Retroperitoneal Fibrosis: A Retrospective Clinical Data Analysis of 30 Patients in a 10-year Period

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Abstract

Background: Retroperitoneal fibrosis (RPF) is an uncommon disease that is characterized by development of fibrosclerotic tissues involving retroperitoneal structures. This study aimed to investigate the clinical features of 30 patients with RPF in a single center in Beijing in a 10-year period.

Methods: We retrospectively analyzed clinical data on demographic characteristics, clinical manifestations, laboratory findings, radiological findings, modalities of treatments, outcomes and prognosis of 30 patients with RPF. Patients were treated in Beijing Chao-Yang Hospital between January 2003 and December 2013.

Results: The mean age of patients with RPF was 56.7 ± 14.4 years. Twenty-three patients were men and seven patients were women. Acute phase reactants were elevated in most patients. Rheumatic factor was positive in 4/25 (16.0%) patients, and antinuclear antibody was positive in 6/22 (27.3%) patients. Elevation of IgG4 was observed in 9/22 (40.9%) patients. The most common type was I + III (n = 13), followed by I + II + III (n = 12). Five patients undertook an ^{18}F-fluoro-deoxy-D-glucose positron emission tomography examination and increased uptake was detected in four patients. Eight patients received combination therapy with glucocorticoids and tamoxifen. Surgical intervention treatments included intraureteral double-J stent implantation (n = 26), percutaneous nephrostomy (n = 2), open ureterolysis and intraperitonealization of the ureters (n = 5) and laparoscopic ureterolysis and intraperitonealization of the ureters (n = 5). Three patients underwent hemodialysis because of renal failure.

Conclusions: Clinical characteristics of RPF patients in our study are similar to those previously reported. Steroids and immunosuppressive therapy combined with ureterolysis could be a viable choice of treatment for RPF. More prospective, multi-center studies with a longer follow-up are warranted.

Key words: Immunosuppression; Retroperitoneal Fibrosis; Tamoxifen

Introduction

Retroperitoneal fibrosis (RPF), also known as Ormond's disease, is an uncommon disease with unclear etiology.[1] The incidence of RPF is approximately 1–2/100,000.[1] RPF was first described in 1905 by the French urologist Albarran and became fully described as an entity in 1948 by Ormond.[2] RPF is a chronic inflammatory process of the retroperitoneum characterized by the presence of fibrosclerotic tissues involving retroperitoneal structures. This leads to encasement of the ureters, which can cause obstructive nephropathy.

Previous studies have suggested that RPF is either idiopathic or secondary to various types of diseases, including autoimmune diseases and atherosclerosis.[1] The pathogenesis of RPF is still unknown, but immune responses may play an important role.[1] Clinical symptoms and physical examinations in most patients with RPF are usually nonspecific. Laboratory findings may show elevation of the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) and variable renal insufficiency. The diagnosis of RPF is based on typical findings on computed tomography (CT) or magnetic resonance imaging (MRI). However, in some cases, biopsy and histopathological examination are required for definitive diagnosis of RPF. The use of corticosteroids and a combination of renal drainage, if needed, are the most preferred treatment strategies, but the optimum treatment has not been established.

We conducted a retrospective analysis to determine the clinical characteristics of 30 patients with RPF, including

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clinical manifestations, imaging findings, laboratory findings, treatments and prognosis in our solitary center in a 10-year period.

Methods

Patients

From January 2003 to December 2013, 30 patients with RPF were admitted and treated in the Department of Urology, Beijing Chao-Yang Hospital, Capital Medical University. After receiving approval from the committee of medical ethics, we retrospectively analyzed the medical records of these 30 patients.

The diagnosis of RPF was based on clinical findings and typical imaging characteristics on CT or MRI. The typical imaging finding of RPF is a well-delimited, but irregular soft tissue periaortic mass extending from the level of the renal hila to the iliac vessel. This mass usually progresses to the retroperitoneum, causing entrapment of the ureters and inferior vena cava, but without displacing the lower abdominal aorta. Retroperitoneal biopsies were performed in patients with atypical imaging findings or in patients with a high suspicion of malignancy.

