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Unilateral perirenal fibrosis without aorta involvement

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Abstract. – Retroperitoneal fibrosis (RPF) located unilateral perirenal without aorta involvement is very rare. We report a case of unilateral perirenal fibrosis which was misdiagnosed as malignancy even after biopsy. RPF should be in mind in dealing with perirenal mass.

Key Words:
Perirenal mass, Perirenal fibrosis, Unilateral, Retroperitoneal fibrosis.

Introduction

Retroperitoneal fibrosis (RPF) is not a common disease[1]. It is characterized by inflammatory fabric tissue enveloping the abdominal aorta, iliac artery, and even nearby organs, such as vena cava, ureter and so on. It is very rare for the RPF located unilateral perirenal[2]. We report a case of unilateral perirenal fibrosis without aorta involvement, which was mistaken for a malignancy and followed with a laparotomy and extended radical nephrectomy.

Case Report

A 45-year-old previously healthy man presented with dull pain in the right abdominal and anorexia 10 months before admitted to our department. Computed tomography (CT) in the local hospital indicated a hematoma of the right kidney. After six months observation and rest, a repeated CT (Figure 1) showed a right renal mass larger than the previous named hematoma. The mass invaded into the renal cortex, involving renal vessels and inferior vena cava. It can be strengthened mildly to moderately. A malignancy was suspected followed with ultrasound guided biopsy. Pathology of biopsy specimen showed hemorrhage. He came to another hospital, in where a magnetic resonance imaging (MRI) showed a perirenal mass with low intense signal in T1/T2 images, surrounding and compressing the inferior vena cava. It can be enhanced slowly. Suspected diagnosis included lymphoma, metastatic tumor, and inflammatory psedotumor (Figure 2). ^{18}F-FDG positron emission tomography/computed tomography (PET/CT) showed an irregular perirenal soft tissue mass situated mainly in the hilum and upper pole of the right kidney with a high FDG intaking (Figure 3). A malignancy was suspected. He received a repeated biopsy guided by ultrasound. Pathology of the specimen showed that the mass was mainly comprised of spindle cells with plenty of inflammatory cells infiltrating into the tumor. Immune staining shows Viminten ++, SMA+++, CD34+/-, S-100-, CD117-, CD68+, LCA+, Ki67-. Pathology consistent with inflammatory myofibroblastomatic tumor. Analplasia nucleus can be seen in some tumor cells. He came to our hospital for operation. On physical examination, he was confirmed with hypertension. His serum creatinine was elevated to 126.10 μmol/L (normal reference: 53.00-115.00 μmol/L) preoperation. He received laparotomy and extended radical nephrectomy. In the operation, the mass was grey white, hard, and firmly adhere to the vena cava. With hard and careful work, the operator removed the mass totally including the right kidney/adrenal in it and a part of vena cava wall (Specimen showed in Figure 4). The vena cava was closed with vascular suture. The patient recovered unevenfully after operation. The pathology showed that the right kidney and adrenal gland were enveloped by fabric tissue with collagen formation. There were plenty of lymphocytes, neutrophils, and a small amount of eosinophils infiltrating into the mass. Fibrous tissue penetrate into renal cortex. The right adrenal was intact. Vascular endothelial erosion can be seen in the affected inferior vena cava wall which infiltrated with a plenty of neutrophils and lymphocytes; Reactive hyperplasia showed in the two retrieved lymph nodes. All consistent with the diagnosis of idiopathic RPF with renal cortex infiltration (Figure 5). His blood pressure returned to normal one month postoperatively with stable level of serum creatinine.

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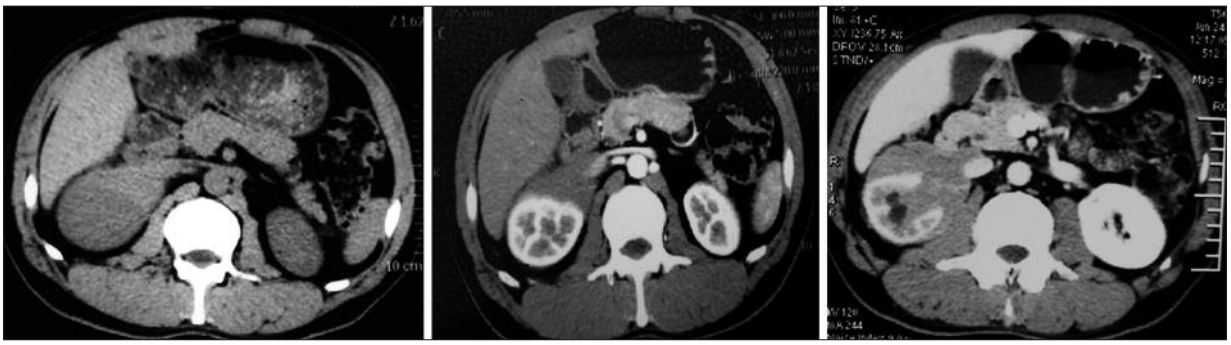

Figure 1. CT shows perirenal (right) mass isodense with muscle ***(A)*** invaded into renal sinus ***(C)***, partly involved the vena cava ***(B-C)***. The mass can be enhanced moderately ***(C)***. The aorta is normal ***(B-C)***.

