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Case Report:

The First Total Situs Inversus Patient Treated with Extended Two-Step Hemihepatectomy

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Abstract

Background: Liver resection is the treatment of choice for patients with primary or secondary liver tumors restricted to the liver. Resection is limited, however, by the need to preserve a sufficient amount of functional remnant liver. If future remnant liver volume (FRLV) is deemed to be small for extended right hepatectomy, occlusion of the right portal vein may induce growth of the left hemi-liver and enable hepatectomy within to ore more months.

Case Report: We present the first reported case of extended left hemihepatectomy in a patient with total situs inversus. In a first step, we did not only divide the right portal vein and bile duct, but also performed parenchymal dissection to induce even more rapid liver growth and perform the final hepatectomy within ten days after the first surgery. The patient underwent two-step hemi-hepatectomy and we observed 63% increase in size of FRLV after ten days. Liver function tests showed functional FLRV and hepatectomy was performed eight days after the first surgery. In the biopsies from the remnant liver we observed hypertrophy and hyperplasia of hepatocytes but not edema. Repeated computed tomography showed further growth of remnant liver up to 200% of initial FRLV after 3 weeks.

Conclusions: We propose the described two-step hepatectomy as an alternative to conventional portal vein occlusion and resection within two or more months, because hepatectomy may be completed within ten days not allowing for tumor growth or spread within this short period.

Key Words: Hemi-hepatecomy – Situs inversus – Portal vein occlusion – Portal vein embolization.

Introduction

LIVER resection is the treatment of choice for patients with primary or secondary liver tumors with tumor load restricted to the liver, or with limited extrahepatic disease [1-4]. Resection is limited, however, by the need to preserve a sufficient amount of functional liver, because excessive resection leads to liver failure and death within a few days after surgery [5].

Although adult hepatocytes are long lived and do not normally undergo cell division, they maintain the ability to proliferate in response to toxic injury, infection, and reduction of overall liver volume as has been shown by the two-thirds partialhepatectomy model in rodents, pioneered by Higgins and Anderson [6] in 1931. Analysis of cellular DNA synthesis shows replication of hepatocytes as early as 12 hours after hepatectomy. This is followed by replication of non-parenchymal cells such as endothelial cells, Kupffer cells, and biliary duct cells initiated within the next 2-3 days [7]. After complete replication, growth (consisting of an increase in cell size), occurs over several additional days [5].

The effects of portal vein occlusion were first applied to the clinical setting by Makuuchi and colleagues to extend the limits of liver resection [8], e.g., in patients with large tumor mass or multiple tumors in segments IV or V through VIII, which were not deemed to survive (extended) right hepatectomy because of a small remnant liver, the right portal vein was occluded and after hypertrophy of the left hemi-liver resection was performed. Selective occlusion of the portal vein may be accomplished by portal-vein embolization or surgical ligation [9].

It has been suggested that liver growth after resection is different from liver growth after portal occlusion [10]. To compare liver growth after resection with portal vein occlusion, we collected data from our own cohort of living donors for liver transplantation and extracted data from the literature [9,11-13]. The analysis clearly showed that after resection, liver growth is increased more than that after vein ligation or occlusion (Fig. 1).

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Therefore, we proposed the following two-step approach in preparation for extended right hemihepatectomy or extended left hemihepatectomy in situs inversus cases, in instances when the potential liver remnant would otherwise be too small. In the first operation, the right portal vein was ligated (or Left portal vein in situs inversus cases) and the liver is split along the future resection margin after complete mobilisation (Fig. 2). When the future remnant liver has undergone sufficient hypertrophy and there are no signs of newly intra- or extrahepatic spread of the tumor, extended right hepatectomy, (extended left hemihepatectomy in our case), is completed in a second operation.

Case Report:

This is the first reported case of extended left hemihepatectomy in a patient with total situs inversus.

During September 2010, the situs inversus patient was evaluated for the above-mentioned procedure (Fig. 1). The indication for liver surgery was huge liver mass in the left liver lobe. Twostep hepatectomy was necessary because of small volume of the right lateral sector and/or expected dysfunction of remaining liver tissue due to steatosis or preceding chemotherapy. The patient was informed about surgical options and gave informed consent to undergo two-step hepatectomy.

Surgical technique:

For the two-step hepatectomy, the surgical technique was as follows:

- 1- Following general anesthesia, a modified left subcostal incision was made.
- 2- A thorough examination was then carried out for the peritoneal cavity to exclude any extrahepatic disease precluding liver resection.
- 3- Assessment of the liver was conducted by visual inspection, bimanual palpation and intraoperative ultrasonography.
- 4- The liver was then mobilized by division of the round, falciform and triangular ligaments (Fig. 3).
- 5- Dissection along the posterior left liver and vena cava was conducted sharply, isolating the left and middle hepatic vein and dividing larger pedicles that enter the vena cava directly from the left liver lobe.
- 6- In the porta hepatis, the right and left hepatic arteries were identified and tributaries to segment IV were ligated.

