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*Case report***Retroperitoneal fibrosis: a case report**Kemal Magden<sup>1</sup>, Ibrahim Yildirim<sup>1</sup>, Derya Demirtas<sup>2</sup>, Utku Erdem Soyaltin<sup>2</sup>, Mustafa Gurkan Haytaoglu<sup>2</sup>, Sennur Budak Kose<sup>3</sup>, Gursel Yildiz<sup>4</sup> and Ender Hur<sup>1</sup><sup>1</sup>Department of Nephrology, Bulent Ecevit University Medical School, Zonguldak, <sup>2</sup>Department of Internal Medicine, Bulent Ecevit University Medical School, Zonguldak, <sup>3</sup>Sisli Etfal Hospital, Clinic of Nephrology, Istanbul, <sup>4</sup>Atatürk State Hospital, Clinic of Nephrology, Zonguldak, Turkey

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**Abstract**

Retroperitoneal fibrosis is characterized by development of extensive fibrosis, leading to entrapment and obstruction of retroperitoneal structures, notably the ureters. In most cases, the etiology is unknown. It is occasionally associated with autoimmune diseases. Response to corticosteroids and immunosuppressive therapy suggest it is probably immunologically mediated. The symptoms and signs associated with retroperitoneal fibrosis are non-specific, and diagnosis requires a high degree of suspicion. We should always have in mind retroperitoneal fibrosis in differential diagnosis of hydronephrosis.

**Key words:** retroperitoneal fibrosis, hydronephrosis, corticosteroid

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**Introduction**

Idiopathic retroperitoneal fibrosis is a subgroup of chronic periaortitis. Chronic periaortitis presents usually with a fibroinflammatory mass which surrounds the abdominal aorta and iliac arteries. Sometimes this mass causes compression of adjacent organs such as ureter and inferior vena cava. There are three subtypes of chronic periaortitis (CP); these are inflammatory abdominal aortic aneurysm (IAAAS), perianeurysmal retroperitoneal fibrosis (RPF) and idiopathic retroperitoneal fibrosis. Aorta aneurysm in the idiopathic retroperitoneal fibrosis is not usual and the retroperitoneal mass can cause compression of adjacent organs. The major difference between IAAAS and RPF is that IAAAS aneurysm sac does not cause compression on adjacent organs and obstruction. Chronic periaortitis pathogenesis is not clear. According to Parums and Mitchinson's hypothesis CP is caused by an autoimmune response directed against ceroid in atherosclerotic plaques [1,2]. The other hypothesis is that CP is a systemic autoimmune disease [3]. According to the first hypothesis patients have critical atherosclerosis. Highly positive anti-nuclear antibody and acute phase reactants are the evidence supporting the second hypothesis. However, this disease is

associated with autoimmune disorders affecting other organs, and the disease is associated with HLA-DRB1\*03. This gene is associated with other diseases such as SLE, autoimmune thyroid disease, type 1 diabetes mellitus and myasthenia gravis [4-7]. Idiopathic retroperitoneal fibrosis is a rare disease. The knowledge about the treatment of this disease is based on case reports or studies of small groups. Thus, we think it is important to present each case of idiopathic retroperitoneal fibrosis.

**Case report**

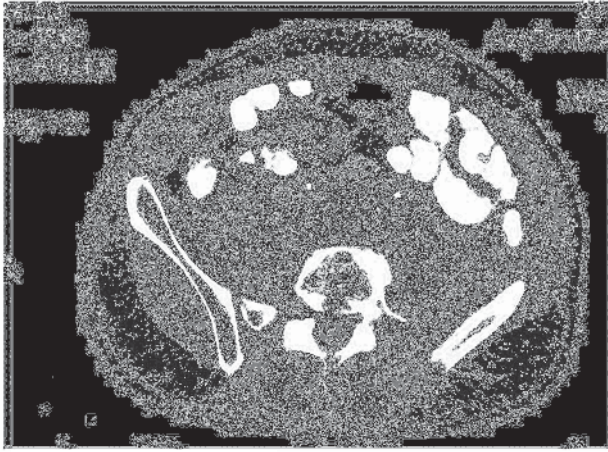
A 65-year-old male patient was admitted to the hospital with complaints of bilateral lower quadrant pain which had started 15 days ago. Creatinine and urea were 5mg/dl and 100 mg/dl, respectively. He also complained on change in bowel habits. There were no previously known kidney diseases, diabetes, hypertension, family history of chronic kidney disease, or previous pyelonephritis, urolithiasis which could explain the high levels of urea and creatinine at presentation. He had no decrease in urine output, or any symptoms of prostatism. He had not used any herbal medicine and had no history of trauma, arthritis, skin rash. Physical examination revealed good general condition; his blood pressure was 130/90 mmHg, pulse 88; body temperature 37°C, with no globe on his urinary bladder. Other system examinations were unremarkable. Uric acid was 9,5 mg/dl, Hb: 11,1 g/dl, CRP: 62 mg/dl; the electrolytes, liver function tests, anti-nuclear antibody tests were normal. Urine pH was 7,5, protein 3 (+), erythrocyte 2 (+), density 1015 on urine strip. Microscopy analysis showed 4-5 leukocytes and 14-15 erythrocytes in the urine. He had no pathological findings on chest radiograph. ECG findings were normal. Urinary catheter was inserted. After the hydration of 10 hours, 1000 cc urine output was recorded. On urinary tract ultrasonography, kidney sizes were (right: 117 x 51 mm, left: 120 x 62 mm) in normal range, parenchymal thicknesses (right: 15 mm, left: 17 mm) were normal, the level of the right renal parenchyma echogenicity increased to grade 1 and bilateral grade 2 hydronephrosis was observed. Based on these results, non-contrast abdominal computerized tomography (CT) was planned. Kidney size and contours

