

*Case report***Calcified Double J Stent after Sequential Liver and Renal Transplantation Associated to Primary Oxalosis: Case Report**Ayse Sinangil¹, Vedat Celik¹, Soykan Barlas², Fatih Altunrende³, Emin Baris Akin² and Tevfik Ecdar¹¹Division of Nephrology, Department of Internal Medicine, ²Renal Transplantation Unit, ³Division of Urology, Istanbul Bilim University, Istanbul-Turkey**Abstract**

Hyperoxaluria type I (HPI) is a metabolic disorder secondary to liver alanine glyoxylate aminotransferase deficiency. Renal failure occurs due to the excessive production and precipitation of oxalate in the kidney. Combined liver-renal transplantation is the correct treatment for this condition when end-stage renal failure occurs since in renal transplantation alone the risk of recurrence of the same pathology in the transplanted kidney would be high. We determined the calcification surrounding the double J stent inserted to the transplant ureter in a short time in a 22-year-old patient who underwent sequential liver and renal transplantation with the diagnoses of oxalosis. In the literature we have not found papers on calcification of double J stent following combined or sequential transplantation. Although after the sequential transplantation the calcification, nephrocalcinosis, and renal stones were practically not of great concern, these patients should be followed up more carefully in terms of stent calcification during the early post-transplant period.

Key word: hyperoxaluria, renal transplantation, double J stent

Introduction

Primary hyperoxaluria (PH) occurs due to an autosomal recessive hereditary disorder of the metabolism of glyoxylate, which causes excessive oxalate production. The most common disorder is due to deficiency of the enzyme alanine: glyoxylate aminotransferase (PH type I), that is specific to hepatic peroxisome [1-4].

The incidence of the disease is at least 1% in the pediatric population with end stage renal disease but it is higher in consanguinious marriages [5].

Considering the higher rate of intermarriages in our country, a higher proportion of such possibility may be considered. The disease leads to deposition of calcium oxalate crystals in the kidney, nephrocalcinosis, progressive renal

failure and systemic deposition of oxalate (oxalosis) [6-8]. Calcium oxalate is not effectively removed by dialysis and isolated kidney transplantation is not the method of choice due to the risk of the recurrence of nephrocalcinosis and nephrolithiasis. Overproduction of oxalate which leads to deposition of calcium oxalate crystals in the kidney continues after isolated kidney transplantation and therefore graft loss occurs frequently [9,10]. Combined liver and renal transplantation (LKT), which has relatively improved graft and patient survival, is the crucial treatment method for patients with PH [11,12]. We present a case of calcification of double J stent inserted to the transplant ureter after a successful sequential liver and renal transplantation in a patient with oxalosis.

Case report

We report a case which demonstrates the disastrous consequences of late diagnosis of hyperoxaluria in a 22-year-old man with nephrocalcinosis, a staghorn calculus and recurrent urinary tract infections. His initial management at another hospital included multiple percutaneous nephrostomies and lithopsies. He had eight siblings. His paternal uncle and one of his eight siblings had renal calculi and were hemodialysis patients. His renal function steadily worsened in the ensuing years and hemodialysis treatment was started when he was 20 years old. In this period due to recurrent severe pyelonephritis total right nephrectomy was performed. The kidney biopsy specimen showed chronic interstitial fibrosis and calcium oxalate crystal deposition was seen in the renal tubules under polarized light microscopy. These findings were consistent with oxalosis. After two years of hemodialysis treatment, he was admitted to our transplant unit for living related liver and renal transplantation from his sisters. They were all healthy; no history of nephrolithiasis and/or no recurrent urinary tract infections. Metabolic evaluation of 24 hour urine collections and genetic analysis were performed. After exclusion of oxalosis from donor candidates sequential transplantation was successfully achieved. Firstly, liver transplantation

from his sister was performed, and two months later he received kidney transplant from his other sister. Before renal transplantation the patient was underwent intensive hemodialysis treatment (two weeks daily/five hours). During renal transplant operation double J stent was inserted in the transplant ureter. ATG was given as an induction therapy and standard immunosuppressive protocol (maintenance therapy) mainly consisted of triple therapy

composed of steroids, a calcineurin inhibitor (tacrolimus) and mycophenolic acid. At the time of discharge, his serum creatinine concentration was 1.4 mg/dl and BUN was 56 mg/dl. Liver function tests were within normal limits. He was admitted to the hospital for removing of the double J stent two weeks later. During the process a calcified stent was detected (Figure 1).