Clinical data collection

The following clinical data were collected for each patient: Demographic information, imaging findings, laboratory findings, and immunological parameters during follow-up and treatment. Demographic information [Table 1] included gender, age, smoking history, presenting symptoms and signs, and comorbidities such as hypertension, diabetes mellitus, coronary heart disease and autoimmune diseases. The presenting symptoms [Table 2] were divided into two categories according to previous studies: Localized and systemic.[3] Localized symptoms included pain, lower extremity edema, and scrotum edema. Systemic symptoms included but were not limited to: Fever, fatigue, nausea, vomiting, and anorexia. Laboratory examinations [Table 3] included routine blood tests (e.g. white blood cell, hemoglobin, and platelet), measurement of the ESR, and measurement of CRP, blood urea nitrogen, creatinine and albumin levels. Immunological parameters included the presence of rheumatoid factor and antinuclear antibody, the level of IgG4 and other auto-antibodies.

Imaging findings

The diagnosis and follow-up assessment of treatment depended on CT or MRI results. All of the patients' images were reviewed and collected. The maximal thickness of the RPF mass was measured in the transverse plane of CT or MRI scans. The imaging findings were classified according to a previously reported classification system[4] [Figure 1].

Class I: Soft-tissue density surrounding the infrarenal aorta and/or iliac vessels;

Class II: Soft-tissue density surrounding the infrarenal vena cava;

Table 1: Demographics of patients with RPF

Feature	Results
Age, years	56.7 ± 14.4
Gender, *n* (%)	
Male	23 (76.7)
Female	7 (23.3)
Smoking history, *n* (%)	
Current smoker	12 (40.0)
Ever smoker	6 (20.0)
Comorbidity, *n* (%)	
Hypertension	16 (53.3)
Diabetes mellitus	2 (6.7)
Dyslipidemia	2 (6.7)
Coronary heart disease	3 (10.0)
Cerebral vascular disease	2 (6.7)
Autoimmune disease, *n* (%)	
Sjogren's syndrome	2 (6.7)
Systemic lupus erythematosus	1 (3.3)
BMI, kg/m^2	21.3 ± 2.9

BMI: Body mass index; RPF: Retroperitoneal fibrosis.

Table 2: Clinical manifestations of RPF patients

Features	Number of patients, *n* (%)
Localized symptoms	21 (70.0)
Pain	16 (53.3)
Flank pain	8 (26.7)
Abdominal pain	4 (13.3)
Back pain	4 (13.3)
Lower extremity edema	4 (13.3)
Scrotum edema	1 (3.3)
Systemic symptoms	21 (70.0)
Anorexia	5 (16.7)
Fatigue	6 (20.0)
Fever	1 (3.3)
Weight loss	9 (30.0)
Nausea and vomiting	5 (16.7)
Paruria	11 (36.7)
Frequency and urgency	3 (10.0)
Oliguria	4 (13.3)
Anuria	2 (6.7)
Hematuria	2 (6.7)
Asymptomatic	4 (13.3)
Presenting signs	
Hypertension	16 (53.3)
Kidney region percussion pain	5 (16.7)
Abdominal tenderness	3 (10.0)
Pyeloureterectasis laterality	
Unilateral	17 (56.7)
Left	6 (20.0)
Right	11 (36.7)
Bilateral	9 (30.0)
None	4 (13.3)

RPF: Retroperitoneal fibrosis.

