


CASE REPORT

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A successful case of deceased-donor liver transplantation from a donor with Marfan syndrome: a case report

Takuma Ishikawa¹, Shinji Itoh^{1*} , Takeo Toshima¹, Yuki Nakayama¹, Katsuya Toshida¹, Yuriko Tsutsui¹, Norifumi Iseda¹, Takuma Izumi¹, Shohei Yoshiya¹, Mizuki Ninomiya¹ and Tomoharu Yoshizumi¹

Abstract

Background Liver transplantation is the definitive therapy for patients with decompensated cirrhosis. Marfan syndrome is a systemic inheritable connective tissue disease associated with fibrillin-1 gene mutations, which cause abnormalities in connective tissue. Vascular changes due to Marfan syndrome occur mostly in the main vessels due to the high amount of connective tissue within the vessel wall and the high pressure and blood flow to which they are exposed. The incidence of changes in visceral arteries is about 0.42% and usually presents with cystic medial necrosis. This report is the first deceased-donor liver transplantation with a donor with Marfan syndrome with a history of abdominal surgery.

Case presentation A patient in his 50s underwent liver transplantation for decompensated alcoholic cirrhosis. The donor, a 50s male with Marfan syndrome, was diagnosed with brain-death due to a cerebral hemorrhage caused by a cerebral aneurysm. The donor's clinical presentation as Marfan syndrome was aortic dissection, with multiple surgical procedures performed from the aortic root to the abdominal aorta. An intraoperative biopsy of the hepatic artery showed no abnormality, so this organ was considered appropriate. The surgery was completed without any problems of the arterial anastomosis. The patient's postoperative course was uneventful, and he was transferred to a hospital for recuperation on the 18th postoperative day. One year after the surgery, the patient is still alive without any complications from the transplantation or arterial problems.

Conclusions Even if the patient had a history of surgery for vascular anomalies extending to the abdominal aorta due to Marfan syndrome, the patient can be a donor for liver transplantation under appropriate judgment, including intraoperative biopsy.

Keywords Liver transplantation, Deceased-donor liver transplantation, Marfan syndrome

Introduction

Liver transplantation (LT) is universally performed as a treatment for end-stage liver disease, acute hepatic failure, hepatocellular carcinoma, and several metabolic disorders [1–6]. In Japan, the proportion of deceased-donor liver transplantation (DDLTL) in LT is lower than in other countries. As a result, many patients are unable to undergo LT [7].

*Correspondence:

Shinji Itoh

itoh.shinji.453@m.kyushu-u.ac.jp

¹ Department of Surgery and Science, Graduate School of Medical Sciences, Kyushu University, 3-1 Maidashi, Higashi-Ku, Fukuoka-Shi, Fukuoka 812-0054, Japan

Marfan syndrome (MS) is an autosomal dominant inheritance of connective tissue disease with an estimated incidence of 1 in 5000; and in 90% of cases, it is caused by mutations in the gene for fibrillin-1, the main component of extracellular microfibrils. The cardiovascular, ocular, and skeletal systems are the main targets of the disease. Early detection and appropriate management are important because patients with MS are prone to life-threatening cardiovascular complications, such as aortic aneurysms and aortic dissection [8]. Therefore, clinical surgeons may often hesitate to perform LT using MS donors in practical terms. However, it was reported that MS cases are likely to have fewer vascular complications in the hepatic artery and other visceral arteries during abdominal surgery [9].

To date there is only one reported case of LT with MS as a donor, and there is little information on whether it is eligible as a donor for LT. This report was first successful liver transplantation donor in a patient with MS with a history of abdominal aortic surgery.

