BANTAO Journal 2008; 6(2): 38-40

BJ
BANTAO Journal

Case report

A case of retroperitoneal fibrosis presenting with uremic encephalopathy

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Abstract

Retroperitoneal fibrosis (RPF) is characterized by the development of dense fibrous tissue that may cause obstruction of retroperitoneal organs, especially the ureters. It is usually primary; but may be secondary to malignancies, collagen vascular diseases, some drugs, radiation and trauma. Clinical presentation is variable. Here we discuss a case presenting with uremic encephalopathy.

A sixty-one years old male was brought to the emergency clinic due to nausea, vomiting, fatigue, decreased urine volume and insensible speech. He was confused with distorted orientation for time and place. Skin was pale and there was flapping tremor. Other physical findings were unremarkable. Uremia, uremic encephalopathy and metabolic acidosis with increased anion gap lead to urgent hemodialysis. Ultrasonography showed bilateral grade 2-3 hydronephrosis without stones and other intraluminal obstructing lesion. Both abdominal computerized tomography and magnetic resonance imaging revealed a retroperitoneal soft tissue mass surrounding the aorta and inferior vena cava. Further investigations provided no further information about the etiology. He had bilateral laparoscopic ureterolysis operation and tissue samples were taken for pathological examination which was consistent with RPF with no findings of malignancy. He is under follow-up with no symptoms and with normal renal functions.

RPF may have highly variable etiology and clinical presentation. Our case represented with uremic encephalopathy. The diagnosis of RPF should lead to a thorough evaluation for a possible underlying disease. Primary cases relapse frequently, so these patients must be followed regularly.

Post renal causes of renal failure especially RPF must always be kept in mind when there is no obvious intraluminal obstructive lesion and diagnostic phase must be as short as possible to prevent loss of renal parenchyma.

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Key words: encephalopathy; fibrosis; primary; retroperitoneal

Introduction

The French urologist Albarran first described retroperitoneal fibrosis (RPF) in 1905, but with Ormond's publication in 1948, the disease became an established clinical entity [1]. Retroperitoneal fibrosis is characterized by the development of dense fibrous tissue all through the retroperitoneum especially in front of the fourth and fifth lumbar vertebrae, surrounding abdominal aorta and iliac vessels. This fibrous tissue may cause obstruction of retroperitoneal organs, especially the ureters [2]. Annual incidence is 1/200000 with peak at 40-60 years of age [3]. It is two times more commonly seen in males. 70% of cases are idiopathic. 8% of cases were associated with malignancies (lymphoma, sarcoma, carcinoma of breast, stomach, lung, colon, bladder, prostate and cervix). Other possible etiologic factors are retroperitoneal inflammation, trauma, some drugs (methysergide, methyldopa, and beta blockers), collagen vascular diseases and radiation [2].

Clinical presentation varies from constitutional symptoms like fever, fatigue and weight loss to symptoms due to venous obstruction, or varying degrees of renal failure, abdominal pain, nausea and vomiting due to ureteric obstruction. Absolute diagnosis depends on pathological examination. Intravenous urography, computerized tomography (CT) and magnetic resonance imaging (MRI) helps to determine the extent of the disease and to follow-up. The primary aim of any treatment is to preserve the renal functions. Stent application to the ureters, surgical ureterolysis, corticosteroids, cytotoxic agents and tamoxifen are main modalities of treatment [4,5]. There are studies reporting 100% five year survival among idiopathic cases with these treatment strategies [6].

Case report

A sixty-one year old male presented with with fatigue, nausea, vomiting, decreased urine volume and confusion. He was without any symptoms until the last two weeks during which progressive fatigue, loss of appetite, nausea

and vomiting developed and followed by decreased urine volume, insensible speech and tremor of the hands for the last few days. His past medical and family histories were unremarkable. He had smoked twenty pack-years.

He was confused when seen in the emergency room with distorted orientation for time and place. Skin was pale and there was flapping tremor. Other physical findings were unremarkable. Abnormal laboratory findings were as follows: blood urea: 74.97 mmol/L, creatinine: 845.72 µmol/L, sodium: 138 mmol/L, potasium: 6.51 mmol/L, calcium: 2.27 mmol/L, phosphorus: 1.94 mmol/L, intact parathyroid hormone: 130 ng/L, hemoglobin: 113 gr/L, hematocrite: 0.31, MCV: 77 fl, sedimentation rate: 55 mm/hour and C-reactive protein: 454000 µg/L. Urine sediment had 10-15 erythrocytes and 4-5 leukocytes per high power field; creatinine clearance was 8.9 ml/minute and he had 586 mg/day proteinuria.

