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A CASE OF CRYPTOGENIC LIVER CIRRHOSIS WITH HUGE SPLENOMEGALY

Minoru Umehara[1)], Shunji Narumi[2)], Michihiro Sugai[1)], Yoshikazu Toyoki[3)], Keinosuke Ishido[3)], Daisuke Kudo[3)], Norihisa Kimura[3)], Tamotsu Kobayashi[1)], and Kenichi Hakamada[1, 3)]

Abstract

Introduction

We never knew how the liver function goes after splenectomy in case of huge splenomegaly because of the dramatic portal flow alteration. In this report, we will present a case of cryptogenic liver cirrhosis with huge splenomegaly.

Case presentation

A 14 years old male was admitted with huge splenemogaly and hepatomegaly. Computed tomography showed enlarged liver and huge spleen that occupied large volume of abdominal and pelvic cavity. His ADL was significantly deteriorated, and the patient could not stand up because of exertional dyspnea. Although the etiology of the patient was not determined in spite of several liver biopsies and laboratorial tests, we planned splenectomy to improve his ADL. Also, the progression of liver insufficiency was expected because the portal flow could be dramatically changed after the removal of the huge spleen. Therefore, we prepared for the living donor liver transplantation at first, and then splenectomy was performed. After the first operation, his general condition was improved and the liver function was not deteriorated for 6 months. Unfortunately, rapid progression of jaundice from his primary disease was occurred, and living donor liver transplantation using left lobe graft from his mother was performed 4 months later. The patient survived the two-stage liver transplantation procedure without major complications.

Conclusion

Our case suggests that we should consider the possibility of liver deterioration after splenectomy for the huge splenomegaly. In case of rapid deterioration of liver function, liver transplantation is the only feasible option.

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Key words: cryptogenic liver cirrhosis; living donor liver transplantation; splenectomy

Introduction

Few reports mention derangement of liver function after splenectomy. In the case of uncontrollable gastroesophageal variceal bleeding due to liver cirrhosis and living donor liver transplantation (LDLT) using a small-size graft, a splenectomy is usually performed to reduce the portal inflow and regulate the portal vein pressure (PVP). However, in the case of huge splenomegaly, it is not possible to predict liver function after splenectomy because of the dramatic alteration in portal flow. Therefore, adequate preparation is necessary in such a situation, because if liver function deteriorates, liver transplantation will be required to save the patient's life.

In this report, we present a case of cryptogenic liver cirrhosis with huge splenomegaly. We prepared for an emergent LDLT before the splenectomy. The patient's general condition remarkably improved after the operation, and

[1)] Department of Pediatric Surgery, Hirosaki University School of Medicine
[2)] Department of Advanced Transplant and Regenerative Medicine, Hirosaki University School of Medicine
[3)] Department of Surgery, Hirosaki University School of Medicine
Address reprint requests to Minoru Umehara
E-mail: mumehara@cc.hirosaki-u.ac.jp
Fax: +81-172-39-5079

