J Cardiol 2005 Apr; 45(4) : 161–164

Esophageal Varices Without Porto-Systemic Venous Pressure Gradient in a Patient With Post-Pericardiotomy Constrictive Pericarditis: A Case Report

Kazuyuki OZAKI, MD
Makoto KODAMA, MD
Fumio YAMASHITA, MD
Tsuyoshi YOSHIDA, MD
Satoru HIRONO, MD
Kiminori KATO, MD
Yoshifusa AIZAWA, MD,FJCC

Abstract

A 51-year-old woman was admitted with intractable congestive heart failure and progressive anemia. She had undergone mitral valve replacement for mitral regurgitation at age 23 years. Subsequently, her mitral prosthesis was replaced twice due to thrombotic stack and valve insufficiency. Signs of congestive heart failure became evident at age 46 years. Gastrointestinal endoscopy revealed esophageal varices, which were treated by endoscopic variceal ligation. Cardiac catheterization disclosed elevated pulmonary capillary wedge pressure (mean 16 mmHg), right atrial pressure (mean 15 mmHg), and hepatic vein wedge pressure (mean 15 mmHg). She died at age 53 years. Autopsy showed severe congestive liver but not liver cirrhosis. Esophageal varices may progress in spite of the absence of porto-systemic pressure gradient in patients with severely high venous pressure.

J Cardiol 2005 Apr ; 45(4) : 161−164

Key Words

■**Heart failure** ■**Hypertension (portal)**
■**Pericarditis (post-pericardiotomy constrictive)**

INTRODUCTION

Development of esophageal varices depends on the portal hypertension whereas heart failure tends to cause liver cirrhosis and occasionally esophageal varices. However, detailed observations of the pathogenesis are scarce. We investigated the pressure gradient between the portal circulation and systemic central vein in a patient with chronic intractable heart failure complicated with esophageal varices.

CASE REPORT

A 51-year-old woman was admitted to our hospital with intractable congestive heart failure and progressive anemia. She was treated under a diagnosis of mitral regurgitation at age 11 years. She underwent mitral valve replacement surgery at the 23 years. Subsequently, her mitral prosthesis was replaced twice due to thrombotic stack episode and valve insufficiency. She received an implanted permanent pacemaker because of complete atrioventricular block at age 45 years. Symptoms and signs

新潟大学大学院医歯学総合研究科 循環器学分野 : 〒951−8510 新潟県新潟市旭町通1−757
Division of Cardiology, Niigata University Graduate School of Medical & Dental Sciences, Niigata
Address for correspondence : OZAKI K, MD, Division of Cardiology, Niigata University Graduate School of Medical & Dental Sciences, Asahimachi-dori 1−757, Niigata, Niigata 951−8510 ; E-mail : k-ozaki@med.niigata-u.ac.jp
Manuscript received February 6, 2004 ; revised August 27 and October 12, 2004 ; accepted October 12, 2004

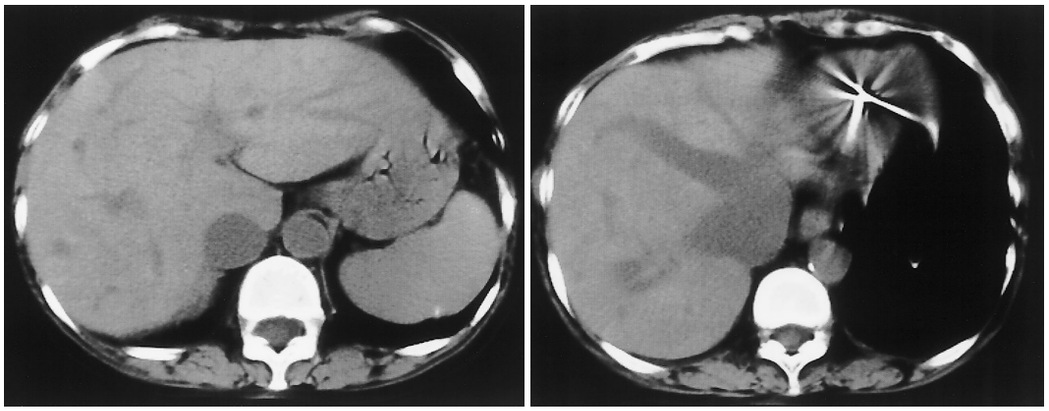

Fig. 1 Abdominal computed tomography scans showing findings of congestive liver, but no findings of liver cirrhosis

of congestive heart failure became evident from age 46 years. Intractable heart failure, progressive anemia and tarry stool appeared 2 years before admission. On admission, she also complained of epigastric discomfort.

