CASE REPORT
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Associating liver partition and portal vein ligation for staged hepatectomy (ALPPS) for pediatric mesenchymal hamartoma: A case report

Alvin Caballes[1], Kristian Alexis De Lara[2]

[1]Department of Surgery, University of the Philippines College of Medicine, Manila, Philippines
[2]Department of Surgery, Philippine Children's Medical Center, Quezon City, Philippines

ABSTRACT

The case involves a one-year-old male with a mesenchymal hamartoma involving the right hepatic lobe. The tumor-free segments comprised 17% of the liver volume, which placed the patient at risk for post-resection liver failure. A staged approach, the associating liver partition with portal vein ligation for staged hepatectomy, was employed. This allowed the interval growth of the liver remnant and thereafter enabled right lobectomy with adequate liver function.

Keywords: Hamartoma, liver neoplasm, hepatectomy, liver regeneration

INTRODUCTION

Resection is the primary curative option for patients with malignant liver tumors. This also applies to many benign lesions that may be complicated by massive growth or potential for malignancy, particularly among pediatric patients. However, resection is not a feasible alternative for cases wherein the remnant liver will have insufficient functional capacity. The latter has been spatially quantified as the proportion of the volumes of the remaining segments compared to the total liver, and is termed the future liver remnant (FLR). The associating liver partition with portal vein ligation for staged hepatectomy (ALPPS) technique was first performed in 2007. It has been shown to cause a substantial increase in the FLR within a relatively short period, enabling resection while avoiding consequent liver failure (1). We report the case of a one-year-old male with a large mesenchymal hamartoma (MHL) and a small initial FLR who underwent ALPPS. To our knowledge, this is only the second patient reported for this procedure and indication worldwide, and the first reported ALPPS case in our country.

CASE REPORT

A one-year-old male had progressive abdominal distention over two months. He was otherwise asymptomatic. A computed tomography (CT) scan done at another institution showed an 11.8x12.4x13.5 cm lobulated cystic mass with septations at the right hepatic lobe. An impression of MHL was given. He was referred to our center for definitive surgical management. On admission, the patient weighed 10.7 kg (Z-score +1), with a height of 76 cm. The upper abdomen was markedly distended. The following laboratory results were obtained: Alanine transaminase (ALT): 23 U/L, aspartate transaminase (AST): 47 U/L, Gamma-glutamyl transferase: 85 U/L, albumin: 45.7 g/L, and alpha fetoprotein: 49.44 mg/L. Upon review of the CT scan, the multi-lobulated cystic tumor was determined to occupy segments IV, V, VII, and VIII (Figure 1). The FLR was estimated to be 17%. In consideration of the latter, a staged resection was recommended. The planned surgery was delayed for several days due to Coronavirus disease-2019 contingencies.

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Corresponding Author
Alvin Caballes

E-mail: abcaballes@up.edu.ph
ORCID ID: orcid.org/0000-0002-9902-754X

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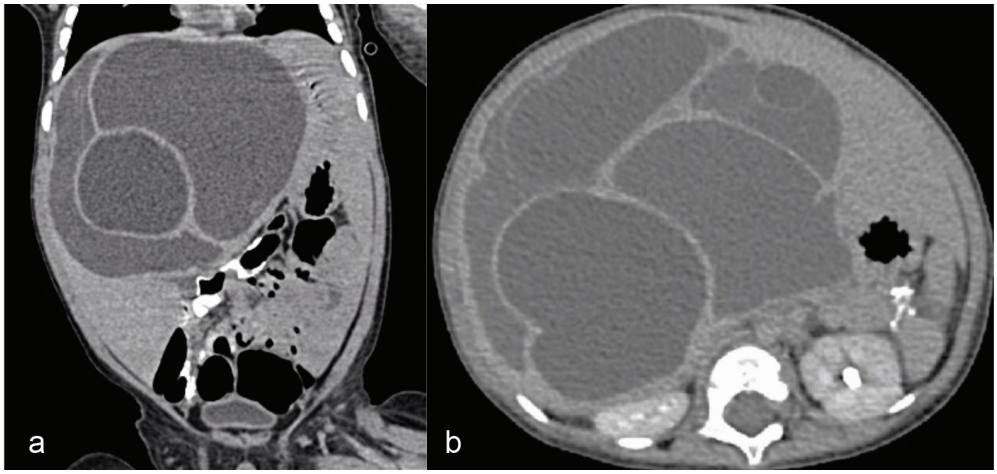

Figure 1. Selected CT scan images of the abdomen demonstrating a massive multi-lobulated cystic tumor at the right liver and the lesion-free segments II and III, a. coronal section, b. axial section.

