Management of Bilateral Ureteral Obstruction After Transplantation of Pediatric En Bloc Kidneys, a Case Report and Review of Available Literature

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There is a large discrepancy between the number of organ donors and number of patients on the kidney transplant list. Based on Organ Procurement and Transplantation Network data as of January 14, 2019, over 94,886 patients are on the waiting list for a kidney transplant in the United States. The growth in kidney transplantation has been sluggish, with roughly only 21,167 cases performed in 2018. The number of living-donor procedures performed has also remained stagnant for several years. Patients who are unable to receive transplants face high mortality and morbidity related to end-stage renal disease needing dialysis. Such discrepancy between organ supply and demand prompts the necessity for novel sources of donors using an expanded criteria for donor organs. Pediatric donors provide a resource to increase the number of kidneys available for transplant. As a group, they have also proven to provide equivalent or superior long-term outcomes compared with standard adult kidneys despite an increase in associated surgical risks due to the potential for ureteral complications, graft thrombosis, and concerns regarding low nephron mass.1–4 Overall, kidneys donated from pediatric donors have demonstrated favorable outcomes.4 Ureteral complications happen because of multiple reasons including congenital abnormalities. In this article, we describe one example of such complication that was diagnosed after the transplant was performed. There has not been an established precedent on how to manage a ureteral pelvic junction obstruction (UPJ)/ureteral stricture in pediatric kidneys posttransplantation. Standard maneuvers were attempted at first. When these measures failed, we had to modify our approach, and after several disappointments our patient finally had success.

CASE REPORT

Patient is a 50-year-old Caucasian female with chronic kidney disease secondary to medullary cystic kidney disease. She was placed on waitlist for deceased donor renal transplant. She received preemptive kidney transplant about 6 months later. Donor was a 11-month-old boy who died of head trauma from motor vehicle collision with terminal creatinine of 0.2 mg/dL and Kidney Donor Profile Index of 68%. Complement-dependent cytotoxicity test and flow T and B cell crossmatches were negative with 2A, 2B, and 1 human leukocyte antigen DR isotype mismatch between donor and recipient. Donor was reported to have a past medical history significant for other congenital anomalies such as tetralogy of Fallot requiring surgical repair shortly after birth. Abdominal computed topography (CT) for trauma evaluation in the donor did not report any renal injuries or anomalies. Both kidneys from this pediatric donor were transplanted en bloc to the patient’s right external iliac vessels. Both ureters were implanted to bladder separately with prophylactic transplant ureteral stents. Cold ischemia time was 12 hours and 19 minutes. Warm ischemia time was 27 minutes. Patient received thymoglobulin induction at a cumulative dosage of 6 mg/kg followed by triple immunosuppressive regimen of tacrolimus, mycophenolate, and prednisone. She had uneventful postoperative course with immediate allograft function. Preoperative creatinine was 4.5 mg/dL. It progressively decreased to 2.6 mg/dL by postoperative day 10.
7, 2.1 mg/dL on day 14, and 2.0 mg/dL on day 30. Serial transplant renal ultrasounds were unremarkable. About 6 weeks postop, she presented with fever and urinary tract infection. Transplant renal ultrasound and CT showed hydronephrosis of both kidneys with partially dislodged stents (Figure 1). She underwent cystoscopy with removal of transplant ureteral stents along with antibiotic treatment for urinary tract infection. The initial thought was that the partially dislodged stents were causing obstruction. Her creatinine remained in the 1.8–2.0 mg/dL range, and ultrasounds showed severe persistent hydronephrosis. Technetium-99 mercaptoacetyltriglycine (MAG3) nuclear medicine renal scan confirmed obstructive uropathy with half clearance times of 36.9 minutes for the superomedial kidney and 26.2 minutes for the inferolateral kidney. Cystoscopy with retrograde pyelogram was performed, which showed bilateral ureteropelvic junction obstructions of both kidneys, and bilateral stents were placed with extreme difficulty. She underwent retrograde exchange of stents about every 3 months for about a year. She then had bilateral nephrostomies placed (Figure 2). The stents temporarily resolved the hydronephrosis; however, they also caused significant scarring of the distal ureters. Given the worsening hydronephrosis of superomedial kidney, she underwent laparotomy with native ureter to superomedial renal pelvis anastomosis. Her hydronephrosis persisted despite this intervention. This is likely due to anomalous/bifurcated renal pelvis. Over the next 2 years, she underwent conversion of nephrostomies to percutaneous nephroureteral stents, serial cutting balloon-assisted ureteroplasty, and progressive upsizing of nephroureteral stents to 16 French size (Figure 2). Eventually, about 3 years posttransplant, tube capping trial and nephrogram were performed prior to removal of the percutaneous nephroureteral stents to confirm collecting system patency and resolution of the obstructive uropathy. Her serum creatinine improved to 1.2 mg/dL range during second year posttransplant and further improved to 0.8 mg/dL during third year posttransplant. Nephroureteral stents and nephrostomies were eventually removed just under 3 years posttransplant. She is now reaching her fifth year anniversary posttransplant and enjoys a serum creatinine of 0.8 mg/dL. Today her ultrasound continues to show no evidence of hydronephrosis with full growth to adult size organs (Figure 3).