Case presentation

A male patient in his 50s was on a waiting list for liver transplantation with decompensated alcoholic cirrhosis. The model for end-stage liver disease score was 30, and the Child–Pugh score was 13 (Grade: C). At that time, a brain-dead donor was available. The donor was a man in his 50s and had MS. He had a history of aortic

dissection related to MS and multiple surgeries. He had previously undergone total aortic replacement of the ascending arch, aortic root replacement, descending thoracic aortic replacement, abdominal aortic replacement, and aortic aneurysmectomy plus artificial vessel replacement (reconstruction of celiac artery, superior mesenteric artery, bilateral renal arteries). Therefore, during hepatectomy, the liver was removed up to the normal artery that is not artificial blood vessel, and the common hepatic artery was used for reconstruction. Only the liver was procured from this brain-dead donor. Because a biopsy of the hepatic artery was performed during surgery and was normal, this organ was considered suitable. The DDLT procedure proceeded without problems and the arterial anastomosis was completed. The operation duration was 621 min, blood loss was 804 ml, and the type of implantation was conventional, recipient's proper hepatic artery and the donor's proper hepatic artery were anastomosed with interrupted by using 8-0 Prolene (totally 12 sutures). The postoperative course was uneventful and a daily ultrasound Doppler examination for 2 weeks confirmed that blood flow in the anastomosed vessels was normal. A computed tomography scan 2 weeks after DDLT also confirmed that there were no problems (Fig. 1). The patient was discharged on the 18th postoperative day without major post-implantation complications. One year after surgery, the patient's systemic condition is still intact.

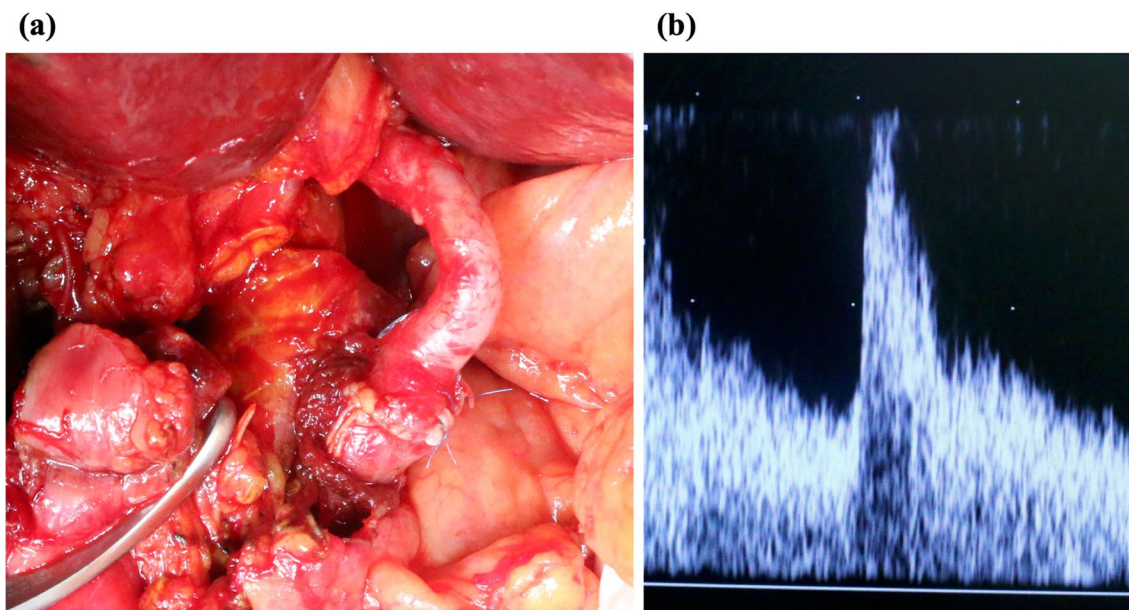


Fig. 1 **a** Anastomosis of the recipient's common hepatic artery with the graft's common hepatic artery. **b** Evaluation of blood flow in anastomosed arteries by Doppler ultrasound

Table 1 Liver transplantation using a deceased donor with Marfan syndrome

Case	Donor				Recipient		
	Age/sex	MS comorbidities	Cause of brain death	Biopsy of hepatic artery	Age/sex	Etiology of liver disease	MELD
No. 1 (Ref. 10)	10s female	Mitral valve deviation, and osteoarticular abnormalities	Hypoxic encephalopathy	No abnormality	40s male	HCV, HCC	N/A
No. 2 (the present case)	50s male	Aortic dissection	Cerebral hemorrhage	No abnormality	55 male	Decompensated alcoholic cirrhosis	30