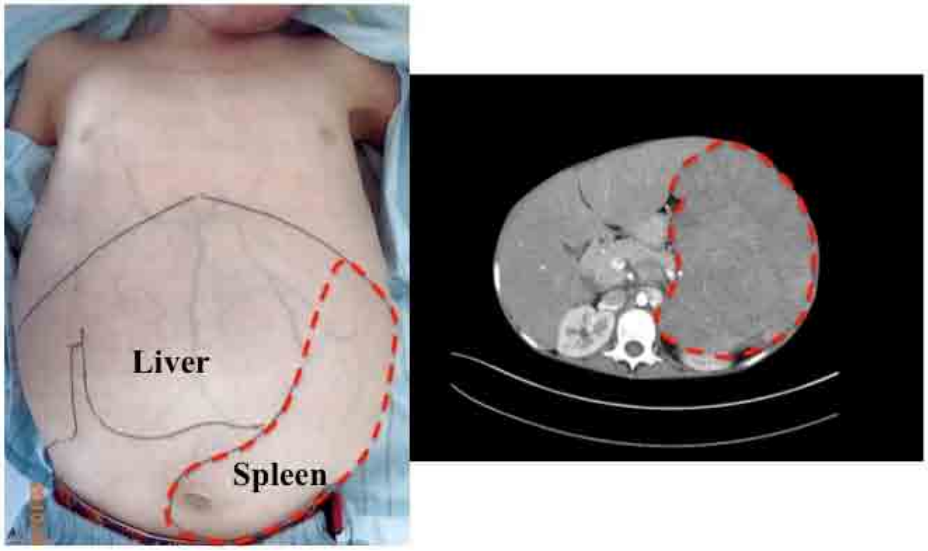

Fig. 1 Huge hepatosplenomegaly. Computed tomography revealed an enlarged liver and a huge spleen that occupied a large volume of the abdominal and pelvic cavities.

10 months later, we could successfully manage the patient with a LDLT performed in a 2-stage operation.

Case presentation

A 14-year-old male was admitted with cryptogenic huge hepatosplenomegaly and recurrent epistaxis due to thrombocytopenia. Computed tomography revealed an enlarged liver and a huge spleen that occupied a large volume of the abdominal and pelvic cavities (Fig. 1). The patient's activities of daily living (ADL) had significantly deteriorated, and he could not stand up because of exertional dyspnea. His medical history began in 2004, but detailed examination, including several liver biopsies, did not result in a confirmed diagnosis. Bone marrow examination revealed foamy cells (a symptom of lysosomal disease), but no abnormal enzymatic activities were detected.

Although the etiology was unclear, we planned a splenectomy to improve the patient's ADL. Progression of liver insufficiency was expected because the portal flow could be dramatically changed after the removal of the huge spleen. Therefore, we prepared for an LDLT before the splenectomy was performed (Fig. 2). PVP was appropriately controlled (pre-splenectomy, 20 mmHg; post-splenectomy, 14 mmHg). Doppler ultrasound examination showed normal hepatic inflow and outflow. After the first operation, the patient's general condition improved, and his liver function did not deteriorate for 6 months. Although liver damage was not detected after the splenectomy, hyperbilirubinemia gradually worsened, possibly due to progression of the underlying disease, and an LDLT was performed 4 months later. The weight of the explanted liver was 2600 g, and a left lobe graft from the patient's mother (400 g, 48.6% of the standard liver volume) was transplanted. The boy experienced 2 episodes of acute cellular rejection (ACR) after the LDLT, but we managed these with steroid pulse therapy and mycophenolate mofetil (MMF). Preemptive therapy was used against cytomegalovirus (CMV) infection. After treatment for ACR, the patient was found to be CMV antigenemia positive (1/150000) without any symptoms. Although administration of

