İstanbul Med J 2017; 18: 37-9
DOI: 10.5152/imj.2016.01643

Idiopathic Retroperitoneal Fibrosis Presenting with Hypertension and Acute Renal Failure

Hipertansiyon ve Akut Böbrek Yetmezliği ile Prezente Olan İdiopatik Retroperitoneal Fibrozis

Engin Onan[1], Saime Paydaş[1], Merve Erkoç[2], Tuba Korkmaz[2], Hasan Bilen Onan[3], Mustafa Balal[1]

Abstract / Öz

Abdominal aortic aneurysm can rarely cause retroperitoneal fibrosis secondary to the leakage of inflammatory cells from aortic plaques to the retroperitoneal area. In this study, we present a case with severe hypertension and acute renal failure secondary to retroperitoneal fibrosis related to abdominal aortic aneurysm.

Keywords: Retroperitoneal fibrosis, acute kidney injury, hypertension, abdominal aortic aneurysm

Abdominal aort anevrizması, aortik plaklardaki inflamatuar hücrelerin retroperitoneal bölgeye kaçağına bağlı olarak nadir olarak retroperitoneal fibrozise neden olabilir. Biz de abdominal aort anevrizması ve buna sekonder retroperitoneal fibrozisi hipertansiyon ve akut böbrek yetmezliği ile prezente olan hastayı sunmayı amaçladık.

Anahtar Kelimeler: Retroperitoneal fibrozis, akut böbrek hasarı, hipertansiyon, aort anevrizması abdominal

Introduction

Retroperitoneal fibrosis is frequently idiopathic, and autoimmune diseases such as Hashimoto thyroiditis, surgeries, drugs, and malignant diseases can cause idiopathic retroperitoneal fibrosis (IRF) (1, 2). Abdominal aortic aneurysm can trigger IRF with the leakage of inflammatory cells from aortic plaques to the retroperitoneal area (3). Retroperitoneal fibrosis usually occurs with pain in the back, flank, or abdomen; vomiting; leg edema; or urinary obstruction induced by fibrosis in the retroperitoneum (1, 4).

Case Report

A 49-year-old-man was admitted to the Nephrology Clinic with complaints of hypertension and abdominal pain. He was previously admitted thrice to other emergency rooms with unrelenting flank pain and hypertension before arriving at our clinic. Parenteral analgesics and antihypertensives were administered. Nausea, weight loss, and anorexia were added to the complaints in the previous 2 months.

On examination, his blood pressure was 180/110 mmHg, and remaining physical examination findings were normal, as were chest X-ray and n electrocardiogram. He had a history of glaucoma and 30 pocket-year cigarette smoking and was administered 2 mg/day doxazosin for hypertension. Contrast agent was not administered. Laboratory test results are summarized in Table 1. There were 2-3 erythrocytes and 4-5 leucocytes observed on urinary sediment analysis. The urinary sodium level was 19 mmol/L. Abdominal ultrasonography revealed normal-sized kidney and parenchyma, grade 1 parenchymal echogenicity, and anteroposterior pelvis diameter consistent with hydronephrosis: 2.4 cm at the right kidney and 2.2 cm at the left kidney. Renal Doppler ultrasonography was normal. Parenteral fluid replacement and urethral catheter were administered, and his blood urine nitrogen (BUN)/serum creatinine levels were 37/2.43 mg/dL on fifth day of hospitalization. Because renal dysfunction and hydronephrosis did not improve, abdominal tomography was performed, which revealed bilateral hydronephrosis, aortitis, and aneurysmatic dilatation of the aorta (Figure 1a, b). The patient refused double J catheter replacement. Serum complement levels were normal, and anti-nuclear antibody, anti-double-stranded DNA, anti-neutrophil cytoplasmic antibody, and antibodies against hepatitis B surface anti-hepatitis B core, anti-HIV, anti-HCV, and hepatitis B surface antigen were negative. Anti-glomerular basal membrane (anti-GBM) antibody was positive. Serum immunoglobulin (Ig) G4 levels were normal. The cause of hydronephrosis was clinically accepted to be IRF, and oral 32 mg/day methylprednisolone was initiated. The diagnosis was not histopathologically proven because the patient refused to undergo a surgery. On the fifth day of methylprednisolone medication and 18th day of hospitalization, BUN/serum creatinine levels improved to 32/1.65 mg/dL, and the patient was discharged. Six months

[1]Division of Nephrology, Department of Internal Medicine, Çukurova University School of Medicine, Adana, Türkiye
[2]Department of Internal Medicine, Çukurova University School of Medicine, Adana, Türkiye
[3]Department of Radiology, Çukurova University School of Medicine, Adana, Türkiye

Address for Correspondence
Yazışma Adresi:
Engin Onan
E-mail: onanmd@gmail.com

Received/Geliş Tarihi:
05.05.2016

Accepted/Kabul Tarihi:
13.06.2016

© Copyright 2017 by Available online at www.istanbulmedicaljournal.org

© Telif Hakkı 2017 Makale metnine www.istanbultipdergisi.org web sayfasından ulaşılabilir.