- 7- The left portal vein and left hepatic duct were then dissected and closed by running suture and/or divided using a vascular stapler (Fig. 4).
- 8- Parenchymal dissection was performed with dissecting scissors used for blunt preparation [14] preserving the venous drainage and hepatic arteries to both hemi-livers.

Seven days after the first operation, a computed tomography (CT) was acquired to assess the volume of the left lateral sector (Fig. 2). In the case, the future remnant liver was deemed to have undergone sufficient hypertrophy and the resection was completed in a second operation eight days after the first operation.

Results

Before surgery, the patient underwent CT (Fig. 2), and volumetric analysis and estimated volume of the future remnant liver (FRLV) was 425mL, which corresponds to a ratio of FRLV to body weight of 0.44%. In the patient, it was concluded that FRLV was not sufficient for standard extended Left hemihepatectomy.

The first surgery was uneventful in the patient, and duration of surgery was 189 minutes. No blood transfusion was necessary in the operation. Postoperative course after the first step was similar to extended hepatectomy. We observed elevation of serum transaminases, reduced bilirubin clearance and prolonged prothrombin time, as expected after major liver surgery.

Within the next four to five days, these values returned to the normal range. Timing of CT was at the discretion of the attending surgeon and CT was conducted after 7 days. Volumetric analysis showed a significant increase of FRLV, by 62% of preoperative FRLV (*p*-value=0.01). In this case, FRLV was deemed sufficient and hepatectomy was performed on the 8 th postoperative day. Extended hepatectomy was considerably shorter because no parenchymal transection was necessary; duration of surgery was 57 minutes. A second surgery was uneventful and no blood transfusion was necessary.

In the patient, we observed mild re-elevation of transaminases after hepatectomy, whereas bilirubin clearance and coagulation function were compromised again. In this patient, prothrombin time returned to normal within 7 days and bilirubin returned to normal within 6 days. The patient had pulmonary embolism and received therapeutic doses of heparin which was later switched to oral phenprocoumon. The patient was discharged 31 days after the first surgery.



Fig. (1): CT Scan before the first Operation.



Fig. (3): The Liver in the left side of the abdomen after left subcostal incision and release of falciform ligament and right and left triangular ligaments with enlarge-ment of left liver lobe.



Fig. (5-A): The left liver lobe after resection.



Fig. (2): CT Scan after the first Operation to asses FLRV.



Fig. (4): The left portal vein and left hepatic duct were dissected and prepared for ligation.



Fig. (5-B): The left liver lobe after resection.



Fig. (6): The resected left liver lobe showing the line of resection.

Histology:

The patient hepatectomy specimen underwent standard pathological work-up. Additionally, biopsies from the FRLV were obtained for histology. Immunohistochemical staining for Ki67 (mouse monoclonal, clone K-2, heat pre-treatment, dilution 1:400, Zytomed System, Berlin, Germany) was performed on formalin-fixed and paraffinembedded tissue sections according to standard protocols. The tissue sections were then evaluated microscopically.

Histologic work-up showed extended hemihepatectomy prepared with 19cm length, minimal differentiated hepatocellular carcinoma, malignant grade 3. There is no liver capsule enlargement, the vessels are free, bile ducts were free and there is no liver cirrhosis. Tumor Classification was T1, G3, N0, M0, L0, V0, R0. Biopsies from the FRLV showed regenerative tissue with broadening of the liver cell trabecules, only mild edema and elevated proliferative activity.

Discussion

Here we present a new approach for extended right hepatectomy (or extended Left hepatctomy in this situs inversus patient), in patients with small FRLV or suspected insufficiency of FRLV, the twostep hepatectomy. Liver resection has been completed in two stages, quite similar to the concept of portal vein occlusion in preparation for major liver surgery. However, in the first stage, we are not only occluding the right portal vein (left portal vein in situs inversus cases) but also perform the dissection of liver parenchyma, ligation of portal and arterial tributaries to segment IV, and ligation of the right hepatic duct (or left hepatic duct in situs inversus patient). Hepatectomy has been completed in a second stage, after less than ten days because of the rapid growth of the FRLV (Figs. 3-6). The basis for the concept of this technique is the clinical observation that, after hepatectomy and portal vein occlusion, regeneration is more rapid than after portal vein occlusion alone [13].

In this case report, we observed increase of remnant liver volume of 66% within seven days after the first surgery. This rapid increase of remnant liver volume is in accordance with our own and Nagasue's [13] observations, which show rapid increase of FRLV within the first one week after hepatectomy, and less increase thereafter.

Conclusion:

• The two-step approach seems to be effective and safe, but it needs further evaluation with a prospective study.

• We recommend careful preoperative evaluation of all patients, so as a known patient of situs inversus would be safely operated upon.

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