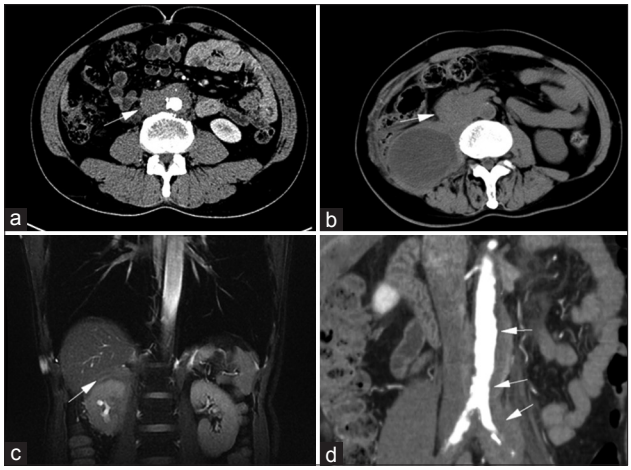

Figure 1: Several typical retroperitoneal fibrosis (RPF) mass imaging. (a) Transverse plane of a computed tomography (CT) scan shows an RPF mass encased aorta and inferior vena cava; (b) Transverse plane of a CT scan shows an RPF mass encasing inferior vena cava. A cystic-solid mass was detected in the right kidney region (postoperative of right-side nephrectomy); (c) Vertical plane of an magnetic resonance imaging scan shows a peri-renal RPF mass; (d) Vertical plane of a CT scan shows a RPF mass encasing the aorta and bilateral iliac arteries.

Class III: Lateral extension of inflammation/fibrosis with compression of one or both ureters;

Class IV: Extension of fibrosis, which includes the renal hilum with compression of the renal artery and/or renal vein.

Continuous variables are shown as mean ± standard deviation. Qualitative variables are shown as a percentage. A $P < 0.05$ was defined as significant. All statistical analyses were performed with SPSS 16.0 software (SPSS Inc., Chicago, IL, USA).

Results

Demographic characteristics of the patients

Clinical data of 30 patients were retrospectively studied and analyzed. Demographic characteristics are shown in Table 1. The mean age of the patients was 56.7 years and most patients were aged between 40 and 70 years. Twenty-three patients were men and seven patients were women. Comorbidities included hypertension (16 cases), diabetes mellitus (2 cases), dyslipidemia (2 cases), coronary heart disease (3 cases) and cerebral vascular disease (2 cases). Three patients had concurrent autoimmune diseases: Two of these patients had Sjogren's syndrome and the other patient had systemic lupus erythematosus. Twelve patients were current smokers and six patients were ever smokers.

Five patients had previously identifiable risk factors for developing RPF [Table 4]. One patient had a history of tuberculosis, one had a history of abdominal surgery, one had a history of a malignant tumor and two had a history of β-blocker use. No patients had a history of previous exposure to asbestosis.

Table 3: Laboratory examination findings at presentation in patients with RPF

Features	Values
White blood cell ($\times 10^9$/L)	8.34 ± 1.89
Hemoglobin (g/L)	114.46 ± 14.80
Platelet ($\times 10^9$/L)	225.56 ± 44.36
Albumin (g/L)	28.91 ± 4.99
ESR (mm/h)	46.80 ± 19.50
CRP (mg/dl)	2.82 ± 2.41
Creatinine (mg/dl)	2.93 ± 4.53
Leukocytosis, *n* (%)	9 (30.0)
Anemia, *n* (%)	16 (53.3)
Hypoalbuminemia, *n* (%)	7 (23.3)
Renal insufficiency, *n* (%)	12 (40.0)
Positive autoantibody, *n* (%)	10/25 (40.0)
Pheumatoid factor	4/25 (16.0)
Antinuclear antibody	6/22 (27.3)
Elevation of IgG4, *n* (%)	9/22 (40.9)

RPF: Retroperitoneal fibrosis; ESR: Erythrocyte sedimentation rate; CRP: C-reactive protein.

Table 4: Patients with predisposing risk factors for developing RPF

Predisposing risk factors	Number of patients, *n* (%)
History of tuberculosis	1 (3.3)
History of abdominal or pelvic surgery	1 (3.3)
History of malignant tumor	1 (3.3)
Prior use of β-blockers	2 (6.7)
Occupation risk with asbestos exposure	0

RPF: Retroperitoneal fibrosis.

Clinical manifestations

The patients' clinical manifestations are shown in Table 2. Twenty-one patients had localized symptoms and twenty-one patients had systemic symptoms. Localized symptoms included pain, lower extremity edema and scrotal edema. Systemic symptoms included anorexia, fatigue, fever, and weight loss. Five patients had nausea and vomiting and eleven patients had paruria. Four patients were asymptomatic. Presenting signs included hypertension, percussion pain in the region of the kidney and abdominal tenderness. Seventeen patients presented with unilateral pyeloureterectasis and nine patients presented with bilateral pyeloureterectasis.