Table 1. Laboratory data during hospitalization

Parameters	Day 1	Day 5	Day 10	Day 18	Month 6
Hematocrit, % (39.5-50.3)	35.7	38	34.6	36.7	41.2
Hemoglobin, g/dL (16-17.2)	10.9	12.1	11	11.6	14
White blood cells, /mm^3 (4.5-10.3)	7.00	6.30	6.50	8.20	9.80
Platelets, x10^3/µL (156-373)	225	219	193	210	183
BUN, mg/dL (8-20)	40	37	36	32	14
Creatinine, mg/dL (0.4-1)	2.94	2.43	1.85	1.65	0.93
Na^+, mmol/L (136-144)	134	134	136	137	139
K^+, mmol/L (3.6-5.1)	4.8	4.6	4.5	4.2	3.9
Ca^{+2}, mg/dL (8.9-10.3)	9.2	10.1	9.7	9.2	9.8
P, mg/dL (2.4-4.7)	4.31	4.0	3.7	4.1	3.6
Uric acid, mg/dL (4.8-8.7)	6.4	-	-	-	5.5
Total protein, g/dL (6.1-7.9)	6.4	7.6	6.9	7	6.4
Albumin, mg/dL (3.5-4.8)	3.6	4.3	3.9	4	4.0
Anti-GBM	++				Negative

BUN: blood urine nitrogen; Na^+: sodium; K^+: potassium; Ca^{2+}: calcium; P: phosphorus; anti-GBM: anti-glomerular basal membrane

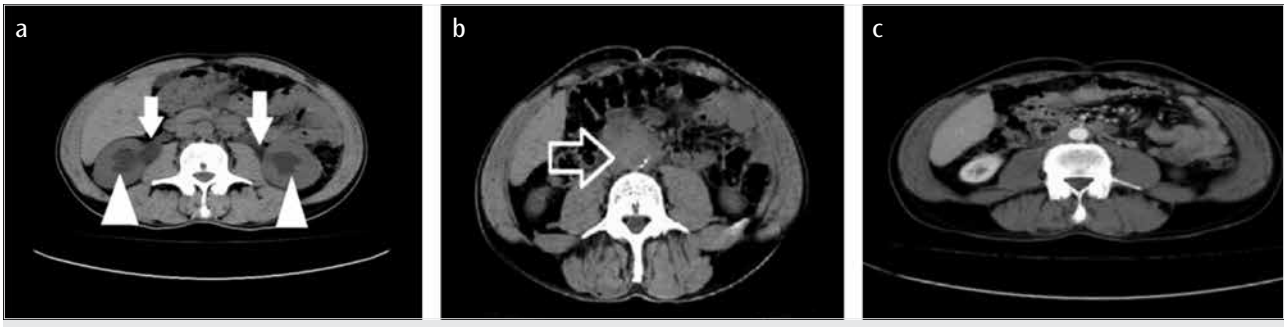

Figure 1. a-c. Bilateral hydronephrosis (white arrowheads) and extended ureters (white arrows) (a). Aortic calcification and aortitis (white arrow) (b). Abdominal tomography at 6 months after discharge showed no hydronephrosis and aortitis (c)

after discharge, he was found asymptomatic. His blood pressure was 110/70 mmHg, and other physical examinations, including pulse rates, were found to be normal. BUN/serum creatinine levels were normal. Control abdominal computed tomography and serum IgG4 antibody titer were normal, and anti-GBM antibody was negative (Figure 1c). The patient continues to return for routine follow-ups. Informed consent was obtained from the patient.

Discussion

Retroperitoneal fibrosis is a rare condition, with an incidence of 1-2/200,000 individuals (1). There are no specific laboratory tests for IRF. C-reactive protein levels may be high or normal and is often used as an indicator of response to therapy (2). In our patient, there was no diagnostic test result. Erythrocyte sedimentation rate was slightly increased. Only anti-GBM antibody was positive. Radiological imaging is the gold standard for diagnosing abdominal aortic aneurysm and IRF. Ultrasonography may be useful for detecting ureteric obstructions but has a limited value for diagnosis. Computed tomography and magnetic resonance imaging with contrast agents are ideal imaging techniques and are effective in demonstrating the involvement of disease and fibrosis. In our patient, we first determined hydronephrosis using ultrasonography followed by computed tomography. We could not histopathologically confirm the diagnosis. Although there was no presence of hypocomplementemia, we could not rule out IgG4-related diseases.

