

# Retroperitoneal Fibrosis in a Chinese Woman with Systemic Lupus Erythematosus: A Case Report

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## Abstract

Systemic lupus erythematosus with retroperitoneal fibrosis, a rare disease with the mechanism obscure, is considered an autoimmune disorder at present. Computed tomography and Magnetic resonance imaging are the two tests of choice in diagnosis and follow-up. A combination of corticosteroids and immunosuppressive drugs is usually applied for therapy. Laparoscopic ureterolysis, or robotic ureterolysis, if available, is the technique of choice to relieve obstruction.

**Keywords:** Systemic lupus erythematosus; Retroperitoneal fibrosis; Computed tomography

## Introduction

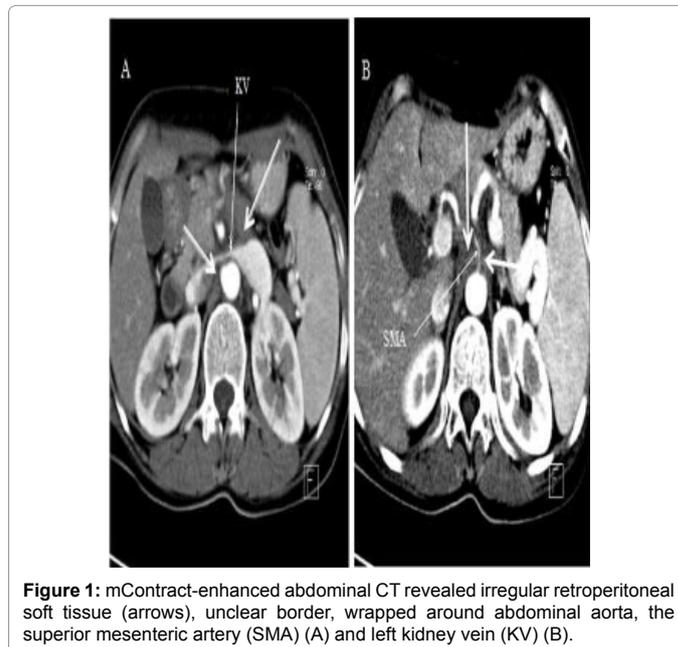
Retroperitoneal fibrosis (RPF) is a rare disease characterized by the presence of a fibro-inflammatory tissue, which usually surrounds the major vessels and organs located within the retroperitoneum. Systemic lupus erythematosus concurrent with retroperitoneal fibrosis is an exceptionally rare disorder with only 7 cases publicly reported so far. The purpose of this report is to present a case of a 52-year-old Chinese female suffering from retroperitoneal fibrosis with history of systemic lupus erythematosus.

## Case Report

A 52-year-old woman, with a history of SLE diagnosed in 2002, was admitted into our hospital for intermittent abdominal pain within the last 10 months. The pain was continuous, blunt in character, and radiated to the loins, nates, and thighs bilaterally, worsening during walking. Her past medical and family history was unremarkable, with the exception of a treatment of corticosteroids for SLE. Physical examination at the time of admission revealed the female patient to be hypertensive with a blood pressure of 158/95 mm Hg in both upper and lower extremities. Examination of the cardiovascular and respiratory systems was unremarkable with the pulse 82 beats per minute, the respiratory rate 20 breaths per minute, and the temperature 37.3°C. The abdomen was soft with epigastric tenderness, but without rebound on palpation. Liver and spleen was not palpated. The peristaltic sound was normal. There was no pain in kidney area on percussion. She had edema in lower extremities. Her initial laboratory test results showed a red blood cell of  $3.32 \times 10^{12}/L$ , hemoglobin 98.5 g/L, white blood cell  $80 \times 10^9/L$ , blood platelet  $80 \times 10^9/L$ , erythrocyte sedimentation rate 123 mm/h, Activated partial thromboplastin time 37.9 seconds, D-dimer 1062 ng/ml. The liver-function and kidney-function tests were normal. Serological tests for hepatitis virus antibody, anti-cytomegalo-virus antibodies, anti-streptolysin O antibodies, and anti-HIV antibodies were negative. The results of immunological tests indicated that ANA was positive with the titer of 1:1000 and anti-nRNP/Sm antibody, anti-SSA antibody, anti-Ro-52, anti-nucleosome antibody, anti-ribosomal P protein, anti-neutrophil cytoplasmic antibody, myeloperoxidase antibody, anti-cardiolipin IgM were positive. Complement and immunoglobulin assay revealed an decrease in C3 at 0.69 g/l (normal range, 0.9-1.8), an decrease in C4 at 0.086 g/l (normal range, 0.1-0.4), an increase in IgG at 17.4 g/l (normal range, 7.0-16.0), an increase in IgM at 2.74 g/l (normal range, 0.4-2.3) and an increase in IgE at 409 IU/ml (normal range, 0-100). A urinalysis presented the following results: ++protein, +++blood, no casts, and microproteinin 24 h urine 800 mg/day (0-80).