Laboratory examinations

Laboratory findings are shown in Table 3. Laboratory examinations at presentation showed leukocytosis, anemia, hypoalbuminemia and renal insufficiency. Rheumatic factor was positive in 4 out of 25 patients and antinuclear antibody was positive in 6 out of 22 patients. Elevation of IgG4 was observed in 9 out of 22 patients.

Radiological features

Radiological features of the patients were analyzed and classified according to a previously reported

study.[4] The patients' radiological features are shown in Table 5. The most common type was I + III (n = 13), followed by I + II + III (n = 12). Twenty-six patients had hydronephrosis requiring ureteral stenting. Renal atrophy was detected in three patients. Five patients undertook an ^{18}F-fluoro-deoxy-D-glucose positron emission tomography (^{18}F-FDG PET) examination and increased uptake was detected in four patients.

Treatments and outcomes

Modalities of treatments for patients with RPF are shown in Table 6. The mean follow-up period was 30 months (range, 4–136 months). Treatments included glucocorticoids, an immunosuppression agent (cyclophosphamide [CTX]), tamoxifen and combination with surgical interventions. Surgical intervention treatments included intraureteral double-J (D-J) stent implantation (n = 26), percutaneous nephrostomy (n = 2), open ureterolysis and intraperitonealization of the ureters (n = 5) and laparoscopic ureterolysis and intraperitonealization of the ureters (n = 5). Three patients underwent hemodialysis because of renal failure. Eight patients received combination therapy with glucocorticoids and tamoxifen. One patient received combination therapy with glucocorticoids and cefotaxime. The average initiation dose of prednisolone was 50 mg daily (20–60 mg/d), and it was tapered to a low dose of <10 mg/d within 3–6 months. The ESR and CRP levels decreased to normal and the retroperitoneal mass decreased in size after treatment. Patients with D-J stent implantation underwent regular stent replacement and D-J stents in five patients were successfully removed after immuno-regulation treatments. One patient received metallic stent implantation. One patient experienced recurrence after surgical intervention alone. Eight patients' final diagnosis was confirmed by histological examinations, which showed ureters encased by fibrotic tissues with chronic inflammation.

Table 5: Radiological features of patients with RPF at diagnosis

Features	Number of patients, *n* (%)
Location of mass	
Periaortic, periiliac	22 (73.3)
Periaortic	2 (6.7)
Periaortic, pericaval	11 (36.7)
Presacral	5 (16.7)
Retrovesical	2 (6.7)
Peripancreatic	1 (3.3)
Paracolic	1 (3.3)
Perirenal	1 (3.3)
Classification	
I	3 (10.0)
I + III	13 (43.3)
I + II + III	12 (40.0)
I + II + III + IV	2 (6.7)
Hydro-ureteronephrosis	26 (86.7)
Bilateral	17 (56.7)
Unilateral	9 (30.0)
Renal atrophy	3 (10.0)
^{18}FDG-PET	5 (16.7)
Increased uptake	4 (13.3)

RPF: Retroperitoneal fibrosis; ^{18}FDG-PET: ^{18}F-fluoro-deoxy-D-glucose positron emission tomography.

Table 6: Modality of treatment for patients with RPF

Modality of treatment	Number of patients, *n* (%)
Glucocorticoids	15 (50.0)
Immunosuppression agent (CTX)	1 (3.3)
Tamoxifen	9 (30.0)
Glucocorticoids + immunosuppression agent (CTX)	1 (3.3)
Glucocorticoids + tamoxifen	8 (26.7)
Surgical interventions	
Intraureteral double-J stent implantation	26 (86.7)
Percutaneous nephrostomy	2 (6.7)
Open ureterolysis and intraperitonealization of ureters	5 (16.7)
Laparoscopic ureterolysis and intraperitonealization of ureters	5 (16.7)
Hemodialysis	3 (10.0)

RPF: Retroperitoneal fibrosis; CTX: Cyclophosphamide.

