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ALPPS for hepatic mesenchymal hamartoma in an infant

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ABSTRACT

Associating liver partition and portal vein ligation for staged hepatectomy(ALPPS) has been recently introduced as a new procedure to improve the surgical management of large hepatic tumors with future liver remnant (FLR) less than 25% in adults. However, there's rare such case report of infants. We present a case of a 9-month-old infant with huge hepatic mesenchymal hamartoma who underwent ALPPS. The patient initially presented with one week history of abdominal distension and vomit after eating. Computed tomography(CT) showed a right hepatic lobe multilocular mass measuring 10 cm*9 cm*10 cm, occupying IV,V,VI,VII,VIII segment. Volumetric assessment of the FLR was 102.31 cm^3, which is 22.74% of the estimated total liver volume. With considerably low FLR, we performed ALPPS procedure for the infant. The first stage of the procedure contained the ligation of the right branch of the portal vein and in-situ liver parenchyma split. The second stage entailing right trisectionectomy after 8 days. The success of this application proved that ALPPS is suitable for little infants with giant benign neoplasm. To our knowledge, this is the first successful case of infant's ALPPS.

1. Introduction

Hepatic mesenchymal hamartoma(HMH) is abnormal hamartomatous growth of mesenchymal tissue, bile ducts and blood vessels of the liver. It is the second common benign neolasm next to hemangioma, and is the second most common benign liver tumors in children, accounting for about 5–8% of pediatric liver tumors [1]. Tumor larger than (10 cm) is defined as very large as in this case. In order to maintain normal liver function, FLR should be no less than 25% [2]. In this case, FLR is 102.31 cm^3, with FLR/SLV(standard liver volume) ratio of 22.74%. As we known, the only way of achieving good prognosis is complete tumor resection or liver transplantation [2].

Insufficient FLR is associated with postoperative liver failure. The conventional approach is usually to perform portal vein embolization or ligation to induce FLR hypertrophy, which normally requires 4–8 weeks for the second stage hepatectomy [3]. However, disease progression could result in drop-out from receiving curative resection. On the other hand, associating liver partition and portal vein ligation for staged hepatectomy (ALPPS) is a novel procedure to induce rapid liver hypertrophy. We present the case of an infant with huge mesenchymal hamartoma of the liver who underwent ALPPS procedure.

2. Case report

A 9-month-old male infant presented with one-week history of abdominal distension and vomiting after eating. He was referred to our institution for disease assessment. His body weight and height was 10 kg and 75 cm. He had poor oral intake and was not gaining. Abdominal ultrasound demonstrated a mass arising from the liver. CT scan was then ordered to further assess of the tumor (Fig. 1), which revealed a 10 cm *10 cm*9 cm mass occupying the segment IV,V,VI,VII,VIII of the liver. Liver function were normal. However, volume measurement of the FLR, the left lateral section(LLS) was only 102.31 cm^3 with FLR/SLV ratio of 22.74%. A laboratory workup was ordered concomitantly which included liver function test exam, lactate dehydrogenase (LDH) and Alpha Fetoprotein (INR: 1.03, Albumin: 33.52 g/L, ALT: 48.91 IU/L, AST: 79.41 IU/L, Total Bili-rubin: 10.49 mmol/L, LDH: 701.60 IU/L, Alpha Fetoprotein: 263.10 mg/L). Preoperative fine-needle biopsy indicated liver cystic lesion. After the initial workup, the primary diagnosis was a huge benign neoplasm. Surgery is the only way to treat the tumor. Stated thus, decision was made to proceed with a ALPPS procedure [4].

After preoperative evaluation, the infant underwent surgery. In the first stage of ALPPS on March 27th, 2018, right portal vein ligation and in-situ liver spilt was performed. The gallbladder was not removed so as to minimize physiological interference [5]. After identification of the portal structures, the right portal vein was transected and marked (Fig. 2). The right hepatic artery and left hepatic vessels were safeguarded. The parenchyma transection was performed along the falciform ligament using ultrasonic scalpel, retaining little liver tissue

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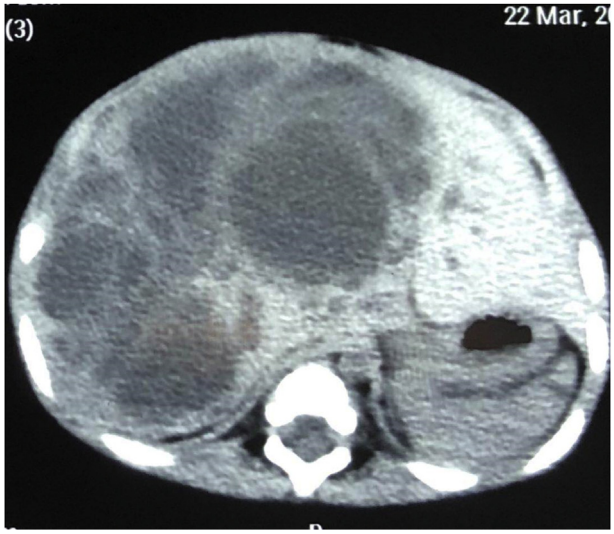

Fig. 1. CT showed the position of liver tumor.