Her gastrointestinal endoscopy suggested chronic erosive gastritis. Contract-enhanced abdominal CT revealed a thickening of the wall of the abdominal aorta, superior mesenteric artery and kidney vein and an increased density of the surrounding fat tissues (Figure 1).

Clinical diagnosis included systemic lupus erythematosus and hemorrhagic erosive gastritis. Diagnosis of the idiopathic RPF was suspected, and primary medical therapy of idiopathic RPF is glucocorticoid 80 mg/d intravenous drip, with the treatment of the original disease with the leflunomide. Abdominal pain was relieved after treatment and the patient discharged to continue drug therapy at home. Patient was followed in clinic for about one year and repeat CT,



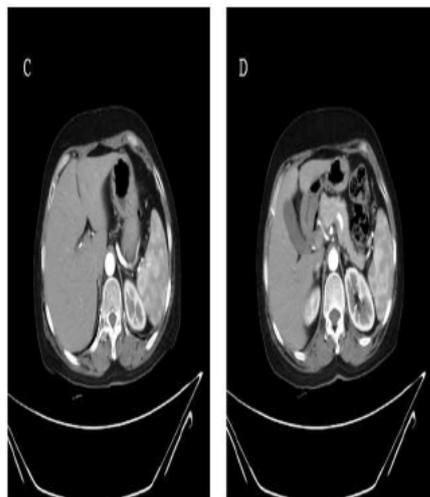
**Figure 1:** mContract-enhanced abdominal CT revealed irregular retroperitoneal soft tissue (arrows), unclear border, wrapped around abdominal aorta, the superior mesenteric artery (SMA) (A) and left kidney vein (KV) (B).

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Received March 14, 2016; Accepted May 17, 2016; Published May 26, 2016

**Citation:** Hu H, Shui H (2016) Retroperitoneal Fibrosis in a Chinese Woman with Systemic Lupus Erythematosus: A Case Report. J Clin Case Rep 6: 787. doi:10.4172/2165-7920.1000787

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**Figure 2:** Enhanced-CT performed 10 months after therapy revealed retroperitoneal soft tissue in regression and continued to improve.

which was performed 10 months after starting the therapy, showed RPF to be in regression and continued to improve (Figure 2).

## Discussion

RPF, first described in 1905 by French urologist Albarran [1], and later formally described by Ormond [2] in 1948, was initially recognized as an the proliferation of retroperitoneal fibrous tissue which could cause ureteral obstruction necessitating surgery. The etiology and pathogenesis of retroperitoneal fibrosis have not yet been studied thoroughly, which is characterized by the presence of retroperitoneal fibrous soft tissue hyperplasia, surrounding the abdominal aorta and the iliac arteries and extending into the retroperitoneum to envelop neighboring structures, such as ureters. Clinically, it was divided into primary RPF and secondary RPF. Secondary RPF, about one-third of total RPF, is secondary to tumor, infection, trauma, radiation, surgery and drug abuse [3,4]. Primary RPF is also known as idiopathic RPF or Ormond's disease. The cause of which is thought to correlate with immune-related diseases because it is often concurrent with autoimmune disease such as ANCA-associated vasculitis, rheumatoid arthritis and so on. Graziano Ceresini [5] systematically analyzed the relationship between RPF and Hashimoto's thyroiditis and considered RPF were highly correlated with autoimmune diseases. The case reported in this paper also has autoimmune disease systemic lupus erythematosus. Autoimmune diseases and RPF are not coincidence, but have correlation. In addition, part of RPF patients [6], IgG4+ plasma cells were significantly increased in the multi-site tissue immunology examination. Therefore, some RPF also belong to the category of IgG4 related diseases(IgG4-RD) and have some characteristics of IgG4-RD, such as IgG4+ plasma cells and multi-organ infiltration when IgG4/IgG>0.40, in accordance with the diagnostic criteria for IgG4-RD. The RPF is considered to be related to abdominal peri-vascular inflammation due to the hyperplasia of retroperitoneal fibrous tissue end with wrapping abdominal aortic [7].