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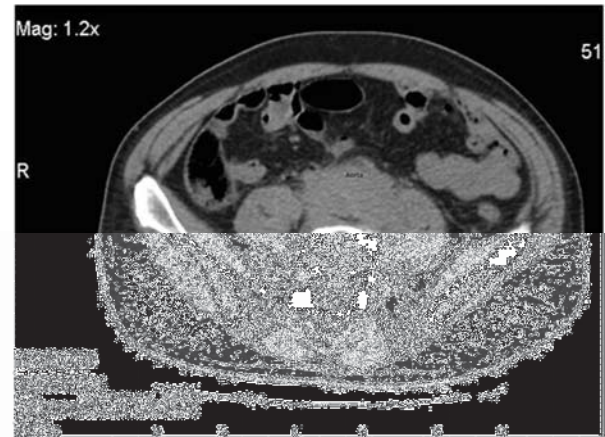
were regular, minimal dilatation of the pelvicalyceal structures was detected in abdominal CT. Irregularly shaped, and increased density of soft tissue around the abdominal aorta, starting from infrarenal level up to the proximal left common iliac artery and to the middle of right common iliac artery were detected (Figure 1).



**Fig. 1.** Non-contrast abdominal computerized tomography of abdomen showing minimal dilatation of pelvicalyceal structures.

The patient had no previous history of trauma and surgery. Blood pressure was normal and there were no signs of peripheral circulatory disorders. The patient was referred to the Vascular Surgery Department for compression of ureters secondary to the intra-abdominal hematoma and abdominal aorta aneurysm. Lesions on the CT were not accepted as an aneurysm and contrast-enhanced abdominal CT scan was recommended. Finally, CT supported the diagnosis of retroperitoneal fibrosis (Figure 2). Bilateral double J catheters were placed in both ureters.

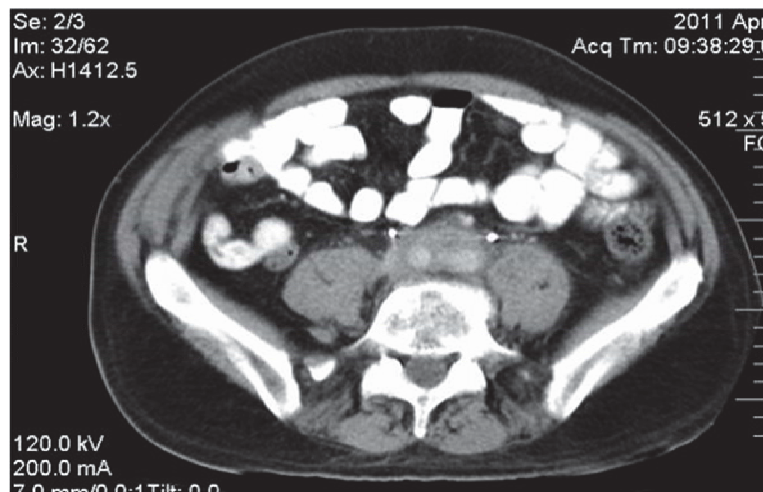
We started the therapy with 0,6/mg/kg of methylprednisolone. Urine output had progressively increased and urea and creatinine decreased to normal levels. Further investigations were planned for exclusion of the malignancy.



**Fig. 2.** Computerized tomography of abdomen excluding hematoma or aneurysm of aorta.

Chest X-ray was normal. Tumor markers were unremarkable. Fecal occult blood test was positive in two consecutive times. However, total colonoscopy revealed no pathologic findings.

Methylprednisolone of 60 mg/day (per oral) was initiated; urea and creatinine levels returned to normal in 10 days and the patient was discharged from the hospital. The dose tapering was planned during the control visits. At the end of the first month, control CT scan revealed a decrease in the lesion size to 15 mm. Three months later, the JJ catheters were removed (Figure 3). Steroid treatment stopped at the end of 6 months and monthly visits showed normal renal functions.