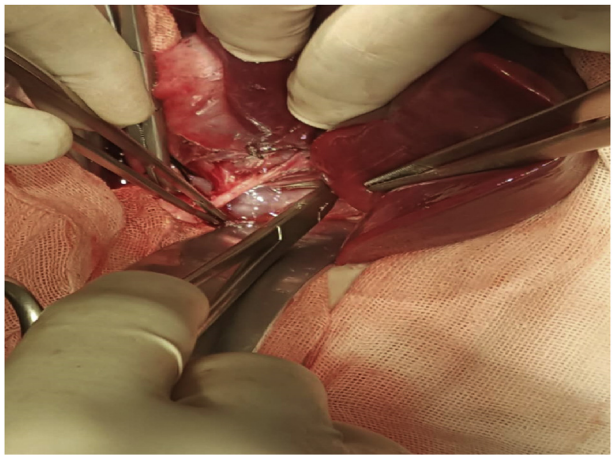

Fig. 2. The dissection of right hepatic vein.

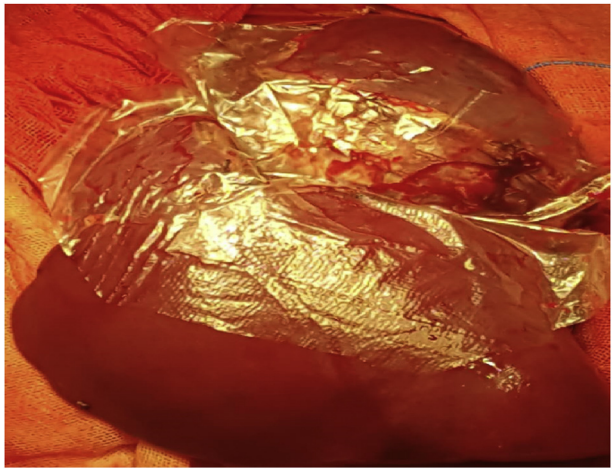

Fig. 3. The plastic membrane separates the liver.

between the first and second hepatic hilum. The transection surface was protected with a plastic film(Fig. 3) instead of a bag and fixed with 2.0 prolene suture to prevent adhesions [6]. The operating duration was 180min. The blood loss was 30 mL, and no blood transfusion necessary during the procedure. The infant was monitored in the pediatric intensive care unit (PICU). The recovery was not good as expected after the first operation. Ventilator is still needed. The average peritoneal exudation in the first 6 days is 1500 ml/d, which reached a peak 1891 ml in day 5. CT volumetry on day 6 revealed that FLR was 179.79 ml, the SLV was 449.93 ml with FLR/SLV ratio of 39.96%. After 8 days, the second stage of ALPPS started with the same incision and mobilization of the tumor mass from the vena cava. The plastic film was removed. First, cholecystectomy was performed without ligating the cystic duct. The right hepatic artery, the right bile duct, the right hepatic vein and the middle hepatic vein were ligated and divided. Liver segment IV-VIII with tumor from the vena cava eventually after the hepatic short vein were ligated and divided. 20% fat emulsion was injected into the cystic duct to exclude bile leakage from the transection surface [7]. Then the cystic duct was ligated and divided. Two silicone drains were placed and the abdomen closed. Patient was sent to the intensive care unit. The operating time was 150min, intraoperative blood loss was 50 ml, and one unit of red cell was transfused during the procedure. CT volumetry on day 9 after second stage revealed FLR was 331.50 ml with FLR/SLV ratio of 73.68%. The postoperative course was uneventful and the infant was discharged at 24 days after the first-stage ALPPS procedure. After 80 days of following up, the infant is thriving with no signs of recurrence. Histopathology of the mass demonstrated a mesenchymal hamartoma of the liver.

3. Discussion

Mesenchymal hamartoma is the second most common benign hepatic neoplasms in children. Eighty percent are discovered during the first two years of age and the remainder by age of 5. There is a slight male: female predominance of 1.5:1 [8]. Mesenchymal hamartoma usually present as an asymptomatic mass lesion. However, as it grows it has the potential to compress adjacent organs, resulting in various symptoms and complications, which include a wide spectrum ranging from respiratory distress to death. Approximately 75% of liver hamartoma occur in the right lobe of the liver. Liver function tests and Alpha Fetoprotein (AFP) usually remain within normal range or be mildly elevated. Imaging studies will demonstrate a large, well circumscribed mass which may contain cyst of various sizes. Treatment consists of a complete surgical resection with clear margins. Pathological biopsy will demonstrate multiple cysts containing yellow, serous fluid. Microscopically, hamartoma shows mesenchymal and epithelial components. The myxomatous stroma contains epithelial and non-epithelial-lined cysts [8].

It is believed that hepatectomy is the best way to obtain long-term prognosis for both primary and secondary malignant liver tumors. The main reason for the failure of surgery is the insufficient FLR, leading to postoperative hepatic failure [9]. The ALPPS procedure has gained popularity for the treatment of patients requiring liver resection with a very low predicted FLR. Schnitzbauer AA etc first reported ALPPS in 2012 [2], and Albert Chan from Queen Mary Hospital reported a pediatric case of 6-year-old [3]. Qazi A Q etc reported a Early multifocal recurrence of hepatoblastoma in the residual liver after R0 liver resection with ALPPS procedure [10,14]. There's no yet any successful case report of infants.

The FLR of the little 9-month-old infant with mesenchymal hamartoma was 22.74%, less than needed. Any further delay of FLR hypertrophy would put him at risk of tumor progression. Considering the rapid growth of the neoplasm(which is the pathological feature of the MH) and progressive poor appetite and vomiting, we decided to apply ALPPS in order to gain a rapid FLR hypertrophy. For a massive benign liver neoplasm, there's no violating the non-touch principle and metastasis problems [11]. We consider ALPPS performed for the gigantic benign liver tumor more beneficial than those to malignant liver tumors.

We faced many difficulties during the treatment. First, it is a challenge because of the age of the boy. As we known that the anatomy,