Laparoscopic Splenectomy for a Patient with Splenomegaly and Hypersplenism due to Idiopathic Portal Hypertension

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Idiopathic portal hypertension (IPH) is a rare disorder which is clinically characterized by portal hypertension, splenomegaly, hypersplenism and the absence of liver cirrhosis. Patients with IPH have massive splenomegaly leading to increased portal venous flow and subsequent portal hypertension. In selected IPH patients with splenomegaly and hypersplenism, splenectomy can be regarded as an effective treatment protocol for decreasing portal hypertension. We report a case of a 44-year-old woman who was diagnosed with IPH accompanied by splenomegaly and hypersplenism. She underwent laparoscopic splenectomy and clinical symptoms and hypersplenism resolved. Our study shows that laparoscopic splenectomy can be considered as a procedure for treating patients with splenomegaly and hypersplenism due to IPH.

Keywords: Idiopathic portal hypertension, Splenomegaly, Laparoscopic splenectomy

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INTRODUCTION

Idiopathic portal hypertension (IPH) is an uncommon disease presenting with clinical symptoms of portal hypertension, such as hepatomegaly, splenomegaly, hypersplenism, pancytopenia, gastric varix, and esophageal varix.¹⁻⁴ Splenectomy is the recommended management procedure for these clinical symptoms of IPH.^{1,5} Laparoscopic splenectomy (LS) has been considered as a relative contraindication for management of massive splenomegaly, because the operation field is too narrow to perform active handling of surgical instruments. However, the improvement of surgical techniques allowed LS with minimal morbidity or mortality. Here we report a case of LS for massive splenomegaly (26 cm/1,517.4 g) generated by IPH.

CASE REPORT

A 44-year old female patient visited the hospital with symptoms including chilling, fever, loss of appetite, and general weakness. In routine physical examination, there was a palpable movable mass located from the left lower abdomen to the upper abdomen. Laboratory findings revealed pancytopenia (WBC 1,030/ μ l, absolute neutrophil count <500, hemoglobin 9.2 g/dl, platelet 100,000/ μ l), elevated liver enzymes (AST 123 IU/L, ALT 61 IU/L) and LDH (808 IU/L). Viral markers for hepatitis A, B, and C were all negative. Abdominal ultrasound and computed tomography (CT) revealed hepatomegaly and massive splenomegaly with multifocal splenic infarctions (Fig. 1A). Based on the results from neck CT, chest CT and bone marrow biopsy, lymphoma was ruled out from the dif-

Fig. 1. (A) Initial CT finding; Contrast enhanced abdominal CT shows marked spleen enlargement with multifocal splenic infarct and suspicious of lymphoma involvement or other hematologic disorder. (B) PET/CT shows diffuse FDG uptake of spleen.

Fig. 2. The histological examination of the liver shows portal inflammation with mild periportal activity (arrow) and moderate lobular activity with ballooning degeneration (arrow head) (hematoxylin and eosin staining, x200).

ferential diagnosis. Diffuse fluorodeoxyglucose (FDG) uptake ($SUV_{max}=3.3$) was shown in positron emission tomography/computed tomography (PET/CT) (Fig. 1B). However, esophageal or gastric varix was not found in endoscopy. Pathology report from the patient's liver biopsy confirmed chronic hepatitis with 1) portal inflammation with mild periportal activity, 2) periportal fibrosis, and 3) moderate lobular activity (Fig. 2).

Authors performed LS to treat hypersplenism due to massive splenomegaly (Fig. 3). The patient underwent operation in the supine position with 4 incisions of trocars (Fig. 4). The enlarged spleen was removed safely via 9 cm of left subcostal incision. The weight and the size of the isolated spleen were 1,517.4 g and 26.4×14.8×4.7 cm, respectively (Fig. 5). There was no specific abnormal finding in pathologic evaluation. After surgery, the clinical symptoms of hypersplenism disap-

peared, and pancytopenia resolved (WBC 8,160/ μ l, hemoglobin 13.3 g/dl, platelet 421,000/ μ l). Six months after surgery, the patient was observed without any abnormalities.

DISCUSSION

Portal hypertension is defined as a portal-caval venous pressure gradient exceeding 5 mmHg. This clinical syndrome leads to the progression of collateral circulations and splenomegaly.¹ The most important causative factor of portal hypertension is hepatic cirrhosis with underlying liver disease. However, portal hypertension can develop without hepatic cirrhosis in various conditions, and this condition is defined as idiopathic portal hypertension (IPH). IPH is generally classified into prehepatic, intrahepatic and suprahepatic portal hypertension based on the site of obstruction.^{1,2}

For the diagnosis of IPH, other liver diseases that can cause portal hypertension should be ruled out. IPH is regarded as sinusoidal portal hypertension of unknown etiology.⁴ Liver biopsy is necessary in IPH to rule out liver diseases that cause hepatic cirrhosis. The presence of periportal fibrosis, obliterated portal venopathy, increased number of portal vascular channels and dilated portal veins herniating into the surrounding parenchyma in the absence of hepatic cirrhosis are the pathologic findings of IPH in liver biopsy.^{1,3,4} Also, as in the present case, liver biopsy can demonstrate periportal fibrosis and portal inflammation without hepatic cirrhosis.

General clinical symptoms and complications of portal hypertension can progress to IPH. Sarin et al. reported the clinical symptoms of IPH, which were splenomegaly (13.5%), upper gastrointestinal bleeding (84.5%), esophageal varices (92%), and gastric varices (22.3%).⁶ However, according to the study results in western countries and Japan, splenomegaly or abnormalities of liver enzymes are more common early symptoms of IPH than gastrointestinal bleeding.^{7,8} Splenomegaly in

Fig. 3. (A) Gross finding seen in laparoscopic view showed massive splenomegaly. (B) Dilated splenic vein was cut with laparoscopic linear stapler.

Fig. 4. The patient is positioned in supine. The surgeon work on the right side of the patient. Four trocars are used, two 5 mm and two 12 mm.

IPH shows a much more severe pattern than it does in portal hypertension with hepatic cirrhosis or in portal vein thrombosis. In our case, the major clinical symptoms were splenomegaly, hypersplenism, neutropenia, anemia and no varix, which were consistent with a report conducted in Japan.

Splenectomy can relieve the clinical symptoms of hypersplenism with splenomegaly.^{1,4,9} Pancytopenia is the most important indicator for splenectomy in hypersplenism patients.⁴ Massive splenomegaly had been considered as a relative contraindication for LS because of technical difficulties including the narrow operation field. However, the development of laparoscopic operation devices and surgical techniques have made LS safe for massive splenomegaly. Hama et al. reported that there is no significant difference in LS and hand-assisted laparoscopic splenectomy (HALS) between patients with splenomegaly and those with a normal sized spleen.⁹ Wang et

Fig. 5. Resected spleen measures 1,517.4 g in weight and 26.4x14.8x4.7 cm in dimensions.

al. introduced a modified HALS method with an additional incision of 6~8 cm for removal of the enlarged spleen when the intact spleen is needed for pathologic evaluation or when conventional LS is not appropriate because of the massive size and dense adhesion around the spleen.¹⁰ In the present case, lymphoma was suspected in preoperative abdominal CT, thus, the authors performed LS with an additional 9 cm incision on the left subcostal region followed by intact removal of the enlarged spleen (1,517.4 g, 26.4×14.8×4.7 cm). Remission of IPH was observed after splenectomy.

Splenectomy would be an effective and curative method for IPH patients presenting with splenomegaly and hypersplenism as major symptoms. In addition, considering the advantages of laparoscopic surgery, LS should be recommended even for cases of massive splenomegaly.

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