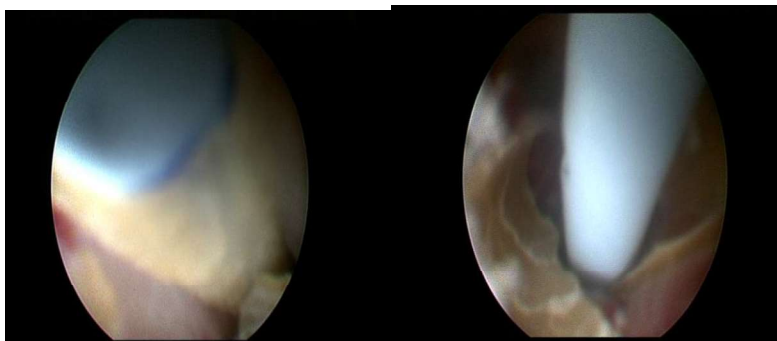


Fig. 1. Calcified double J stent

The stones surrounding the calcified double J stent inserted into the transplant ureter were successfully treated by laser lithotripsy and the stent was removed. The history of renal calculi was not detected during the post-transplant follow-up period. The patient is being still followed-up in the liver and renal transplantation outpatient polyclinics and his liver function tests are within normal limits; basal serum creatinine concentration is 1.3 mg/dl.

Discussion

The excess oxalate that is produced in PH is primarily excreted by the kidneys. Urinary calcium oxalate supersaturations occur in the presence of excess urinary oxalate excretion and this supersaturation leads to crystal aggregation, urolithiasis, and nephrocalcinosis [2]. Calcium oxalate crystals are also deposited within the renal interstitium and renal tubule cells. Nephrocalcinosis or recurrent urolithiasis can cause renal parenchymal inflammation and fibrosis and, if persistent, end-stage renal disease [2]. Other urinary complications associated with urolithiasis, such as infection and obstruction, also contribute to renal damage in affected patients. Intervention is required when stones obstruct the urinary tract. Nephrostomy, ureteroscopy, and ureteral JJ stent are preferred interventions for stone removal. Open surgical removal may precipitate acute renal failure and extracorporeal shock-wave lithotripsy (ESWL) may harm the kidney because of the potential presence of nephrocalcinosis and microlithiasis within the kidney [13]. In our case recurrent nephrolithiasis, nephrocalcinosis, hematuria, urinary infections and rapid development of renal failure are the prominent clinical manifestations. Firstly, the history of renal stone was detected at the age of seven and

eleven years and the patient underwent a couple of surgical intervention and ESWL due to the recurrent stones. The efficacy of treatment in PH is dependent on early diagnosis. Initiation of medical management as soon as possible leads to protection of renal functions, postpones the end-stage renal disease (ESRD) and minimizes nonrenal sequelae [2]. The patient had late diagnosis, after developing all complications when he was 20 years old by microscopic examination of kidney biopsy specimen obtained by total right nephrectomy due to recurrent episodes of urinary tract infection. So he had been already on regular hemodialysis when he was diagnosed. The definitive cure for PH type 1 is liver transplantation as the donor liver provides the missing enzyme, which lowers oxalate production to the normal range, but liver transplantation itself has significant complications and potential mortality. It is performed when the glomerular filtration rate is less than 40 ml/min/1.73 m². Sequential transplantation (liver followed by kidney transplantation), are performed in patients with PH type 1 and ESRD [14-16]. The rationale underlying this approach is the initial liver transplant allows intensive dialysis to clear the stores of tissue oxalates, thereby reducing the risk of kidney injury after renal transplantation [16-18]. In our case, firstly liver transplantation and two months later renal transplantation were performed from patient's two sisters.

The daily oxalate production is approximately 3500 to 7500 micromol in patients with type 1 PH, but the maximal oxalate elimination is 950 to 1440 micromol/day via conventional hemodialysis and also peritoneal dialysis [19]. As a result, standard maintenance dialysis therapy is not sufficient for lowering the plasma oxalate level. Higher plasma oxalate level increases the risk of systemic oxalosis. Intensive dialysis (eg, more than four-hour

daily HD sessions or a combination of PD and HD) is required to minimize plasma oxalate level. However, such intensive treapy is not effective to reduce the daily oxalate load. It should be instituted before renal transplantation in order to decrease the plasma oxalate level and oxalate deposition which leads to injury of the renal allograft [20]. Before renal transplantation the patient was taken to the intensive hemodialysis treatment (two weeks daily/five hours). Double J stent was inserted into the transplant ureter during the surgery but stent calcification of was observed within a short period (two weeks).

Mobilization of tissue oxalate deposits ensues gradually after transplantation and high urinary oxalate excretion may persist even more than two years after transplantation until tissue stores are removed [21].

In our case, double J stent might have been calcified because of the possibility of elevating urinary oxalate excretion.

We should be careful in terms of calcification of double J stent, inserted during the combined or sequential liver and renal transplantation, which is the actual treatment of oxalozis. During the early period after transplantation they should be closely monitored.

Conflict of interest statement. None declared.

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