Post-Renal Transplant Lymphorrhea and Lymphocele: An Unusual Association with Autosomal Dominant Polycystic Kidney Disease

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Abstract

Lymphorrhea and lymphocele are unusual post renal transplant complications; Lymphorrhea is lymphatic fluid leakage around renal allograft, while lymphocele is cavity of lymphatic fluid collection around graft, lacking epithelial lining, caused by severed lymphatic vessels during transplant surgery. Numerous etiological factors had been defined and the association with Autosomal Dominant Polycystic Kidney Disease (ADPKD) is accepted as a risk factor, but mechanism is still to be established. A young doctor who himself under went renal transplant surgery due to end stage renal disease by ADPKD. He experienced the symptomatic lymphorrhea and underwent various stepwise therapeutic options and faced variety of complications. The treatment of Lymphorrhea and lymphocele evolved with time and experience and now laparoscopic fenestration is well established technique with minimal morbidity.

Keywords: Lymphorrhea; Lymphocele; Autosomal dominant polycystic kidney disease (ADPKD)

Introduction

Post renal transplant lymphorrhea and lymphocele were first documented in 1968 [1]. Lymphorrhea is the lymphatic fluid leakage around renal graft (perivascular and/or hilar) caused by severed lymphatic vessels during transplant surgery, while lymphocele is cavity of lymphatic fluid collection around graft, lacking epithelial lining. The meticulous dissection for preparation of iliac vessels of recipient damages the lymphatic channels or otherwise dissection of renal vessels of donor results in lymphocele [2]. The overall incidence of lymphorrhea documented in different studies varies between 0.5% to 33.9%.

Various etiological factors has been established like acute allograft rejection, delay in graft function, graft biopsy, acute tubular necrosis, administration of heparin, cyclosporine and mechanistic Target of Rapamycine (mTOR) based immunosuppression, Cytomegalovirus infection, re-transplantation, edema of the lower extremities and adult polycystic kidney disease as an original disease [3-5]. Lymphatic complications may be related to diabetes, obesity, blood coagulation abnormalities, anticoagulation prophylaxis, high dose of diuretics, and immunosuppressive drugs.

Post-transplant lymphocele in ADPKD is belief due to presence of polycystic kidney in retroperitoneal space leading to increase in the abdominal pressure and post-operative retroperitoneal scarring and fibrosis impair the lymphatic drainage. Role of proteins, albumin and colloid oncotic pressure has a key role in the development of lymphocele but detailed understanding needs more work to establish understanding [6].

Most lymphoceles are small and asymptomatic or improved unreported. In small lymphocele physical examination is usually unremarkable and detected by routine ultrasonography. Sometimes it may be large size and manifest as mild heaviness or fullness in flank. The present case study highlights the association of polycystic kidney disease leading to chronic kidney disease followed by allograft kidney transplant, lymphorrhea and its management. The study highlights the personal experience of author’s presentation, diagnosis and use of different treatment modalities.

Case Presentation

A 32-year-old male young doctor was diagnosed as a case of hypertension. The primary investigation revealed bilateral polycystic kidneys. In due course of time he developed chronic kidney
disease at the age of 50 years. He was advised for renal transplant for his deranged renal profile. His tissue matching was compatible with his wife. Preliminary investigations revealed left anterior descending coronary artery was blocked 70%. Non medicated coronary stenting was successfully performed. Meanwhile ethical committee approved his request of transplantation.

The surgery was planned. Right nephrectomy was done and haemodialysis started. After few days of nephrectomy, renal transplant surgery was performed. Post renal transplant immunosuppression started with corticosteroids, tacrolimus (macrolide) and mycophenolate mofetil (inosine monophosphate dehydrogenase inhibitor). During early post-renal transplant period he developed lower abdominal discomfort with continuous fluid discharge in the intra-abdominally placed drain. Ultrasonography showed a fluid collection at the lower pole of transplant kidney. The draining fluid analysis revealed that it’s a lymph fluid. Percutaneous catheter drainage of fluid around the kidney was performed. Wait and watch policy with stepwise approached was adopted. In a due course of time the amount of fluid reduced up to 50 ml in 24 hrs. The drain was removed.

