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A Case of Nephrotic Syndrome Presenting with Pulmonary Embolus in a Kidney Transplant Patient

Böbrek Nakli Sonrası Pulmoner Emboli ile Prezente Olan Bir Nefrotik Sendrom Olgusu

ABSTRACT

Recurrence of glomerulonephritis (GN) is considered an important cause of allograft failure in kidney transplant recipients. It can present after transplantation with complications which are difficult to manage.

A 53-year-old woman was admitted to the hospital with abdominal pain and swelling in the right lower quadrant at the first month of kidney transplantation. Following hospitalization, she complained of sudden onset dyspnea and chest pain. Thorax CT examination revealed pulmonary embolism and anticoagulation was started. The 24-hour urine proteinuria was quantified at 16695 mg/day and she was diagnosed with nephrotic syndrome due to a low albumin value. However, kidney biopsy could not be performed due to a bleeding tendency. With a probable diagnosis of recurrent focal segmental glomerulosclerosis, plasmapheresis was performed. A total of 8 plasmapheresis sessions resulted in full recovery regarding the proteinuria.

Recurrence of glomerulonephritis and development of pulmonary embolus due to nephrotic syndrome are well known complications but difficult to manage. Nephrology follow-up after transplantation is crucial.

KEY WORDS: Nephrotic syndrome, Pulmonary embolus, Kidney transplantation

ÖZ

Nüks glomerülonefrit, böbrek nakilli hastalarda allograft kaybının önemli bir sebebidir. Nakil sonrası yönetimi zor komplikasyonlarla ortaya çıkabilir.

53 yaşında kadın hasta kliniğimize böbrek naklinden 1 ay sonra gelişen karın sağ alt kadranda ağrı ve şişlik şikayetiyle başvurdu. Hastanın yatışı yapıldıktan sonra dispne ve göğüs ağrısı şikayetleri gelişti. Çekilen toraks BT pulmoner emboli olarak yorumlandı ve hastaya pulmoner emboli tanısıyla antikoagülasyon başlandı. Hastanın 24 saatlik idrarında proteinüri 16695 mg/gün olarak saptandı, albümin seviyesi düşen hastaya nefrotik sendrom tanısı konuldu ancak, kanama olasılığı yüzünden böbrek biyopsisi yapılmadı. Olası nüks fokal segmental glomerüloskleroz tanısı ile plazmaferez başlandı. 8 seans plazmaferez sonrası proteinüride tam remisyon sağladı.

Böbrek nakli sonrası glomerülonefrit nüksü ve nefrotik sendrom zemininde plumoner emboli gelişmesi bilinen ancak yönetimi zor komplikasyonlardır. Nakil sonrası nefrolojik takip elzemdir.

ANAHTAR SÖZCÜKLER: Nefrotik sendrom, Pulmoner emboli, Böbrek nakli

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INTRODUCTION

Recurrence of glomerulonephritis (GN) following kidney transplantation is a well-known complication and is considered to be an important cause of allograft failure in kidney transplant recipients (1). Occasionally, it can present with a devastating clinical picture and the management can be difficult (2). In this report, we present a kidney transplant patient with pulmonary embolus (PE) due to severe nephrotic syndrome.

CASE REPORT

A fifty-three-year-old female patient who had a living donor kidney transplantation from her husband one month ago (May 2017) presented to the outpatient clinic with abdominal pain and swelling in the right lower quadrant. She had a history of left radical nephrectomy during childhood. The cause of nephrectomy was not documented. The patient had been followed-up by our nephrology outpatient clinic since 2010. Initially, she presented with proteinuria in the nephrotic range, hypoalbuminemia, oedema and hyperlipidaemia. A kidney biopsy was scheduled. However, kidney biopsy could not be performed because of cysts in her single kidney. Her kidney function deteriorated gradually until hemodialysis was initiated in January 2017.

During investigation of abdominal pain, a 16x8 cm of homogeneous fluid collection was detected between the urinary bladder and transplanted kidney in the right lower quadrant during ultrasonography. The patient was hospitalized for further investigation of the fluid. A sample from the abdominal fluid obtained by puncture was evaluated as transudate.

