Case Report

Combined liver-kidney transplant in a 21-month-old child with type 1 primary hyperoxaluria—The perioperative challenges

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ABSTRACT

Primary hyperoxaluria type 1(PH 1) is the most common indication for a paediatric combined liver-kidney transplant. It is a technically challenging procedure. We describe the challenges in managing a 21-month-old female child weighing 7 kg for a combined liver-kidney transplant from two related living donors.

Key words: Anesthesia for combined liver and kidney transplant, combined liver-kidney transplantation, primary hyperoxaluria type 1 (PH 1)

INTRODUCTION

Primary hyperoxaluria type 1(PH 1) is a rare, congenital condition due to deficiency of the hepatic enzyme alanine-glyoxylate aminotransferase (AGT), which necessitates a combined liver-kidney transplant (CLKT) in childhood.[1-3] CLKT is a technically demanding procedure characterized by massive fluid shifts, blood loss, and post-transplant renal failure due to oxalate load. We report the perioperative challenges faced during CLKT in a 21-month-old female child with PH 1.

CASE REPORT

A 21-month-old female child (weight 7 kg) with PH 1 was admitted for CLKT and right nephrectomy. At the age of 1 year, she was diagnosed as a case of primary autosomal recessive polycystic kidney disease with renal failure and hypertension and was started on multiple anti-hypertensives and continuous abdominal peritoneal dialysis (CAPD). Recurrent abdominal infection, catheter leaks, and umbilical hernia necessitated multiple surgeries Soon haemodialysis (HD) was initiated through a subclavian catheter after stopping CAPD. However, worsening renal status and failure to thrive prompted further investigations. The presence of oxalate crystals on the retinal scan, bilateral renal calcification on X-ray and computed tomography (CT) of abdomen, and serum oxalate of 157 ug/l drew attention towards a diagnosis of PH. [Figure 1]. Alanine--glyoxylate and serine--pyruvate aminotransferase (AGXT)

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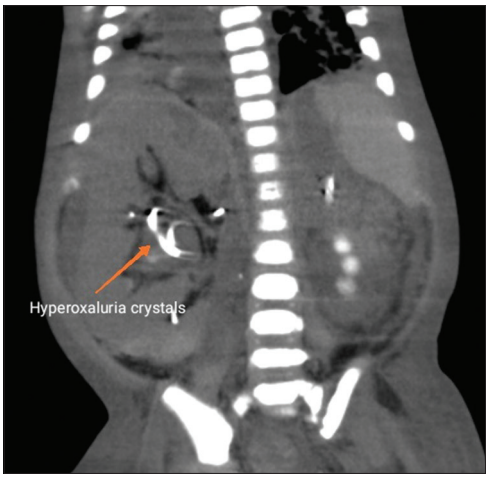

Figure 1: CT Scan shows oxalate crystals in the kidney

gene mutational analysis study confirmed PH 1. Echocardiogram showed left ventricular hypertrophy with mild diastolic dysfunction.

The child's father (34 years) and paternal grandmother (54 years) were worked up as the liver and renal donor, respectively. After securing peripheral line and left radial artery cannula under sedation, anaesthesia was induced using a combination of fentanyl (12 μg), sevoflurane (2%), ketamine (4 mg), and atracurium (3.5) mg. Difficulty was encountered on attempting to intubate with a 4 mm uncuffed endotracheal tube (ET), which persisted with smaller size tubes. A size one LMA was placed, making ventilation possible. A paediatric fiberoptic bronchoscope (2.8 mm, Karl Stroz, Germany) showed severe subglottic stenosis. An emergency tracheostomy was done using a 4 mm internal diameter uncuffed tracheostomy tube. Anaesthesia was continued using a combination of air, oxygen, sevoflurane, fentanyl (3 mcg/kg/hour), and atracurium (2 mg/hour). Two central lines (5 Fr, 8 cm), one in the right internal jugular vein (RIJV) and the other in the right femoral vein (RFV) were established. An additional arterial line was placed in the left femoral artery (22G). Noradrenaline infusion was titrated to maintain mean arterial pressure (MAP) above 65 mmHg. Fluid warmers and warming blankets were used to maintain temperature. Piperacillin-tazobactam, teicoplanin, and fluconazole were used as antimicrobial prophylaxis. Thromboelastogram (TEG) was used to monitor coagulation profile and guide blood product transfusion.

After removal of the diseased liver and the right kidney, the left lateral segment of the donor liver (segments 2 and 3, 240 grams, graft recipient weight ratio 3.8) was implanted. The central venous pressure (CVP) was maintained at 4-5 mmHg. The total warm ischaemia time, cold ischaemia time (CIT), and anhepatic time were 43, 58, and 75 minutes, respectively. Reperfusion was uneventful. After hepatic vascular anastomosis and restoration of hepatic circulation, Central venous pressure (CVP) was raised to 9-10 mmHg avoiding fluid overload. Mannitol 0.5 g/kg and frusemide 0.5 mg/kg were administered. The kidney was placed in the retroperitoneum on the right side. The renal artery was anastomosed using a side clamp on the aorta and similarly the renal vein to the inferior vena cava (IVC). The warm ischaemia time and CIT for kidney were 1 hour 22 minutes and 1 hour and 48 minutes. Urine output was good following renal reperfusion. The bile duct anastomosis and uretero-vesical anastomosis were then performed. The child received 280 ml of packed cells, 150 ml of fresh frozen plasma, and 1 unit of platelets. Blood loss was 300 ml and urine output was 350 ml. Plasmalyte® (1500 ml) was also given during the procedure, which lasted 17 hours. Methyl prednisolone (70 mg) was administered before the liver implant and basiliximab (10 mg) was given before the renal implantation.

