CASE REPORT

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Renal Transplant in a Case of EHPVO with Severe Pancytopenia

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Abstract

Renal transplantation in patients with extrahepatic portal vein obstruction (EHPVO) is rare and surgically challenging due to portal hypertension, hypersplenism, and altered vascular anatomy. We report a 32-year-old male with chronic kidney disease stage V on maintenance haemodialysis, associated with EHPVO, portal cavernoma, and severe hypersplenism, who successfully underwent live donor renal transplantation along with simultaneous mesocaval and lienorenal shunt surgery. Preoperatively, the patient had severe anaemia (Hb 5.5 g/dL) and thrombocytopenia (20,000/mm^3). Intraoperative findings included extensive collateralisation and splenomegaly. The postoperative course was uneventful with immediate graft function, improvement in renal parameters (serum creatinine 0.8 mg/dL at discharge), and rising platelet counts. No rejection or infectious complications were observed. This case highlights that renal transplantation is feasible in patients with complex portal hypertensive disorders when combined with appropriate portal decompressive procedures and multidisciplinary planning.

Key words: Extrahepatic portal vein obstruction; renal transplantation; portal hypertension; hypersplenism; mesocaval shunt; Lienorenal shunt.

Introduction

Extrahepatic portal vein obstruction (EHPVO) is a major cause of non-cirrhotic portal hypertension and is commonly associated with portal cavernoma and hypersplenism[1,2]. Renal transplantation in such patients is rarely performed due to technical challenges including distorted vascular anatomy, high bleeding risk, and cytopenias[3,4]. Severe thrombocytopenia further complicates the perioperative management. Literature on simultaneous renal transplantation with portal decompressive procedures remains limited. We report a rare case highlighting the successful management with a combined surgical approach.

Case Report

A 32-year-old male with chronic kidney disease stage V on maintenance haemodialysis via right internal jugular permacath was planned for renal transplantation. The patient was evaluated and found to have severe anaemia (haemoglobin 5.5 g/dL) and marked thrombocytopenia (platelet count 20,000/cumm). His liver function tests were normal and fibroscan showed a kPA value of 5.6 indicating a normal liver elastography.

He was further evaluated and ultrasonography revealed chronic EHPVO with portal cavernoma, splenomegaly, and features of portal hypertension. Triple phase CT abdomen demonstrated features of portal hypertension with massive splenomegaly. The persistent thrombocytopenia was attributable to hypersplenism after ruling out other causes.

His immunological workup including complement-dependent cytotoxicity crossmatch, flow crossmatch, and donor-specific antibodies was negative. Due medical and surgical clearance was taken from all the departments. A multidisciplinary meeting was held and given the presence of significant portal hypertension and extensive collateralisation, a combined surgical approach was planned.

The patient received induction immunosuppression with basiliximab with initiation of low dose tacrolimus and prednisolone for immunosuppression. Injection filgrastim was given twice per week for one month prior to surgery. Adequate platelet along with packed red blood cell were also transfused prior to surgery and intraoperatively to manage pancytopenia.

Intraoperatively, extensive mesenteric and splenic collaterals, portal cavernoma, and massive splenomegaly were noted. The patient underwent exploratory laparotomy followed by mesocaval shunt and lienorenal shunt (side-to-side) creation for portal decompression. Renal transplantation was then performed with the donor kidney anastomosed to the recipient's internal iliac artery and external iliac vein. Ureteroneocystostomy was done with placement of a double-J stent.

The immediate post-operative period was marked by good

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urine output (up to 7 litres/day) and satisfactory graft perfusion. Serum creatinine improved from 3.8 mg/dL on post-operative day 0 to 1.0 mg/dL by day 5 and 0.8 mg/dL at discharge. On post-operative day 1, four units platelets were transfused and platelet counts showed gradual improvement to 40,000/mm^3 on post-operative day 2 to 65000/mm^3 on day 9. Mycophenolate mofetil was withheld on the day of transplantation due to concerns of exacerbating cytopenias and was introduced at a low dose on post-operative day 4, once platelet and leukocyte counts demonstrated an improving trend.

There were no episodes of acute rejection or infection. Doppler ultrasonography demonstrated a well-perfused graft (resistive index 0.61) and patent mesocaval and lienorenal shunts.

He was discharged on post-operative day 9 in stable condition with normal graft function and improving haematological parameters. At 1-month post-transplant patient has a good urine output of 1.8 - 2 litres/day with a serum creatinine of 0.9 mg/dL on triple drug immunosuppression with prednisolone, tacrolimus and mycophenolate mofetil and maintains his haematological parameters as Hb 9.8 g/dL, TLC 4400/cumm and platelet count of 1,25,000/cumm.

Presently, at 3 months follow-up, patient has urine output of 1.5 - 2 litres/day with a serum creatinine of 0.8 mg/dL on triple drug immunosuppression with prednisolone, tacrolimus and mycophenolate mofetil and maintains his haematological parameters as Hb 10.1 g/dL, TLC 5,600/cumm and platelet count of 1,46,000/cumm.

Discussion

EHPVO is characterised by chronic thrombosis of the portal vein leading to cavernoma formation and portal hypertension. Hypersplenism frequently results in cytopenias, particularly thrombocytopenia, which significantly increases surgical risk[1,2]. Renal transplantation in such patients poses unique challenges.

First, vascular anatomy is often distorted due to extensive collateral formation, making surgical dissection difficult[3]. Second, severe thrombocytopenia increases the risk of intraoperative and post-operative bleeding and thirdly, portal hypertension may predispose to haemodynamic instability[4].

Simultaneous portal decompressive procedures, such as mesocaval or lienorenal shunts, can alleviate portal hypertension and improve hypersplenism[5]. In the present case, combined shunt surgery likely contributed to improved platelet counts and reduced perioperative complications.

There is limited literature describing renal transplantation in patients with EHPVO. Most reported cases emphasize staged procedures; however, simultaneous surgery, as in this case, may reduce overall morbidity by avoiding multiple interventions.

Immunosuppression management is also crucial, as these patients may have altered liver function and increased infection risk and our more prone for bleeding post-operatively. In our patient, standard triple immunosuppression was well tolerated without complications.

In the presence of pancytopenia with an increased risk of bleeding and bone marrow suppression, the choice of induction agent – rabbit anti-thymocyte globulin (rATG) versus basiliximab – requires careful consideration. While rATG provides potent anti-rejection efficacy in the immediate post-operative period, it is associated with a higher risk of cytopenias and infections. In contrast, basiliximab, though relatively less potent, offers a safer profile in patients with pre-existing pancytopenia[6]. Therefore, a balanced risk – benefit assessment is essential when selecting the induction strategy.

This case highlights the importance of a multidisciplinary approach involving hepatobiliary surgeons, renal transplant surgeons, and nephrologists to achieve optimal outcomes in complex cases of EHPVO with end-stage renal disease undergoing transplantation.

This case underscores the importance of:-

- Careful preoperative planning
- Multidisciplinary collaboration
- Expertise in complex vascular and transplant surgery

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