Four days after admission to the hospital, she complained of sudden onset chest pain and dyspnea. With clinical suspicion of pulmonary embolus (PE), a thorax CT was performed and the radiological findings were consistent with PE. Anticoagulation with enoxaparin 0.6 ml bid subcutaneously was started. On echocardiographic examination, the ejection fraction was 55% and a thrombus was suspected in the right atrium. Transoesophageal echocardiography did not show vegetations or thrombi. A work-up for thrombophilia profile was performed. Protein S (81.5%), antiphospholipid IgG (1.26 U/ml), antiphospholipid IgM (0.26 U/ml), anticardiolipin IgG (1.05 GPL U/ml) and anticardiolipin IgM (0.69 MPL U/ml) levels were normal and protein C (159.6%) was high. Her laboratory findings were as follows: urea 50 mg/dl, creatinine 1.09 mg/dl, serum albumin 4.03 gr/dl, hematocrit 35%, leucocytes 6100/mm^3, and platelets 181000/mm^3. Urinary sediment revealed 4 leucocytes and 7 erythrocytes per high power field and urinary protein excretion was positive. After 24-hour urine collection, proteinuria was present at 16695 mg/day and microalbuminuria 12709 mg/day. Activated partial thromboplastin time was 30.8 seconds and international normalized ratio (INR) was 0.83. Her blood albumin level had decreased to 3.43 mg/dl.

A diagnosis of nephrotic syndrome with a possible cause of focal segmental glomerulosclerosis (FSGS) was considered. However, because of the anticoagulation therapy and risk of bleeding, a biopsy could not be done. Based on this clinical judgment and also considering the high probability of FSGS, eight plasmapheresis sessions were applied in 4 weeks. Eventually, the proteinuria regressed significantly and the clinical picture of nephrotic syndrome remitted completely. The final laboratory findings revealed that the proteinuria had decreased from 16695 mg/day to 348 mg/day and microalbuminuria from 12709 mg/day to 187 mg/day.

DISCUSSION

Thrombosis in patients with nephrotic syndrome can arise from many causes including enhanced platelet activation and aggregation, enhanced activation of the coagulation system via accumulation of high molecular weight coagulation factors, decreased endogenous anticoagulants, and decreased activity of fibrinolytic system. Intravascular volume depletion due to nephrotic syndrome, changes in the glomerular hemostatic system and exposure to corticosteroids are other exacerbating factors for a thromboembolic event (3,4).

Glomerulonephritis is a common cause of allograft failure in up to 20% of kidney transplant recipients. Nevertheless, the risk of recurrence depends significantly on the primary glomerular disease (1). In recent years, the establishment of protocol biopsy programs and registry databases have provided more precise data regarding the incidence and impact of recurrent GN on allograft failure. Post-transplant proteinuria is a common finding which has a significant impact on allograft failure and patient survival (5,6). In our case, because of a single kidney being present prior to transplantation and being on anticoagulation treatment after transplantation, a kidney biopsy could not be performed. Therefore the exact pathological diagnosis of the kidney disease is not known. However, focal segmental glomerulosclerosis (FSGS) is considered based on the clinical evidence and rapid response to the treatment. Additionally, it is also well known that FSGS recurs in 30-50% of kidney recipients and is associated with 2 types of clinical presentation. More commonly it occurs within hours or days after transplantation and results in rapid allograft loss if left untreated. Neither post-transplant duration of hemodialysis nor the immunosuppressive treatment choice changes the probability of graft survival or recurrence of FSGS (7,8). It can also have an insidious onset and develops in months or years and presents similarly to the early recurrence (7,8). An undefined permeability factor is considered in the pathogenesis of recurrent GN after kidney transplantation and plasmapheresis is therefore recommended in the treatment. In a meta-analysis of 423 patients, 71% of patients showed a complete or partial remission after the appropriate use of plasmapheresis for the recurrence of FSGS after kidney transplant (9).

In conclusion, we have discussed the treatment of a patient with a probable FSGS diagnosis who presented with recurrent GN, nephrotic syndrome and pulmonary embolism one month after a kidney transplant in this report. Plasmapheresis treatment ensured remission of the disease.

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