The child was shifted the paediatric intensive care unit (PICU) after surgery for elective ventilation. Haemodynamics were maintained with noradrenaline (0.2 mcg/kg/min). She was initiated on slow low-efficiency dialysis (SLED) in the immediate postoperative period. On the fifth postoperative day (POD), she was tapered of inotropes and was started on haemodialysis. The peak postoperative level of bilirubin was 3.5 mg/dl, aspartate transaminase (AST, 393 IU), international normalized ratio (INR, 4.15), and creatinine (1.59 mg%). Serial Doppler showed satisfactory flows. On postoperative day (POD) 10, she had an intestinal perforation for which she underwent an exploratory laparotomy with ileostomy. She was ventilated for 10 more days and then placed on noninvasive ventilation from which she was weaned off by POD 19.

Serum oxalate levels became normal at discharge. Bronchoscopy done 4 weeks later showed normal airway so she was de-cannulated. She was discharged on the POD 52 with a functioning ileostomy and is doing well on follow-up.

DISCUSSION

The first simultaneous kidney-liver transplant was done by R Margeiter in 1983.[4] Although CLKT is

routine in adult transplant programs, only about 88 procedures have been done in the children aged 0-5 years till 2016.[5] The most common indication for paediatric CLKT is PH 1.[2]

PH 1 is an autosomal recessive metabolic disorder caused by deficiency or mislocalization of the liver-specific AGT.[3,5] This results in the conversion of glyoxylate to oxalate, which forms insoluble calcium salts. This results in recurrent nephrocalcinosis, nephrolithiasis, or end-stage renal disease (ESRD).[1,3] In severe infantile form of PH 1, 80% of these children develop ESRD by the age of 3 years.[2,5]

Haemodialysis, peritoneal dialysis, or a combination of the two can be considered.[1] However, the high rate of oxalate production exceeds removal by dialysis. Hence, the only curative treatment is CLKT.[1,3]

Multiorgan transplantations present a great challenge, especially in young children.[6] The normal-sized adult kidney and 240 gm liver had to be accommodated within the abdominal cavity of a 7 kg child.[2] The space within the abdomen was further compromised by adhesions from previous surgery.[6] Thus, a large protruding abdomen with upward displaced diaphragm may compromise the postoperative respiratory function. Hence, right nephrectomy was done and the neo kidney was placed in the space vacated.

CLKT is almost exclusively performed with organs from deceased donors.[2] We used live donors due to a shortage of deceased donors. Although technically possible, the procurement of part of the liver and a kidney from the same living donor is considered a risky procedure for the donor.[2] Therefore, we decided to go ahead with a live donation from two donors.

The liver has an immunoprotective effect on the neo-kidney graft when both organs come from the same donor.[2,3,6,7] In this case, it is not clear whether any such protection exists when two related donors are involved.

Multisystem involvement, complex surgical procedure, and difficult fluid replacement strategy added to the anaesthesiologist's woes.[3,7] Restricted fluid replacement (CVP 5-7 mmHg) is recommended in the pre-anhepatic phase.[2,7,8] After implantation of the kidney, CVP was raised to 9-10 mmHg. Central venous oxygen saturation (ScvO2) was targeted at around 70%. The lactate levels and the base excess could not be relied upon entirely as these values tend to increase during the anhepatic phase. The targeted haemoglobin level during the intraoperative period was about 9 gm%. Blood loss was carefully monitored and arterial blood gas (ABG) was repeated every three hours Modern techniques of cardiac output assessment including arterial waveform-based stroke volume analysis are not validated in children.[9]

The difficulty in intubation during the surgery was unexpected. The child had undergone repeated intubations in the past. This could have contributed to subglottic oedema and narrowing.[10] The normal findings during bronchoscopy before discharge were probably due to clearance of airway edema.

An important technical challenge in CLKT is the minimization of CIT of the organs, as it is known to impact both early and long-term functions of transplanted organs.[2,6] Since we were transplanting from two donors, the transplants were timed in such a way that CIT was less. The renal donor was induced only when the liver transection was almost complete. Vascular anastomoses and reperfusion of the liver were performed first followed by that of the kidney. The bile duct and vesicoureteral anastomosis were performed after the establishment of perfusion to both the transplanted organs resulting in minimal CIT.

Late recurrence of renal oxalosis due to post-transplant mobilization of tissue-bound calcium oxalate may complicate successful combined CLKT.[2,3,5,6] SLED rather than HD was started in the immediate postoperative period as hypotension during the dialysis could compromise perfusion of the transplanted liver. HD later replaced SLED till the oxalate levels returned to normal. Hyper hydration of the patient has to be maintained until the urinary oxalate/creatinine ratio is stabilized.[1]

CONCLUSION

The complexity of the surgical procedure coupled with the fragile and tenuous condition of a small child can make the anaesthetic management, a nightmare. Managing both expected as well as unexpected complications during the course of surgery made the case very challenging and unique for the anaesthesiology team.

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Conflicts of interest

There are no conflicts of interest.

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