Her height was 156 cm and weight was 44 kg. Blood pressure was 104/60 mmHg and heart rate was 70 beats/min. Chest examination revealed no rales. There was a grade 3/6 holo-systolic murmur at the apex. Liver was palpable 3FB under the right costal border and the hepato-jugular reflux was observed. Her abdomen was soft and flat, but fluctuation was not evident. Pretibial edema was not noted. Chest roentgenogram revealed cardiothoracic ratio of 68%. Laboratory examinations did not suggest liver cirrhosis or hepatitis of viral or autoimmune origin. Abdominal computed tomography scanning showed findings of congestive liver, but splenomegaly and ascites were not shown, and there were no findings of liver cirrhosis (**Fig. 1**). Gastrointestinal endoscopy was performed because she complained of epigastric discomfort and revealed esophageal varices (F_2, C_b, L_S, RC sign +) (**Fig. 2**).

Transthoracic echocardiography showed remarkably dilated left atrium (7.6 cm), moderate mitral regurgitation of perivalvular leakage and severe tricuspid regurgitation. Cardiac catheterization disclosed elevation of pulmonary capillary wedge pressure (mean 16 mmHg), right atrial pressure (mean 15 mmHg) and mild left ventricular dysfunction (**Table 1**). Left ventriculography demonstrated second degree mitral regurgitation. Coronary angiography revealed no significant stenosis. Hepatic vein wedge pressure was measured in the

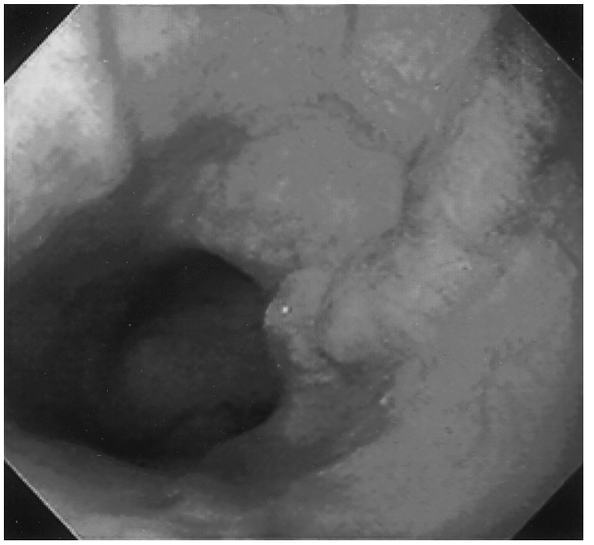

Fig. 2 Gastrointestinal fiberscope image showing esophageal varices

Table 1 Hemodynamic data from cardiac catheterization

Pulmonary capillary wedge pressure	a14v25 (mean 16) mmHg
Pulmonary artery pressure	39/15 (mean 24) mmHg
Right ventricular pressure	36/EDP 13 mmHg
Right atrial pressure	a12v25 (mean 15) mmHg
Left ventricular pressure	98/EDP 14 mmHg
Aortic pressure	99/47 (mean 65) mmHg
Cardiac index	2.9 *l*/min/m^2
Left ventricular end-diastolic volume index	126 ml/m^2
Left ventricular end-systolic volume index	71 ml/m^2
Ejection fraction	44%

EDP=end-diastolic pressure.

right hepatic vein. Mean hepatic vein wedge pressure was 15 mmHg, which implied portal hypertension. Cardiac catheterization was performed when her condition of heart failure was stable. Endoscopic ligation of the esophageal varices was performed. Her esophageal varices disappeared and she returned to the outpatient clinic.

Her congestive heart failure was intractable. One and a half years after discharge, ascites and emaciation became evident. Protein and albumin levels of ascites fluid were 5.2 and 3.5 g/d*l*, respectively, and serum protein and albumin levels were 7.1 and 4.4 g/d*l*. She died of congestive heart failure at age 53 years. At the autopsy, thickened adhesive pericardium which firmly surrounded the heart was observed. The autopsy showed high degree of atrophy, degeneration, and necrosis of the liver cells, and fibrosis of the central zone. The degree of central fibrosis varied significantly from region to region. Thrombosis of hepatic veins and portal veins were not observed. Reconstruction of the lobes was also not observed. Severe congestive liver was found but no evidence of liver cirrhosis. Other esophageal varices were not present. The final diagnosis was post-pericardiotomy constrictive pericarditis with congestive liver.

DISCUSSION

Congestive heart failure affects the liver through two ways: forward failure through decreased hepatic blood flow and backward failure through congestive liver. Both lead to hepatocellular hypoxia. Ischemic damage and passive congestion result in liver fibrosis around the perivenular area and in the space of Disse. The process impairs the blood-to-hepatocyte diffusion of oxygen and nutrients. As a result, cardiogenic liver cirrhosis develops[1–4]. Generally, liver dysfunction occurs in patients with heart failure persisting for 60 days. Fibrosis of the central zone progresses and liver cirrhosis develops after 3 years[5]. Currently cardiac liver cirrhosis is quite rare. The chief causes of cardiac cirrhosis are ischemic heart disease (31%), cardiomyopathy (23%), valvular heart disease (23%), restrictive lung disease (15%), and pericardial disease (8%)[1].