**Fig. 3.** Computerized tomography of abdomen (one month after treatment) showing retroperitoneal fibrosis recovery.

## Discussion

Waist, abdomen, lumbar pain, constitutional symptoms,

weight loss and fever may be present in idiopathic retroperitoneal fibrosis. In this case, there are complaints on abdominal pain. The most common complication of idio-

pathic RPF's is hydronephrosis and renal failure. 75% of patients with RPF are expected to lose renal function at the time of diagnosis. The treatment of idiopathic RPF includes steroid therapy, immunosuppressive agents and invasive urologic procedures (ureteral stent insertion, percutaneous nephrostomy). There is not a strict guideline for treatment. Fry, *et al.* suggested usage of corticosteroids alone [8]. Maillart, *et al.* recommended immunosuppressive treatment in addition to corticosteroids, which is superior to corticosteroid therapy alone (97%-70%) [9]. Steroids can be stopped if steroid drugs are used in combination with immunosuppressive agents (such as azathioprine, cyclophosphamide, methotrexate, cyclosporine, and micophenolate mofetil (MMF)) other than steroids alone [10,11]. Recently there has been an increasing evidence of the benefit of MMF, which made it the treatment of choice in these patients. However, a few studies have shown that azathioprine was highly effective in idiopathic retroperitoneal fibrosis [11]. Another advantage of azathioprine over MMF is its lower cost. There is no difference between side effects of these two drugs. Moroni, *et al.* showed the treatment response with azathioprine in their six patients' study [12]. In the present study, the patient responded to methylprednisolone therapy. In our opinion, steroids as the first-line therapy could be given alone in RPF patients. If needed combination with MMF or azathioprine may be suitable. Perhaps the only common sense in the treatment of RPF that comes with urinary obstruction, with no major metabolic disorders, is the steroid therapy alone as an initial therapy. However, in more severe cases, steroid treatment with intravenous pulse cyclophosphamide therapy may be the treatment of choice [13].

## Conclusions

Post-renal acute kidney injury needs careful evaluation. We wanted to emphasize that RPF has to be taken into consideration in the differential diagnosis of post-renal acute kidney injury although it is not very common. Appropriate treatment prevents progression of renal injury to further chronic kidney disease.

*Conflict of interest statement.* None declared.

## References

1. Parums DV, Chadwick DR, Mitchinson MJ. The localisation of immunoglobulin in chronic periaortitis. *Atherosclerosis* 1986; 61: 117-123.
2. Parums DV, Brown DL, Mitchinson MJ. Serum antibodies to oxidized low-density lipoprotein and ceroid in chronic periaortitis. *Arch Pathol Lab Med* 1990; 114: 383-387.
3. Meier P, Vogt B, Blanc E. Rethinking the triggering inflammatory processes of chronic periaortitis. *Nephron Exp Nephrol* 2007; 105: 17-23.
4. Morris DL, Taylor KE, Fernando MM, *et al.* Systemic Lupus Erythematosus Genetics Consortium, Criswell LA, Vyse TJ. Unraveling multiple MHC gene associations with systemic lupus erythematosus: model choice indicates a role for HLA alleles and non-HLA genes in Europeans. *Am J Hum Genet* 2012; 91: 778-793.
5. Kong YC, Brown NK, Flynn JC, *et al.* Efficacy of HLA-DRB1\*03:01 and H2E transgenic mouse strains to correlate pathogenic thyroglobulin epitopes for autoimmune thyroiditis. *J Autoimmun* 2011; 37: 63-70.
6. Weber P, Ambrosova P, Canov P, *et al.* GAD antibodies in T1D and LADA-relations to age, BMI, c-peptide, IA-2 and HLA-DRB1\*03 and DRB1\*04 alleles. *Adv Gerontol* 2011; 24: 312-318.
7. Fekih-Mrissa N, Klai S, Zaouali J, *et al.* Association of HLA-DR/DQ polymorphism with myasthenia gravis in Tunisian patients. *Clin Neurol Neurosurg* 2013; 115: 32-36.
8. Fry AC, Singh S, Gunda SS, *et al.* Successful use of steroids and ureteric stents in 24 patients with idiopathic retroperitoneal fibrosis: a retrospective study. *Nephron Clin Pract* 2008; 108: 213-220.
9. Maillart E, Laueriere L, Kassis S, *et al.* [Is there an evidence-based management of idiopathic retroperitoneal fibrosis?]. *Rev Med Interne* 2006; 2: 854-857.
10. van Bommel EF, van Spengler J, van der Hoven B, Kramer P. Retroperitoneal fibrosis: report of 12 cases and a review of the literature. *Neth J Med* 1991; 39: 338-345.
11. Kurzawska-Firlej D, Suchanek-Krawiec A, Piernik T, Oko A. Outcome of immunosuppressive treatment of a patient with renal failure due to retroperitoneal fibrosis. *Pol Arch Med Wewn* 2004; 112: 845-848.
12. Moroni G, Dore R, Collini P. Idiopathic retroperitoneal fibrosis. *J Nephrol* 2005; 18: 794-808.
13. WarnatzK, Keskin AG, Uhl M, *et al.* Immunosuppressive treatment of chronic periaortitis: a retrospective study of 20 patients with chronic periaortitis and a review of the literature. *Ann Rheum Dis* 2005; 64: 828-833.