Discussion

Retroperitoneal fibrosis is a rare chronic inflammatory disease involving the retroperitoneum and causing compression of the retroperitoneal structures, especially the ureters, this frequently leads to pyeloureterectasis. RPF patients have a male predominance and a male-to-female ratio of approximately 2–3:1. RPF can be idiopathic or secondary to various reasons, such as surgery, drugs, malignant neoplasms, radiation and infections.[5] Chronic infectious diseases, malignancies and drug consumption, should be excluded to confirm diagnosis of idiopathic RPF.[1] The pathogenesis of RPF is uncertain. However, various recent studies have suggested that RPF may be caused by chronic inflammatory conditions that are induced and maintained by autoimmune responses.[6]

Our study results showed that RPF occurred between the age of 40 and 70 years and the male-to-female ratio was 3.3:1, which is similar to previous studies.[4] Preliminary researches have suggested a relationship between RPF and cardiovascular disease risk factors.[1,7,8] In our study, risk factors of RPF included hypertension, diabetes mellitus, coronary heart disease, dyslipidemia, cerebral vascular disease, and tobacco exposure. Our study showed that predisposing factors of RPF comprised of history of tuberculosis, abdominal surgery (uterectomy), malignant tumor (carcinoma *in situ* of the urinary bladder), and previous use of β-blockers.

The most common symptom in our study was pain (flank, abdominal, and back), which is similar to previous reports.[4,9] This pain, which is typically dull, noncolicky and unchanged with posture, may radiate to the lower abdomen or groin. Four patients in our study had lower extremity

edema, indicating a compression effect of the ilio-femoral veins, caused by the RPF mass. Paruria was detected in 11 patients (frequency and urgency in 3 patients, oliguria in 4 patients, anuria in 2 patients, and hematuria in 2 patients), and this may have been caused by urinary tract infection induced by D-J stents implantations. Pyeloureterectasis induced by ureteral encasement by the RPF mass was observed in 26 patients (unilateral in 17, bilateral in 9).

Laboratory examinations showed that acute-phase reactants such as the ESR and the CRP levels were increased in most patients. When the ESR and CRP levels decreased after immunoregulation therapy, the maximal plane of the RPF mass decreased and the symptoms of the patients were relieved. This suggested an association between acute-phase reactants levels and the patients' progression. An *et al*.[10] showed that the changes in the ESR and CRP levels were strongly correlated with changes in CT/MRI. However, Pelkmans *et al*.[11] believed that patients with RPF who had elevated acute-phase reactants were more symptomatic and neither acute-phase reactants nor their initial changes could be taken as a major predictor for treatment success. Therefore, the ESR and CRP levels lack sensitivity and specificity for the diagnosis of RPF, and further investigations are required. Renal insufficiency was observed in 12 (40%) patients in our study, which is higher than that in previous studies.[8,12] Rheumatoid factor was positive in 4 out of 25 patients and antinuclear antibody was positive in 6 out of 22 patients in our study, Vaglio *et al*.[5] believed that positive autoantibodies do not portend the presence or development of clinical manifestations of RPF.

Recently, an international consensus has been reached to use the term "IgG4-related disease (IgG4-RD)" by an international symposium that aimed to improve current understandings and facilitate communications.[13] The term "IgG4-RD" encompasses several disorders, depending on the organ involved. Diseases such as RPF, type I autoimmune pancreatitis, and some forms of inflammatory orbital pseudotumor share pathological, sclerotic, and clinical features including tissue infiltration with IgG4-positive cells.[14] Serum IgG4 levels often elevate to greater than 1.35 g/L in IgG4-RD. However, high serum levels of IgG4 are not entirely specific to IgG4-RD. In our study, elevation of IgG4 was detected in 9 out of 22 patients.