After ten days, abdominal discomfort again started and gradually increased. Ultrasonography revealed collection of fluid around the renal transplant lead to mild hydrenephrosis. The renal profile was also disturbed. They planned for placement of percutaneous catheter under ultrasound guidance for lymphocele drainage and to relieve the pressure effects on the graft. The renal profile normalized next day. The accumulated fluid, drained was more than 500 ml from lymphocele, pigtail catheter drain kept for next one month. Aspirated fluid was sent for bacteriological and chemical examination and has blood urea nitrogen and creatinine and electrolytes level similar to plasma. For this period the draining fluid decreased to 80 ml from 300 ml/24 hrs. Trans-catheter sclerotherapy was planned. Bleomycin 60 ml diluted in normal saline was inserted through catheter and kept for 12 hrs. Unfortunately systemic absorption of Bleomycin manifested with high grade fever, severe rigors and persistent vomiting. Initial investigations revealed pancytopenia but renal profile was normal. He was managed conservatively and advised to wait for one month again for fluid to decrease in amount.

Following one month of Bleomycin therapy, Lymphorrhea failed to become less than 100 ml/24 hrs. Laparoscopic peritoneal window was planned and performed successfully. He was improved and became asymptomatic. 31 months follow up with repeat base line laboratory and Doppler ultrasounds were normal.

Discussion

Autosomal Dominant Polycystic Kidney Disease (ADPKD) is a hereditary disease with prevalence of 1:400 to 1:1000 in white population. The gene responsible in 85% of cases PKD1 located on chromosome 16, while remaining associated with PKD2 located on chromosome 4. In PKD2 progression to end stage kidney disease is 10-15 years later as compared to PKD1. Our patient developed renal failure at the age of 50 years after diagnosis of ADPKD in 32 years. Renal replacement therapy is needed in 5% to 10% of patients suffering from ADPKD [7].

There is a general consensus that pre-transplant native nephrectomy, especially bilateral nephrectomy, unless serious complications like infection or bleeding arise. Others suggest a ‘sandwich’ technique with single-sided nephrectomy before and removal of the remaining polycystic kidney after transplantation. In our patient right sided nephrectomy was performed before renal transplant. The left kidney without any complication was not operated. In a study conducted by Neeff HP, on 3156 patients of renal transplant 9% followed by ADPKD with end stage renal failure and lymphocele was developed in 4% of cases [8-11].

The treatment of lymphorrhea and lymphocele evolved with time and experience. The options included is simple aspiration or external drainage and instillation of sclero therapeutic agent, internal drainage either opens surgical or laparoscopic. [12] among these various treatment options recommended worldwide, no gold standard exists. Per cutaneous simple aspiration is associated with 50% to 100% recurrence rate, while additional sclerotherapy instillation in lymphocele cavity reduces the rate to 6% to 25%. Bleomycin, fibrin glue, povidone iodine or ethanol instillation with or without addition of factor XIII and fibrinogen sodium tetradecyl sulfate, Au colloid, tetracycline are usual sclero therapeutic agents used. The first description of laparoscopic fenestration in 1991 is considered to be the optimal treatment tool now, replacing the convention laparotomy. Recent studies favour the laparoscopic deroofing of lymphocele with complete cure [13]. The treatment options depend upon symptoms, renal functions and ultrasound findings.

In our patient stepwise approach was adopted. Initially conservative treatment was done. Later on when pressure effects observed on renal graft and renal profile is deranged, ultrasound guided aspiration and continuous external drainage done. Bleomycin instillation was performed in the next step of stepwise management. Bleomycin systemic absorption leads to toxic manifestation. Later on laparoscopic internal drainage was performed, which was successful.

Conclusion

Transplant surgeons must pay particular attention during surgery of renal vascular pedicle in both graft and recipient to avoid the dissection of the lymphatic vessels. The symptomatic lymphocele should be early treated by laparoscopic surgery fenestration procedure rather than repeated percutaneous aspiration or external drainage and instillation of chemical agents in lymphocele cavity, which not only effects the quality of life of transplanted patient but also increase the risk of life threatening infections.

Specifically, the autosomal dominant polycystic kidney disease leading to end stage renal disease needs renal transplant and complicating in lymphorrhea and lymphocele needs further clinical studies to establish the association, mechanism of development and prevention.

References


