contrast echocardiography was performed by intravenous injection of hand-agitated saline while visualizing the atria. Contrast appears in the left atrium 3 to 5 seconds after it is seen in the right atrium, suggestive of pulmonary arteriovenous fistula (Fig. 1).

Discussion

HPS is defined as the clinical triad of advanced liver disease, arterial deoxygenation and intrapulmonary vascular dilatation.[1] HPS is a well-recognized complication of liver cirrhosis, but its pathogenesis is not completely understood. It is one of pathophysiologic mechanism that pulmonary capillary is dilated upto 500 μm (normal range : 8-15 μm) and the vascular resistance reduced, so abnormal hyperdy-

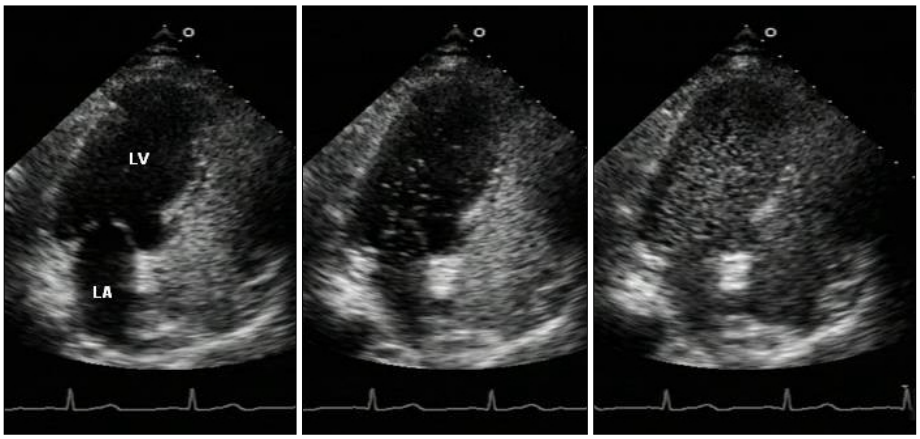

Fig. 1. Contrast echocardiography in hepatopulmonary syndrome. Delayed opacification of the left-sided heart (5 beats after the opacification of the right-sided heart in this figure) was showed during contrast echocardiography using hand-agitated saline.

namic pulmonary circulation results in shunting of deoxydized venous blood into systemic arterial network. Excessive pulmonary nitric oxide production seems to be one of the factors that contribute to the intrapulmonary vascular dilatation. Other mediators such as endothelin-1 and the heme oxygenase-1/carbon monoxide system have recently been found to be important contributors.[4)]

The prevalence of HPS among patients with liver cirrhosis has been reported to be between 5.3 and 17.5%.[5)6)] HPS may also occur in liver diseases such as acute viral hepatitis and fulminant hepatic failure.[7)8)] A few cases with noncirrhotic portal hypertension complicated by HPS has also been re-ported.[9)10)]

The major clinical manifestations are arterial hypoxemia, clubbed fingers and spider nevi. Orthodeoxia is also the characteristic clinical feature.

For the detection of intrapulmonary vascular dilatations and shunting, contrast echocardiography is the method of choice because it is of easy execution and it can differentiate between the intrapulmonary and intracardiac communications.[3)11)] In the case of HPS, contrast appears in the left atrium 2 to 5 s after it is seen in the right atrium, while simultaneously or within 2 heart beats in intracardiac shunt. Our case was also approved by hand-agitated saline contrast echocardiography.

99m Technetium macroaggregated albumin (Tc-99m MAA) lung perfusion scan can further specify the diagnosis of HPS and quantify the magnitude of shunting. On the other hand, Koksal and collegues have reported that high resolution computed tomography (HRCT) may be helpful in the diagnosis of HPS by demonstrating the dilated peripheral pulmonary vessels or increased pulmonary artery to bronchus ratios in patients with liver disease and hypoxemia.[12)]

Despite of multifarious trial, no clearly effective medical treatments have been found. Although liver transplantation seems feasible to reverse this situation,[11)] it is associated with increased postoperative morbidity and mortality. Swanson et al reported that preoperative arterial oxygen tension of 50 mmHg or less and Tc-99m MAA shunt fractions of 20% or more are strong predictors of postoperative mortality that can be used to stratify patients with better outcome.[13)] Therefore, early detection of HPS can be related with reduction of morbidity and postoperative mortality.

KEY WORDS : Hepatopulmonary syndrome · Contrast echocardiography · Right-to-left shunt.

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