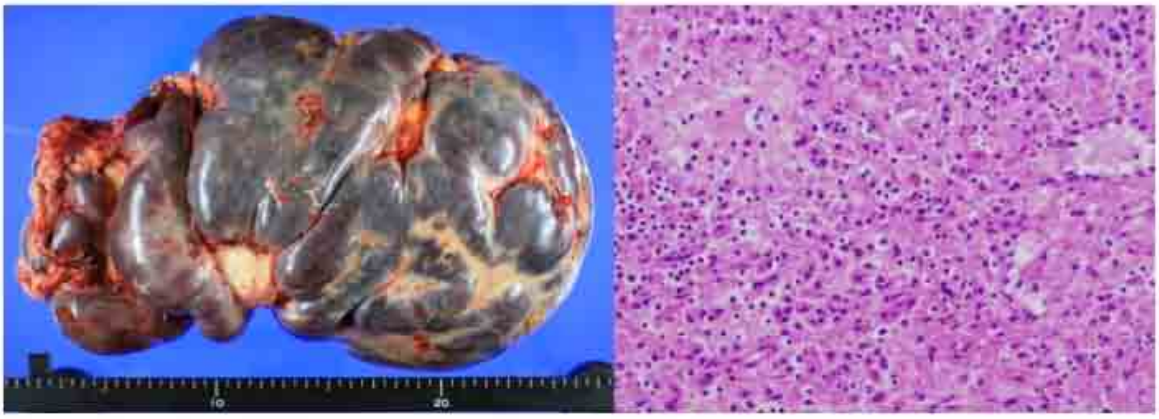

Fig. 2 The weight of the resected spleen was 4125 g. Histological findings were splenic congestion, chronic inflammation, and severe fibrosis. Examination of a liver biopsy specimen showed chronic hepatitis with bridging fibrosis and moderate activity.

ganciclovir (5 mg/kg, every 12 h) was effective, we reduced the dosage due to pancytopenia, which we presumed was an adverse effect of ganciclovir. The patient was discharged on postoperative day 62 with immunosuppressive therapy consisting of tacrolimus (3.5 mg/day) and predonine (5 mg/day). He was subsequently admitted twice for treatment of acute rejection episodes. These were treated with steroid pulse therapy and MMF, and his liver function improved. However, severe thrombocytopenia occurred because of platelet-associated IgG (PAIgG) production. This was managed with steroid therapy and platelet transfusions. The patient is currently doing well 6 months after transplantation.

Discussion

Elevated PVP in the early phase of LDLT is strongly associated with poor patient survival and can be attributed to small-for-size grafts (SFS). Excessive wall shear stress of sinusoidal endothelial cells appears to play a key role in the occurrence of SFS syndrome, which includes massive ascites, meteorism, gastrointestinal bleeding, increased infection, and renal dysfunction. To prevent SFS syndrome, Ogura et al. suggested that PVP be maintained at <15 mmHg.[1] However, there is no indicator for insufficient portal flow. In our casepatient, huge splenomegaly and liver dysfunction coexisted before the operation; this complicated our expectations of the splenectomy results. We thought that splenectomy might reduce the portal flow to the liver and worsen the liver dysfunction. However, the PVP was maintained at 14 mmHg after splenectomy, and no liver deterioration was observed. In case of deterioration, we planned an emergent LDLT to save the patient.

We could not determine the etiology of the patient's liver cirrhosis despite careful examination. Yalamanchili et al. reported that patients who undergo transplantation for cryptogenic or NASH-related cirrhosis do well after transplantation.[2] Despite a greater prevalence of risk factors for cardiovascular disease, both short- and long-term survival are surprisingly similar to those of patients who undergo transplantation for other indications. In case of an unknown underlying disease, recurrence of the disease is an important issue for the survival of both the graft and the patient. In this context, patients who undergo transplantation for cryptogenic cirrhosis are

less likely to die from recurrent liver disease than are those who undergo transplantation for other indications. We have not found any sign of recurrent disease in our patient at present, but close observation and follow-up are still required.

The spleen is the largest accumulation of lymphoid tissue in the body. Indeed, innate splenic function may represent the ultimate defense against bacterial infections. Without the capacity to phagocytize opsonized encapsulated organisms in the spleen, the cumulative risk of a serious infection for a variety of indications is around 33%.[3,4,5] Overwhelming post-splenectomy infection is a serious fulminant process with a high mortality rate. Infections due to encapsulated organisms such as *Streptococcus pneumoniae, Neisseria meningitides, Haemophilus influenzae,* and *Streptococcus pyogenes* lead to uninhibited bacterial overgrowth. Preventive strategies including vaccinations are therefore important for splenectomized patients.

Conclusion

A case of cryptogenic liver cirrhosis with huge splenomegaly was presented. The findings of the present case suggest that the possibility of liver deterioration should be considered after splenectomy for huge splenomegaly. In case of rapid deterioration of liver function, liver transplantation is the only feasible option. The optimal treatment and possible benefit of splenectomy in this context should be discussed among pediatricians, hepatologists, and transplant surgeons.

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