HCC: hepatocellular carcinoma, HCV: hepatitis C virus, MELD: model for end-stage liver disease, MS: Marfan syndrome,

N/A: not available

Discussion

We experienced DDLT with a donor with MS. DDLT with a donor with MS is rare, with only one case reported in the past (Table 1) [10]. Vascular changes due to MS occur mostly in the main vessels (30–50% incidence). This is associated with the high amount of connective tissue within the vessel wall and the high pressure and blood flow to which the artery is exposed [11–13]. The wall of the aorta is composed of three layers, with a large number of elastin fibers in the tunica media in the middle. The tunica media provides elasticity to the arteries and binds the outer and inner membranes together. In MS, the fibrillin-1 gene is abnormal, which prevents the formation of elastin fibers in the tunica media, making the aortic wall brittle. As a result, aortic aneurysms are more likely to form, and aortic dissection is more likely to occur [14]. When aortic cannulation for perfusion, we need careful cannulation for possible dissection of abdominal aorta because of possible abnormality in the structure of aorta. However, in this case, the common iliac artery had normal elasticity on intraoperative palpation, and the lumen did not feel any change from normal. We think that good perfusion was achieved by careful and careful debridement to avoid divergence and intimal detachment. The incidence of visceral artery changes is approximately 0.4–2.0% and usually manifests with cystic medionecrosis in MS [11–13]. If the biopsy result was strong degeneration with cystic medionecrosis, it is not suitable for anastomosis and liver transplantation may not be performed as planned. The frequency of aneurysms of abdominal viscera is around 0.01–0.2% at autopsy. The patient will continue to undergo computed tomography scan and other imaging evaluations.

In the literature, only two hepatic artery complications are described in patients with MS [15, 16]. Santiago-Delpin described a case of incomplete MS with multiple aneurysms of the aorta and its branches, in which a hepatic artery aneurysm perforated into the common bile duct. Ruschen published a case in which a spontaneous hepatic artery rupture occurred in a patient with MS.

In the present case, our rationale to accept a donor with MS was based on the low incidence of hepatic artery complications and total absence of hepatic dysfunction in patients with MS. The normal hepatic artery biopsy was also important in the acceptance process. Fibrillin-1 gene mutations persist in the transplanted graft and, theoretically, may increase the risk of vascular complications such as dilation, aneurysmal changes, dissection, and intimal tears. Farese et al. [17] suggested another potential complication occurring in this setting: the possibility of antibodies developed against mutated fibrillin-1 donor liver cells. To date, we have not detected any posttransplant liver dysfunction and the recipient has not presented graft rejection. Three cases of kidney transplantation from MS have been reported. These three patients showed good progress with no postoperative complications [17]. In a case of heart transplantation, Marfanoid aneurysm in donor aorta after transplantation was reported [18]. This report was that the donor has the possibility to be MS due to his physique and other factors, and organ transplantation with main vessels from MS patients may have a certain risks but is feasible.

In summary, a patient with MS and a history of abdominal aortic surgery can still be a donor for DDLT with appropriate judgment. Considering reports of LT using MS donors are infrequent, a biopsy should be performed when transplanting a liver from an MS patient. In addition, a detailed follow-up of the transplanted vessels by Doppler ultrasound is advocated for patients undergoing such transplants [10]. In addition, before organ procurement, recipients should receive adequate informed consent regarding MS. There is no information on the number of DDLT using MS as a donor or their long-term outcome, and this information needs to be gathered in the future.

Conclusion

We experienced a successful case of DDLT in which a patient with a history of abdominal aortic surgery due to MS was used as a donor. Patients with MS and a history

of abdominal aortic surgery can still be donors for DDLT with appropriate judgment based on an intraoperative hepatic artery biopsy.

Abbreviations

DDLT	Deceased-donor liver transplantation
LT	Liver transplantation
MELD	Model for end-stage liver disease
MS	Marfan syndrome

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Author contributions

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Competing interests

The authors declare no conflicts of interest in association with the present study.

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