

Idiopathic Retroperitoneal Fibrosis Causing Obstructive Nephropathy and Involving Surrenal Gland: A Case Report

Obstrüktif Nefropatiye Neden Olan ve Sürrenal Glandı Tutan İdiyopatik Retroperitoneal Fibrozis: Olgu Sunumu

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ABSTRACT

A 41-year old male patient was admitted to our hospital with the complaints of reduced urine, swelling and increased urea and creatinine values. Bilateral hydronephrosis was detected by ultrasonography, but the surrounding tissues couldn't be clearly examined. Ureteral catheter and then bilateral percutaneous nephrostomy were applied. The patient's complaints reduced in the follow-up period. The intravenous urogram was performed to explain the reason of hydronephrosis. The left kidney was nonfunctioned while the right kidney was functioning. An abdominal computed tomography scan showed a soft-tissue mass encasing the aorta, the left ureter and left surrenal gland and causing hydronephrosis. The appearance observed by magnetic resonance imaging scan was considered consistent with the diagnosis of retroperitoneal fibrosis. The entire body Ga-67 scintigraphy demonstrated an uptake in the same area. Steroid treatment was started to the patient. A clinical recovery was observed during the follow-up period. Regression in retroperitoneal fibrosis was confirmed by the post-therapy abdominopelvic computed tomography scan and Ga-67 scintigraphy.

In addition to malignancies, retroperitoneal fibrosis should be considered in the differential diagnosis of an abdominal mass observed in case with acute renal failure secondary to obstructive nephropathy.

Keywords: Acute renal failure, retroperitoneal fibrosis, obstructive nephropathy, surrenal gland

ÖZET

Kırk bir yaşında erkek hasta, idrarda azalma, tüm vücutta şişlik, üre ve kreatinin değerlerinin yüksekliği nedeni ile kliniğimize başvurdu. Ultrasonografide bilateral hidronefroz tespit edilen ve çevre dokular net değerlendirilemeyen hastaya üreteral kateter ve daha sonra bilateral perkütan nefrostomi kateteri takıldı. Takiplerde yakınmaları kaybolan hastanın hidronefrozunun nedenini açıklamak amacı ile yapılan intravenöz pyelografi incelemesinde sol böbrek nonfonksiyone, sağ böbrek fonksiyone idi. Abdominal bilgisayarlı tomografi incelemesinde aorta, sol üreter ve sol sürrenal glandı saran, aynı zamanda hidronefrozise neden olan yumuşak doku kitlesi saptandı. Manyetik rezonans incelemede saptanan bulgular retroperitoneal fibrozis ile uyumlu olarak kabul edildi. Yapılan tüm vücut Ga-67 sintigrafisinde, manyetik rezonans inceleme ile saptanan bölgelerde tutulum görüldü. Hastaya steroid tedavisi başlandı. Hastada izlem süresinde klinik olarak düzelmeye gözlemlendi ve abdominopelvik bilgisayarlı tomografi ve Ga-67 incelemesi de gerilemeyi doğruladı.

Obstrüktif nefropatiye bağlı gelişen akut böbrek yetmezliği olgularında, batında kitle tespit edildiğinde, malignensilere ek olarak retroperitoneal fibrozis de ayırıcı tanıda yer almalıdır.

Anahtar kelimeler: Akut böbrek yetmezliği, retroperitoneal fibrozis, obstrüktif nefropati, sürrenal bez

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Introduction

Retroperitoneal fibrosis (RPF) is a rare entity that develops with several pathologies and results in fibrosis in the retroperitoneal area (1,2). Renal failure develops in a vast majority of patients (%75) who have RPF (3). This

usually occurs due to the unilateral or bilateral ureteral obstruction caused by the fibrous tissue. The obstruction is generally observed in the middle or lower 1/3 part of the ureter (4). Various symptoms such as abdominal or back pain, malignant hypertension and pedal edema may be seen (1). The classical triad is as follows; hydronephrosis, medial deviation of the ureter and shrinkage of the ureter owing to the thickening of the surrounding tissue of the middle segment of the ureter (1). Here we present a case of RPF causing obstructive nephropathy and involving surrenal gland.