DISCUSSION

To our knowledge, bilateral ureteral obstruction after pediatric en bloc kidney transplantation has not been reported in the past. In our case, the etiology is definitely not clear. Mechanical, ischemic, or congenital anomalies are possible causes for ureteral obstruction. Other confounders such as an injury from dislodged ureteral stents cannot be ruled out. Donor medical history was significant for other congenital anomalies such as tetralogy of Fallot, although its association with bilateral ureteral obstruction has not previously been reported in literature. Additionally, we could not verify the radiologist’s reading of no obvious renal abnormalities on the donor’s abdominal CT, due to the inaccessibility of the CT images. It is possible that partial obstruction could have been missed during the radiologist’s initial interpretation. Mechanical kinking from torsion was excluded with cross-sectional imaging. Ischemic causes and crossing vessels are also a possibility; however, the initial dynamic images seemed to suggest that

FIGURE 1. Partially dislodged stents and hydronephrosis shown on imaging. A, Ultrasound showing severe hydronephrosis of the en bloc transplanted kidneys and (B) coronal noncontrast CT scan of transplanted kidneys with severe hydronephrosis. CT, computed topography.
Congenital UPJO is a more likely cause as the distal ureters looked normal in initial images and developed strictures after further instrumentation.

Congenital UPJO are rare entities that are found in 1 to 1000–1500 live births of newborn infants. It is about 4 times more common in male infants. The management of this anomaly has
been well established in the literature. Dismembered pyeloplasty has been considered the gold standard surgical intervention. The new reality of blood supply after transplantation as well as the unpredictability of the kidney anatomy after growth for a year or so makes these options very unlikely to succeed and fraught with complications.

Percutaneous nephrostomies have been thought of as temporary measures as a bridge to surgical management. We, however, described using aggressive, successive, and rather rapid serial dilation of the ureteral obstructions. This, accompanied by oversized nephroureteral stents, succeeded in resolving the stenosis. The kidney function was preserved in that manner.

This procedure has been attempted once before in adult kidneys, but never, to our knowledge, in pediatric en bloc kidneys. The pathophysiology may be different. As in adult kidneys, the etiology is usually acquired scarring from infectious or ischemic insults. Also crossing vessels can play a factor. Antegrade dilation has been described for lower ureteral abnormalities posttransplant. This, however, was not described for congenital cases of bilateral ureteral obstruction.

CONCLUSIONS

Utilization of pediatric donor kidneys comes with a number of inherent risks, one of which is unidentified congenital anomalies that could lead to bilateral ureteral obstructions. This case is an example in which an anomaly was encountered and could have led to graft loss. The morbidity of management of an anomaly like ureteral obstruction posttransplant is significant, and persistence is needed. This is especially true because there is no established precedent for management. The events of this case will hopefully shed a light on this potential pitfall and guide management in future cases.

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