Arterial blood gas analysis revealed metabolic acidosis with respiratory compensation (pH: 7.25, HCO_3: 13 mmol/l, pCO_2: 26.4 mmHg, pO_2: 106 mmHg). Hemodialysis therapy was initiated due to uremic encephalopathy and immediately afterwards laboratory and radiological examinations for evaluating the etiology of renal failure were commenced.

Bilateral grade 2-3 hydronephrosis was reported after the urinary system ultrasonography; which also demonstrated normal echogenity and parenchymal thicknesses of the kidneys. The abdominal CT was showed an enlarged liver (180 mm) and spleen (144 mm) and bilateral grade 3 hydronephrosis. Ureters could be followed as dilated to the aortic bifurcation level. A retroperitoneal soft tissue mass surrounding the aorta and inferior vena cava at the aorto-iliac bifurcation, and descending caudally as long as the main vasculature was seen during abdominal MRI (Figure 1). Ureters were ending within this soft tissue density.

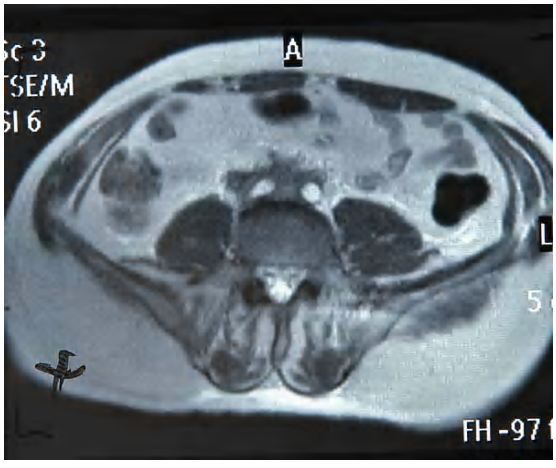

Fig. 1. Abdominal MRI showing a retroperitoneal mass

With these radiological findings he was diagnosed as RPF and bilateral ureteric catheters were placed after three sessions of hemodialysis. The urine volume increased progressively after the procedure leading to the normalization of blood urea nitrogen and creatinine levels. In the mean time upper and lower gastrointestinal series revealed no pathology. Tumor markers were negative. Autoantibodies were studied and anti-nuclear antibody, anti-double stranded DNA antibody, and antineutrophil cytoplasmic antibody were found to be negative. For definitive diagnosis of RPF and treatment, he had bilateral laparoscopic ureterolysis during which tissue samples were taken for pathological examination. Pathology report was consistent with RPF with no findings of malignancy. Currently he is under follow-up with no symptoms and with normal renal functions.

Discussion

Retroperitoneal fibrosis is most common in people aged 40 - 60, and men are twice as likely to develop them as women [3]. It may have highly variable clinical presentation ranging from constitutional symptoms such as severe pain in the lower back, abdominal, and flank areas, swelling in one or both legs to different degrees of renal failure (three quarters of patients) [6]. Our male case had presented with uremic encephalopathy without any prior constitutional symptoms.

Post-renal causes of renal failure must always be kept in mind and diagnostic phase must be as short as possible to prevent loss of renal parenchyma. Since bilateral grade 2-3 hydronephrosis was reported after the urinary system ultrasonography in our case, a post-renal etiology was sought. Although rare, RPF is a treatable cause and must be remembered when there is no obvious reason for obstruction.

The diagnosis of RPF should lead to a thorough evaluation for a possible malignancy, collagen vascular disease, autoimmune disease; and history of trauma and drug use should be asked insistently [4]. Our patient had no history of trauma or any drug use. Moreover, he did not have any significant laboratory result regarding the etiology of RPF; so he was regarded as primary.

Primary RPF cases relapse frequently, so these patients must be followed regularly for relapse and for an underlying disease which may be detected years after the diagnosis of RPF.

Conclusions

We wanted to share this case to remind approach to the etiology of renal failure, especially of post-renal causes. RPF must be kept in mind in cases without obvious reason for obstruction.

Conflict of interest statement. None declared.

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