Case

A 41 year-old male patient was admitted to our clinic in September 2003 with complaints of reduction in urine, swelling in the body and high urea and creatinine values. Two months ago, an abdominal computed tomography (CT) scan was performed in our clinic when the patient complained of weight loss. CT showed an extensive soft-tissue mass encasing the aorta, the left ureter and left surrenal gland and causing hydronephrosis. The laparotomic biopsy had been diagnosed as a chronic lymphadenitis. The laboratory values were normal. Physical examination indicated 140/80 mmHg blood pressure. Physical examination showed a left axillary and cervical lymphadenopathies measuring approximately 1x1 cm. Bilateral pretibial (3+), pedal (3+) and periorbital edema were also observed. Urethral catheter was applied, but the patient was anuric.

Hemoglobin level was 10.9 g/dl, erythrocyte sedimentation rate was 86 mm/h, and white sphere and platelets were normal. Biochemical values were as

follows; serum urea nitrogen 85 mg/dl (range 5-20), creatinine 15.78 mg/dl (range 0,5-1,4) potassium 5.84 mEq/L (range 3,5-5,5), total protein 7.5 g/dl (range 6-8.5) albumin 3.8 g/dl (range 3,5-5), phosphorus 7.3 mg/dl (range 2,7-4,5), calcium 9.6 mg/dl (range 8,5-10,5), uric acid 9.3 mg/dl (range 2,3-7), and serum electrophoresis yielded normal albumin band. Coagulation parameters, aspartate and alanine aminotransferase, gamma-glutamyl, alkaline phosphatase, amylase, lipid profile, thyroid hormones, Beta-hCG and other hormones, iron parameters, tumor determinants, and direct urine analysis were normal. In the 24-hour urine analysis, proteinuria, which is not at the nephrotic level was detected. (360 mg/G). In the serological tests; antinuclear antibodies, human immune deficiency virus, hepatitis B and C, p and c-ANCA were negative and anti DNA, C3, C4 were normal.

In the abdominal ultrasonographic examination, right kidney was measured as 137x61 mm, the left kidney as 122x71 mm, the parenchyma echogenity was increased (grade 1), and hydronephrosis was detected in the right kidney and hydroureteronephrosis in the left kidney. Although it was urgently tried to apply bilateral ureteral catheter (Double J stent) to the patient, right ureteral catheter was placed, but the left ureteral catheter was not. The patient commenced to urine output and urea nitrogen and creatinine levels returned to normal rapidly. Urea was measured as 15 mg/dl, creatinine as 1,4 mg/dl and edema disappeared.

The abdominal pelvic CT scan and magnetic resonance imaging scan (MRI) performed for control purpose, showed that the soft tissue mass locating in the paraortic region, surrounding the left ureter and

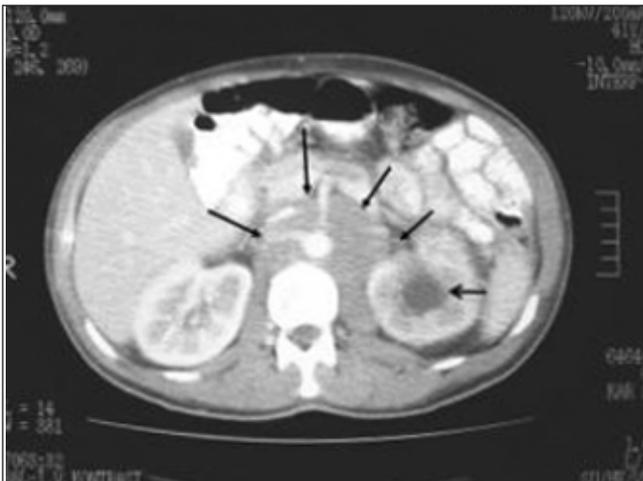


Figure 1. Abdominal CT showing retroperitoneal fibrosis and involvement of the left kidney.