Patients with cardiogenic liver cirrhosis rarely show systemic signs of porto-systemic shunts, such as caput medusae[2]. None of 28 autopsied cases disclosed esophageal varices[6]. These observations may imply that cardiogenic liver cirrhosis is not involved in the porto-systemic pressure gradient. However, chronic and severe right heart failure is believed to involve liver cirrhosis with esophageal varices occasionally[1]. Five cases of esophageal varices related to congestive heart failure without liver cirrhosis were reported among consecutive adult autopsies[4].

Development of esophageal varices depends on the portal hypertension and on the pressure gradient between the portal circulation and systemic central veins. Once liver cirrhosis has developed, portal hypertension may subsequently progress to exceed the congestive central vein pressure. If the pressure gradient between the portal circulation and central vein becomes significant, porto-systemic shunt may develop. Pressure in the esophageal varices is 15.5 mmHg in cirrhotic patients, significantly lower than the main portal pressure of about 18.8 mmHg[7]. Right heart failure actually leads to portal hypertension through congestive liver, but does it produce porto-systemic pressure gradient? There is no low or normal pressure region in the systemic central veins in patients with severe congestive right heart failure. Detailed observation of the pathogenesis of esophageal varices in congestive heart failure is rare.

In this case, we investigated the pressure gradient between the portal circulation and systemic central veins in a patient with chronic intractable heart failure complicated by esophageal varices. Portal hypertension was evident, but the hepatic vein wedge pressure was equal to the right atrial pressure and pulmonary capillary wedge pressure. There was no pressure gradient between the portal circulation and systemic central veins.

The esophageal varices in this case were thought to have appeared with the progression of congestive heart failure. Gastrointestinal endoscopy showed no findings of esophageal varices at age 46 years. From that time, signs of congestive heart failure became evident. Esophageal varices had developed at age 51 years in spite of the absence of porto-systemic venous pressure gradient. Why did the porto-systemic shunt vessels develop in this patient without a pressure gradient? The intermittent pressure gradient may have occurred only after daily postprandial periods. Another hypothesis is that a period of porto-systemic pressure gradient occurred during progression of congestive heart failure. If congestive liver leads to significant portal hypertension in the early phase of congestive heart failure with not so high venous pressure, a period of sig-

nificant pressure gradient would occur. Another possibility is that the venous hypertension and portal hypertension caused dilation of the submucosal veins even in the absence of a pressure gradient.

To clarify the pathogenesis of esophageal varices in patients with severe chronic right heart failure and without porto-systemic pressure gradient, examination of hemodynamics at several time points will be necessary during the progression of congestive liver.

要　　約

門脈系静脈系圧較差が認められずに食道静脈瘤が発現した心膜切開後収縮性心膜炎の1例

尾± 和幸　小 玉 誠　山下 文男　吉 田 剛
広 野 暁　加藤 公則　相澤 義房

症例は51歳，女性．難治性のうっ血性心不全および進行性の貧血により入院した．23歳時に僧帽弁閉鎖不全症により僧帽弁置換術を受けた．その後，人工弁血栓症および人工弁不全により2回の再僧帽弁置換術を受けた．46歳より心不全症状が出現した．入院後，上部消化管内視鏡により食道静脈瘤が認められ，内視鏡的結紮術を施行した．心臓カテーテル検査では，肺動脈楔入圧（平均16 mmHg），右房圧（平均15 mmHg），肝静脈楔入圧（平均15 mmHg）の上昇が認められた．53歳で死亡した．剖検により高度のうっ血肝が認められたが，肝硬変はなかった．静脈圧が高度に上昇した心不全症例では，食道静脈瘤は門脈系と静脈系の圧較差が存在しなくても増悪する可能性が示された．

J Cardiol 2005 Apr; 45(4): 161−164

References

1) Naschitz JE, Slobodin G, Lewis RJ, Zuckerman E, Yeshurun D: Heart diseases affecting the liver and liver diseases affecting the heart. Am Heart J 2000; **140**: 111−120
2) Giallourakis CC, Rosenberg PM, Friedman LS: The liver in heart failure. Clin Liver Dis 2002; **6**: 947−967
3) Sekiyama T, Nagano T, Aramaki T: Congestive(cardiac) cirrhosis. Nippon Rinsho 1994; **52**: 229−233(in Jpn with Eng abstr)
4) Luna A, Meister HP, Szanto PB: Esophageal varices in the absence of cirrhosis, incidence and characteristics in congestive heart failure and neoplasm of the liver. Am J Clin Pathol 1968; **49**: 710−717
5) Shiina Y, Kataoka T, Kounosu M, Kaneko K, Kawazoe K: A case report of successful surgical treatment of valve disease associated with cardiac cirrhosis. Shinzo 2003; **35**: 691−695(in Japanese)
6) Sherlock S: The liver in heart failure: Relation of anatomical, functional, and circulatory changes. Br Heart J 1951; **13**: 273−293
7) Sherlock S, Dooley J: The portal venous system and portal hypertension. *in* Diseases of the Liver and Biliary System, 10th Ed. Blackwell Science, Oxford, 1997; pp 135−180