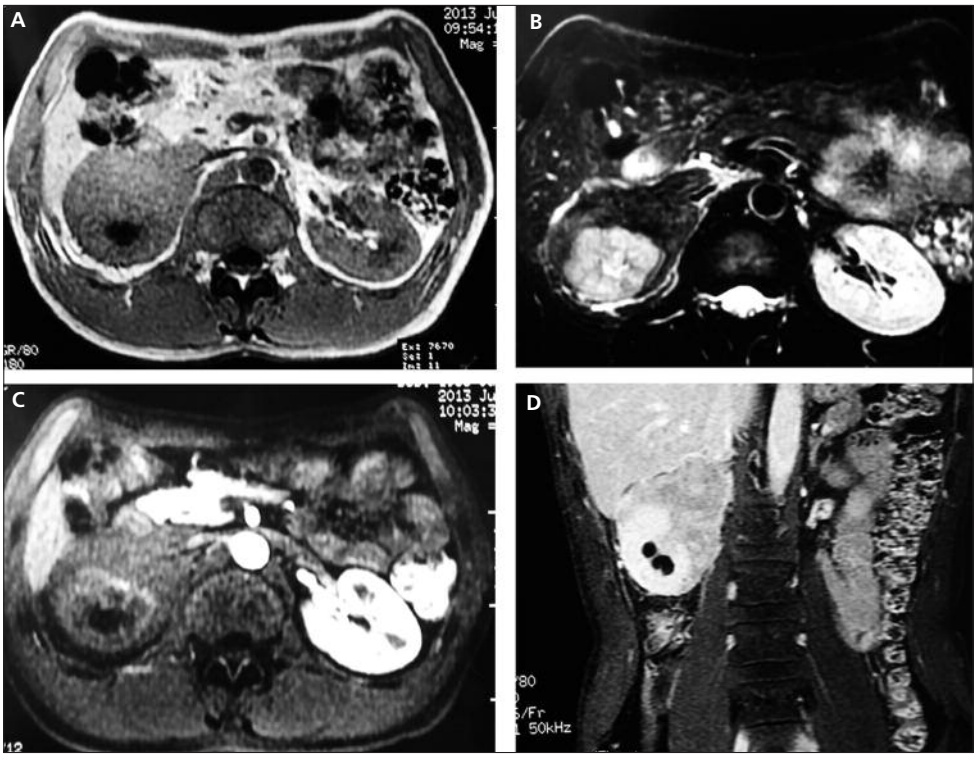

Figure 2. MRI shows the perirenal mass (*right*) with low intense signal in T1W/T2W images ***(A, B)***, mainly situated in the hilum and upper pole of right kidney ***(D)***. The mass involved part of the wall of vena cava without intruded into its luminal ***(A, B)***, The right kidney with hydronephrosis was enhanced poorly compared with the left one ***(A, C)***.

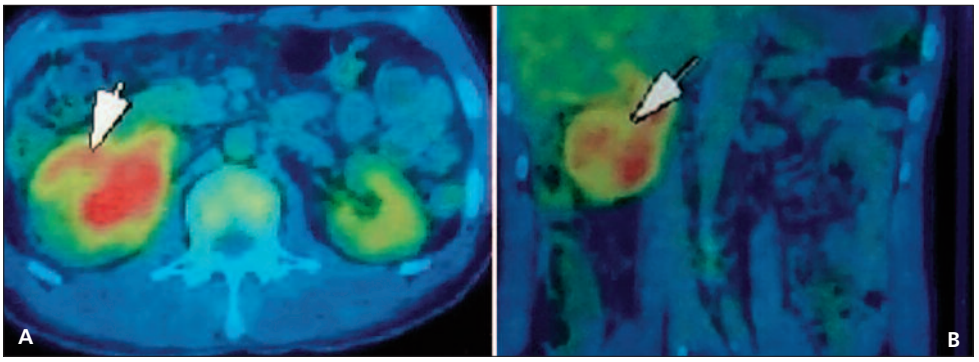

Figure 3. ***A, B,*** PET/CT shows the perirenal mass (*right*) with high FDG intaking, adhere to the lateral and anterior wall of vena cava.