There was no eosinophilia or eosinophiluria related to acute interstitial nephritis. Retroperitoneal involvement for IgG4-related disease is 20% and is frequently observed as a mass in the retroperitoneal area, lymphadenopathy, or inflammation of vessels, e.g., periaortitis/periarteritis. IgG4-related diseases are present in 55%-57% of patients with IRF. Retroperitoneal involvement in patients with IgG4-related IRF usually begins in the periaortic or periiliac regions and may progress to ureteral involvement. Increasing evidences have shown that noninfectious aortitis is related to IgG4, and this process may also affect iliac/mesenteric arteries. Pathologic findings of tissue samples, obtained by re-evaluating archived and clinical data, were correlated with the IgG4-related disease. Diagnostic criteria include (i) increased levels of IgG4-positive plasma cells as >50 magnification ×400 for lungs, (ii) IgG4/IgG ratio of >0.40 or >0.30 with in situ hybridization, and (iii) characteristic histological findings such as lymphocytic infiltrate, spindle fibrosis, and obliterative phlebitis in the tissue. Interventional procedures were refused by our patient; hence, we could not explore the potential association among aortitis, retroperitoneal fibrosis, hydronephrosis, and IgG4-related disease.

Anti-GBM antibody was positive before treatment, but after six months of treatment, anti-GBM antibody was negative, with the regression of hydronephrosis. Takeuchi et al. reported that anti-GBM antibody was positive with hydronephrosis, and with the regression of hydronephrosis, it was negative. They also observed the development of crescentic glomerulonephritis if the positivity for the antibody persisted (5). These findings were similar to those of our case.

IRF-related ureteric obstruction can be treated with steroids, invasive stent, or open ureterolysis. Prednisolone inhibits fibrosis by suppressing inflammation. Our patient had nausea, vomiting, and bilateral hydronephrosis. Despite intravenous fluids and urethral catheter, there was minimal decrease in BUN/serum creatinine levels. Using radiological examinations, abdominal aneurysm and retroperitoneal fibrosis were detected; however, because our patient refused interventional procedures, 32 mg/day oral methylprednisolone was administered. In the following days, BUN\serum creatinine levels rapidly improved. We also radiologically detected an improvement in the process of fibrosis, and the patient's kidneys were functioning normally 6 months after diagnosis.

The recommended treatment duration is minimum 1 year for IRF. Treatment options include methotrexate, azathioprine, and cyclophosphamide. Immunosuppressive treatments can be used with ureterolysis or invasive stents. Open ureterolysis is also effective, but laparoscopic ureterolysis is a better option for the geriatric population because of its minimal complication rates (6, 7).

Conclusion

Idiopathic retroperitoneal fibrosis is a rare cause of post-renal acute kidney injury. With or without abdominal aortic aneurysm, IRF may cause hydronephrosis and acute kidney injury. Along with abdominal pain, severe hypertension, acute renal dysfunction, hydronephrosis, and abdominal aortic aneurysm, the possibility of retroperitoneal fibrosis should also be considered.

Informed Consent: Informed consent was obtained from patients who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - E.O.; Design - E.O.; Supervision - S.P.; Materials - T.K.; Data Collection and/or Processing - M.E.; Analysis and/or Interpretation - H.B.O.; Literature Review - S.P.; Writing - E.O., S.P.; Critical Review - S.P.; Other - M.B.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

Hasta Onamı: Hasta onamı bu çalışmaya katılan hastalardan alınmıştır.

Hakem Değerlendirmesi: Dış bağımsız.

Yazar Katkıları: Fikir - E.O.; Tasarım - E.O.; Denetleme - S.P.; Malzemeler - T.K.; Veri Toplanması ve/veya İşlemesi - M.E.; Analiz ve/veya Yorum - H.B.O.; Literatür Taraması - S.P.; Yazıyı Yazan - E.O., S.P.; Eleştirel İnceleme - S.P.; Diğer - M.B.

Çıkar Çatışması: Yazarlar çıkar çatışması bildirmemişlerdir.

Finansal Destek: Yazarlar bu çalışma için finansal destek almadığını belirtmiştir.

References

1. Burkhardt SS, Fehr A, Brandt AS, Roth S. Retroperitoneal fibrosis. Aktuelle Urol 2007; 38: 221-31. [CrossRef]
2. Warnatz K, Keskin AG, Uhl M, Scholz C, Katzenwadel A, Vaith P, 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: 828-33. [CrossRef]
3. Rasmussen TE, Hallett JW. Jr Inflammatory aortic aneurysms: a clinical review with new perspectives in pathogenesis. Ann Surg 1997; 225: 155-64. [CrossRef]
4. Swartz RD. Idiopathic retroperitoneal fibrosis: a review of the pathogenesis and approaches to treatment. Am J Kidney Dis 2009; 54: 546-53. [CrossRef]
5. Takeuchi Y, Takeuchi E, Kamata K. A possible clue for the production of anti-glomerular basement membrane antibody associated with ureteral obstruction and hydronephrosis. Case Rep Nephrol Dial 2015; 5: 87-95. [CrossRef]
6. Stifelman MD, Shah O, Mufarrij P, Lipkin M. Minimally invasive management of retroperitoneal fibrosis. Urology 2008; 71: 201-4. [CrossRef]
7. Tobias-Machado M, Correa WF, Korkes F, Juliano C, Muller S, Pompeo AC. Transmesocolic ureteral intraperitonealization: a new approach for laparoscopic treatment of retroperitoneal fibrosis. J Laparoendosc Adv Surg Tech A 2011; 21: 341-4. [CrossRef]