CT: Computed tomography

The first stage was performed through a transverse upper laparotomy. The dilated right portal vein was isolated, transected, and the proximal stump was closed with a running suture (Figure 2). The left hepatic artery, hepatic duct, and portal vein, which were stretched out by the expanded right liver lobe, were dissected and traced to the left liver lobe to safeguard these during liver partition. Retrohepatic veins to the right lobe were divided and ligated. Liver separation was performed with blunt dissection, following the falciform ligament. Pringle's maneuver was not employed. Only a partial split was done, as the hepatic veins and adjacent parenchyma were spared (Figure 3). A plastic film was laid over the transection area, on which a silicone drain was placed. The operative time was 255 minutes, and the blood loss was 170 mL.

The patient's postoperative course was difficult, with hemodynamic instability and high ventilatory support requirements. The initial international normalised ratio was 1.56. Liver enzyme (ALT: 519 U/L, AST: 963 U/L) and bilirubin (DB: 4.29 umol/L, IB: 19.45 umol/L) levels were elevated. Hospital-acquired pneumonia also set in. Thus, while a CT scan showed that the FLR had increased by 47% by the 14th day after surgery, the second stage could only be done after another week.

On re-laparotomy, considerable adhesions were encountered over the right liver lobe, as well as on the previously freed-up main vascular and biliary structures going to the left liver. The left hepatic duct was injured during the dissection and duly repaired. The cystic duct and artery were transected and the gallbladder was left *in situ*. The right hepatic artery and duct were divided. Right hepatectomy was completed following the separation of the remaining parenchymal bridge and transection of the right and middle hepatic veins. The operation lasted 280 minutes, with the total blood loss being 750 mL. There was a rapid recovery and the patient required only a brief intensive care unit stay. A single episode of hematochezia occurred on the 10th postoperative day. Bile was noted at the peritoneal drain at the thirteenth postoperative day. These developments were managed conservatively. The patient was sent home 15 days after surgery.

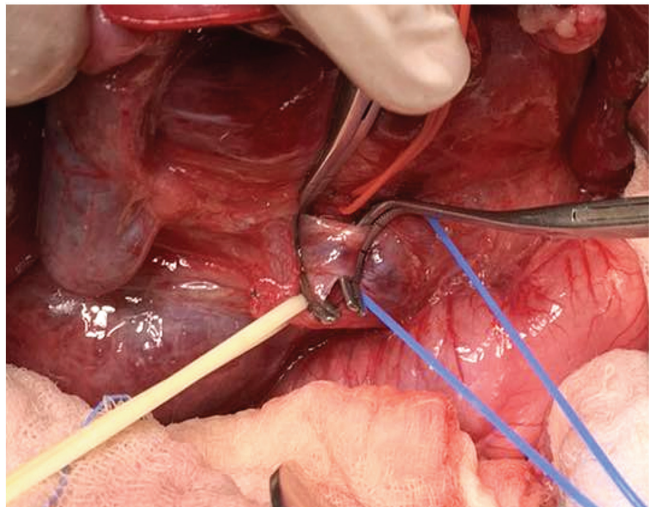

Figure 2. The right portal vein has been isolated and vascular clamps are applied proximal and distal to the point of transection. Also shown are the common bile duct (yellow tag), right hepatic artery (red tag), and portal vein (blue tag).

The lesion was confirmed to be MHL on histopathology, with margins clear of tumor. There are no indications of hepatic decompensation two years after the procedure. However, a recent CT scan performed at another institution had findings suggestive of portal vein stenosis. Additional studies to confirm this are forthcoming.

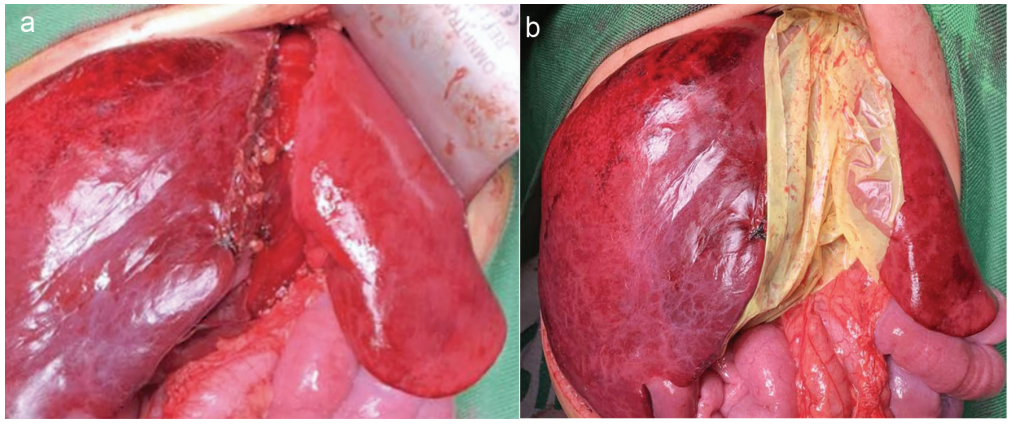

Figure 3. a. Partial hepatic separation accomplished. b. Plastic film and a drain tube are placed over the separation area.