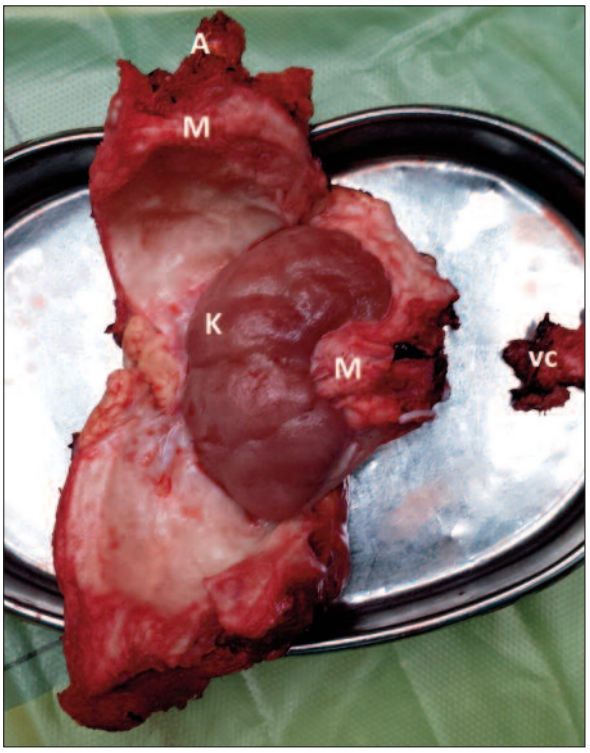

Figure 4. The specimen shows the mass (M) enveloped the whole right kidney (K) and right adrenal gland (A). The mass invaded into the renal sinus. A piece of wall of vena cava was ablated with the undetachable fibrotic tissue (VC).

Discussion

RPF is a rare condition. The most common presenting symptoms were nonspecific dull pain in back, abdominal or flank. Weight loss and nausea is also common[3]. Physical examination is always normal. The laboratory test may show system inflammation, such as elevated level of C-reactive protein, erythrocyte sedimentation rate and leukocytosis. The diagnosis of RPF relies primarily on imaging studies. Nearly 90% of RPF presented with periaortic mass and/or perireteral mass localized to the infrarenal large arteries[3]. RPF is diversity in images for its disease stages. On CT scans, the mass, isodense with muscle, can be variably enhanced depending on the stage of the fibrotic process[4]. In the early stage, with more vascular the mass enhances to a greater degree[5]. On MRI, the T1-weighted image shows hypointense.The T2-weighted images are high intense in early stage and low intense in late stage. Whereas areas affected by active inflammation demonstrate high T2 signal intensity and early contrast enhancement, areas of fibrosis show low T2 signal intensity and delayed contrast enhancement[6]. Perirenal involvement is not common. Perirenal fibrosis that occurs in association with periaorta retroperitoneal fibrosis or as part of multifocal fibrosclerosis is not difficult to detect at imaging[7,8]. However, the imaging features of isolated perirenal fibrosis are nonspecific and a biopsy may be required to achieve a definitive diagnosis[2]. Biopsy guided by CT or ultrasound is the first line procedure to obtain the specimen[2,6,9,10]. Some needed laparotomy[11]. For extremely rare character of localized unilateral fibrosis without aorta involvement[2], a nephrectomy maybe adopted for the malignancy possibility[12]. In our case, the serum creatinine is stable after the right nephrectomy, and the blood pressure returns to normal. His condition is thought as a renal and/or renal vascular hypertension. Postoperative medicine therapy and long term follow-up should be advised to the patient for the recurrence possibility.

In the early stage of perirenal fibrosis, the patient may be asymptomatic with normal renal function[9]. But the disease may progress with hydronephrosis, renal atrophy, renal failure, and compression effect of the affected organs[3,13]. The medicine treating is effective in the early stage of disease, and hardly irreversible in late stage. So, a closely surveillance and medicine therapy mainly including corticosteroid and/or tamoxifen should be taken for the disease especially in the early stage. Surgical intervention is considered for steroid-resistant cases or to relieve ureteral obstruction.

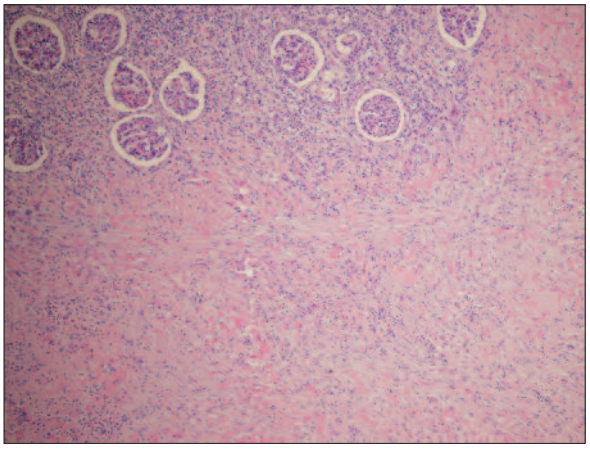

Figure 5. Pathology shows the fibrosis tissue infiltrated into renal cortex (hematoxylin-eosin stain, original magnification ×100).

We reported this very rare case in order to emphasis the possibility of perirenal RPF other than in the ordinary places of retroperitoneal. The needle biopsy may miss the typical part of RPF. New innovative work is urged regarding diagnostics and therapy of RPF[14].

Conflict of Interest

The Authors declare that there are no conflicts of interest.

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