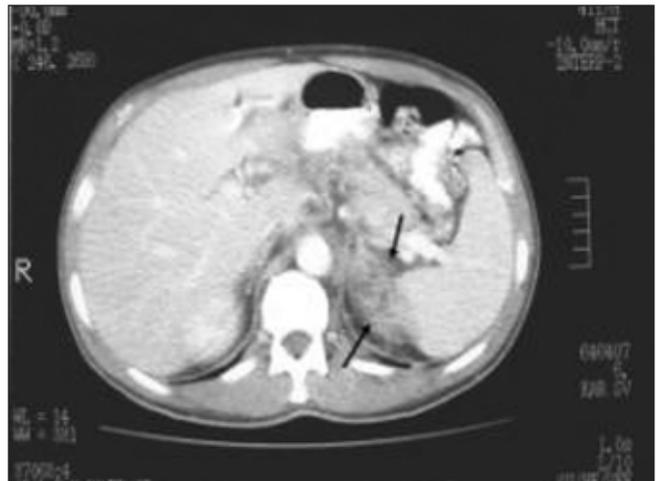


Figure 2. Abdominal CT showing retroperitoneal fibrosis involving the left surrenal gland.

involving left surrenal gland persisted. No significant increase in size of the mass was observed when comparing to the earlier CT. The radiologic appearance was considered consistent with retroperitoneal fibrosis, and involvement of the left kidney and surrenal gland was reported (Figure 1-2). In the Ga-67 scintigraphy, an uptake was observed in these regions. Intravenous pyelography showed that the right kidney was functioned and hydronephrotic and the left kidney was nonfunctioned. The left ureter could not be visualized. Scintigraphic examination showed that the right kidney was 70% functioned and the left kidney 30% functioned. Twenty days after the placement of ureteral catheter, the urine decreased and the urea-creatinine values were increased. Retrograde pyelography showed that there was no intraluminal obstruction. Upon the examination of the patient, ureteral catheter dysfunction was considered (due to pressure on ureters) and bilateral nephrostomy catheter was placed. The patient started to urine output again; the right nephrostomy catheter was functional and the left nephrostomy catheter was non-functional. Biochemical parameters returned to normal. For the treatment of RPF, methylprednisolone, 500 mg/day for 3 days and then, prednisolone, 20 mg/day were administered to the patient. On the 3rd day of the treatment, it was observed that urine output started through the left nephrostomy catheter and the bladder tract. Regression in RFP was observed in the post-therapy abdominopelvic CT and Ga-67 scintigraphy.

Discussion

RPF is idiopathic in 60 to 70% of the cases. In the remainder the more common associations include drugs, malignancies, periaortitis and abdominal aorta aneurysm (1). It may also be associated with systemic inflammatory conditions (e.g. SLE, vasculitis, collagen vascular diseases, sarcoidosis). Recently, the association of RPF with other diseases, including ANCA-associated RPF (5-7), sclerosing pancreatitis-associated RPF (8), and primary biliary cirrhosis-associated RPF (9) has also been reported in the literature. In our patient, as the serological tests were negative, the urine sediment was normal and proteinuria was absent, the glomerular pathologies were excluded, therefore renal biopsy was not done. Direct pulmonary graphy and thorax CT were normal and then sarcoidosis was excluded.

Renal involvement in RPF generally occurs through extrinsic compression and as a result of ureteral distorsion. Rarely, obstructive nephropathy develops through the

direct inflammation of the ureter. In both conditions, ureterolysis or stent application alone may not be successful in the treatment (10). Steroid treatment individually or combined with this treatment, known to be effective (1,11). In our case, left ureteral stent application could not be performed due to extrinsic pressure. Retrograde pyelography showed that there was no intraluminal obstruction. The urine output of the patient decreased and the renal functions deteriorated. Since the right ureteral stent become non-functional due to extrinsic pressure, bilateral percutaneous nephrostomy catheter was placed. The renal functions of the patient improved, but the left kidney was non-functional. Following the steroid therapy, the urine output of the patient increased and the nephrostomy catheter in the left kidney started to function. Urine output through the bladder tract was observed.

In conclusion, in addition to malignancies, RPF should be considered in the differential diagnosis of an abdominal mass observed in case with acute renal failure secondary to obstructive nephropathy.

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