DISCUSSION

Liver tumors comprise 1% of all pediatric solid tumors and are more commonly malignant. MHL is the second most frequent type of benign liver tumor in children, surpassed only by hemangioma (2). As with the present case, the usual presentation is a large abdominal mass in an otherwise asymptomatic preschool child. There have been a few reports of fetal and newborn MHL cases, with most having severe presentations, such as respiratory distress or bleeding. Adult MHL is rare. Imaging often demonstrates non-contrast-enhancing septated cysts in the affected liver segments. Serum AFP levels may be elevated, but not to the same extent as those of malignant tumors. Given the age group and presentation of patients, hepatoblastoma is a likely differential diagnosis. However, the AFP levels are considerably higher and cystic lesions are uncommon with hepatoblastoma.

Spontaneous regression of MHL has been reported, especially for highly vascular lesions. Only some reduction in size, rather than actual tumor resolution, has been documented within the short monitoring period of these cases (3). Progressive enlargement is the norm for MHL. Likewise, it is associated with the occurrence of undifferentiated embryonal sarcoma of the liver, a malignant tumor of mesenchymal origin. As such, resection of the involved liver segments is the recommended treatment for MHL (3,4). This, however, can be challenging for massive tumors wherein the resulting FLR may be inadequate.

The minimum FLR often recommended for a hepatectomy is 20%. Methodological and clinical concerns have nonetheless been raised regarding the estimation of liver volumes (5). While there may not be a definitive cut-off, optimizing the residual liver size is an important consideration for hepatic resections. There are several surgical options for liver tumors with insufficient FLR. Transplantation or when suitable two-stage or central hepatectomies may be utilized (5,6). Should these not be practicable, a procedure that increases the FLR prior to resection can be a reasonable option.

Following partial hepatic resections, hepatocyte regeneration is known to occur in the remaining segments. This is triggered by an increased portal venous inflow, leading to a greater influx of intestine-derived growth factors and other changes that promote regeneration. Portal vein embolization or occlusion has thus been employed to deliberately stimulate hypertrophy of the anticipated liver remnant prior to the required resection. The desired level of liver regeneration may take several weeks to be achieved, and this can be detrimental for patients with malignant lesions. As such, other approaches that more substantially redirect hepatic circulation, including transarterial embolization, liver venous deprivation, and radiation lobectomy, have been utilized to accelerate liver regeneration (6,7). With ALPPS, which incorporates ipsilateral portal venous occlusion with collateral vascular interruption through liver separation, portal venous flow is exclusively directed to the contralateral normal segments. Hypertrophy of the latter is thereby further hastened. Among 45 adult patients who had weekly CT scans after the first stage of ALPPS, 85% reached the required FLR by the seventh postoperative day (8). Contingent on the documented adequacy of FLR hypertrophy, resection could be undertaken within a week or two after the first stage of the procedure.

While ALPPS has been utilized in many adult cases, it has not yet been extensively applied in pediatric patients (9). The first reported pediatric ALPPS case was a 6-year-old child with hepatoblastoma (10). A systematic review covering pediatric patients, compared 12 who had ALPPS and nine who underwent only portal vein embolization or ligation before resection. The mean FLR was lower for the ALPPS group (11). Three pediatric patients who had centrally-located hepatoblastoma and who would have been transplant candidates based on PRETEXT stage were recently reported to have successfully undergone ALPPS (12). A partial ALPPS, with preservation of the liver parenchyma

adjacent to the hepatic veins during the first stage, was previously performed in a 54-day-old infant with hepatoblastoma (13). A 9-month-old with a massive MHL and an FLR of 22.7% also underwent ALPPS (14). As had happened with our patient, the course immediately following the first surgery was turbulent. The latter may be attributed to an acute hepatic insufficiency, which does not deteriorate into irreversible liver failure.

There were procedural aspects in the current case that could have been improved. There was undue delay in the performance of the second stage, which undermined the temporal advantage of ALPPS. The handling of the left hepatic vascular and biliary structures during the first stage should have been minimized to lessen the consequent inflammatory reaction and scarring that hampered the subsequent surgery.

CONCLUSION

The case adds to the growing number of pediatric patients with liver tumors who have been treated with ALPPS. While the procedure is utilized more for adults with malignant tumors, its applicability for benign lesions in pediatric patients, is further highlighted in the current report.

Ethics

Informed Consent: Informed consent was obtained.

Footnotes

Author Contributions

Concept - A.C., K.A.D.L.; Design - A.C., K.A.D.L.; Supervision - A.C., K.A.D.L.; Data Collection or Processing - A.C., K.A.D.L.; Analysis or Interpretation - A.C., K.A.D.L.; Literature Search - A.C., K.A.D.L.; Critical Review - A.C., K.A.D.L.; Writing - A.C., K.A.D.L.

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