The initial clinical manifestations of RPF were hidden and varied, and the early symptoms are non-specific, bringing difficulties for early clinic diagnosis. After the emerge with SLE, both SLE-related symptoms and RPF performance were overlapped, which easily lead to missed diagnosis and treatment. Pain in the abdominal or back with no metastasis and radiating is then most common manifestation among

patients with RPF. Some patients also manifest with edema, such as lower extremity edema [8], or scrotum edema [9] and even cerebral edema. The general symptoms include anorexia, fatigue, weight loss, etc. Some patients also showed symptoms of urinary system such as oliguria, dysuria and hematuria. Hypertention may be found in many patients due to renal artery impingement. The ureters was occasionally pressed by the mass when the disease develops into the ureter, which may lead to ureteral stricture [10] that causes acute renal dysfunction or chronic renal failure. Since the clinical symptoms of retroperitoneal fibrosis are nonspecific, the diagnosis of RPF depends on laboratory and imaging examination. The results of routine laboratory examination, such as ESR or CRP, may mainly indicate it is whether they are inflammatory lesions or at active phase, which is beneficial for monitoring the progress of the disease. Due to the urinary system is often involved, urinalysis and renal-function test, in a certain extent, contribute to the indication of renal and ureteral involvement when excluding other causes of impaired renal function. Autoimmune tests are important for the diagnosis of autoimmune disease, autoimmune antibodies such as anti-nuclear antibodies can help to identify whether it is idiopathic RPF. RPF is often associated with ANCA-associated vasculitis [7,11] and ANCA is also positive in the female patient reported in this article, suggesting vasculitis lesions. Some indicators such as connective tissue growth factor (CTGF), matrix metalloproteinases (MMP), were significantly increased in RPF patients, which indicates the degree of fibrosis [12]. Laboratory examination results can only serve as an auxiliary diagnosis; therefore, imaging examination has important value in the discovery of the disease. Imaging examination is very important for the diagnosis of RPF, which also can be used to the identification of primary RPF and secondary RPF. At present, it is considered that enhanced CT is the most direct method for the diagnosis of RPF. It's typical characteristic is that the abdominal aorta and iliac arteries are enveloped by homogeneous density of retroperitoneal hyperplastic tissue and there is often a shift or stricture of the ureter. MRI can clearly show the adjacent tissue, MRI has an increased T1 signal in RPF patient; T2 signal is reduced in the quiescent period, increased in the active period [13]. The pathologic examination is the gold standard for diagnosis of RPF. Color Doppler ultrasound or CT-guided biopsy is the preferred method for obtaining pathological diagnosis. Currently, the treatment of RPF depends mainly on the drug and surgery, which aim to relieve the obstruction caused by fibrosis, terminate the progression of the fibrotic process and prevent recurrence. For the mild condition, hormones and immunosuppressive therapy is efficacious [14]. Glucocorticoid, tamoxifen [15], methotrexate, mycophenolate mofetil, azathioprine, cyclophosphamide and combination therapy are commonly used at present. Vaglio [16] carried out a randomized controlled trial found that prednisone can effectively prevent the RPF recurrence. Therefore, it is recommended as the first chosen to treat RPF. If the imaging examination results of the patients suggested that the ureter was severely blocked, the ureteral decompression was needed. Surgical ureterolysis can obviously relieve the obstruction of ureter, and can prevent the further deterioration of renal function, but it cannot prevent the progression of the primary disease [17]. In summary, RPF is a scarce disease, especially when concurrent with RPF. The pathogenesis of RPF remains unclear and inflammation coexisting with fibrosis is the major feature. Inflammatory exudate collects around the abdominal aorta, ureter and other abdominal organs, promoting fibrosis and resulting in perivascular inflammation and ureteral obstruction. The clinical manifestations are nonspecific. The most common clinical manifestation is abdominal pain in primary RPF and renal insufficiency or renal failure secondary to ureteral obstruction. Vascular complications, such as deep venous thrombosis, are occasionally found in some patients. Laboratory

findings have good value for the diagnosis of autoimmune diseases and can help to diagnose immune-related RPF. Imaging examination is of great value in discovering and diagnosing this disease, and CT scan can be the first method of diagnosing RPF. Color Doppler ultrasound- or CT-guided biopsy is the gold standard for diagnosis of RPF. At present, primary medical therapy of idiopathic RPF is glucocorticoids. If the renal function is severely damaged, surgical ureterolysis can relieve the obstruction.

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