Ultrasonography is simple to perform and is minimally-invasive but has poor overall sensitivity in detecting RPF.[1,15,16] Intravenous urography and retrograde pyelography have been replaced by cross-sectional imaging. Currently, multi-detector CT and MRI are considered the mainstay of choice for the diagnosis of RPF. The typical morphological characteristics of idiopathic RPF and most forms of benign secondary forms of RPF consist of a well-delimited but irregular soft-tissue density mass extending from the level of the renal hila to the iliac vessels, and often progressing through the retroperitoneum to encase the ureters and inferior vena cava.[17-19] Enhancement of the RPF mass depends on the stages of the disease in enhanced CT scan. Different stages of enhancement could be helpful in evaluating the patient's response to therapy.[19] MRI is equivalent to CT, and provides an ideal choice in accessing the characteristics of an RPF mass and its effects on adjacent structures, but it is superior to CT in its high contrast resolution. Early soft-tissue enhancement reflects the degree of inflammatory activity in T2-weighted imaging.[19] Moreover, MRI features may be helpful in distinguishing between RPF and lymphoma.[20] Brandt *et al*.[21] showed that dynamic enhancement analysis MRI is able to distinguish patients with different response rates to medical treatment of idiopathic RPF, and might be helpful to individualize therapeutic decision-making. ^{18}F-FDG PET is a functional imaging modality with high sensitivity, but low specificity. Therefore, this technique is not helpful in distinguishing between idiopathic and other benign forms of RPF.[19,22,23] ^{18}F-FDG PET is superior to CT/MRI in revealing active inflammation and predicting post-treatment outcomes.[19] ^{18}F-FDG PET may be also be used to evaluate responses of treatment and recurrence during follow-up.[19,24,25] The most common type of radiological finding in our research was type I + III, which is similar to the previous study.[8] In our study, increased uptake was detected in four out of five patients who underwent the ^{18}F-FDG PET examination. However, further studies concerning the applications of the ^{18}F-FDG PET are warranted.

Inhibiting or relieving obstruction of the ureters or other retroperitoneal structures, switching off the acute-phase reaction and its systemic manifestations and preventing disease from relapse are the aims of treatment of idiopathic RPF.[1] To date, medical treatments of RPF consist of corticosteroids and/or immunosuppressive therapy. Steroid therapy is considered as the first line treatment for RPF, regardless of its causes. Immunosuppressive strategies previously reported for treating RPF included the azathioprine, cyclosporine, CTX, methotrexate, mycophenolate mofetil and tamoxifen.[26-29] van Bommel *et al*.[29] conducted a single-center prospective, observational study of 55 patients with idiopathic RPF who were treated with tamoxifen for 2 years. This study showed that tamoxifen was a safe and viable therapeutic option in the treatment of RPF. Vaglio *et al*.[30] concluded that prednisone is more effective in the prevention of relapses than tamoxifen in patients with idiopathic RPF. They suggested that prednisone should be considered as the first-line treatment for patients with newly diagnosed idiopathic RPF. In our study, 15 patients received prednisone therapy, 9 patients received tamoxifen therapy, and 8 patients received a combination therapy of prednisone and tamoxifen.

To date, no widely accepted therapeutic schedule for RPF has been established. Obtaining biopsies of the RPF mass during release of the ureters and their transposition inside the peritoneal cavity, which is completed with ureters wrapped with omentum to prevent a new entrapment, are the main goals of surgical treatment of RPF.[31] Arvind *et al*.[32] concluded that laparoscopic ureterolysis and omental

wrapping in the setting of obstructive uropathy were safe and effective alternative with a high success rate at mid-to-long term follow-up. Another study reported the first documented use of SurgiWrap to wrap the ureters for minimizing the future possibility of recurrent fibrosis, compression, and ureteral obstruction.[33] Haddad *et al*.[34] presented three RPF patients who had failed initial endourologic/surgical management underwent insertion of simultaneous bilateral subcutaneous pyelovesical bypass grafts and gained an improved quality of life. Bourdoumis *et al*.[35] reported the first study using thermo-expandable Memokath 051 stents in patients with RPF, and concluded that it can be considered as a safe, minimally invasive, and effective long-term means of managing ureteral obstruction in RPF. However, Williams *et al*.[36] reported a case of RPF with spontaneous remission without any medical management. Therefore, further large-sample, prospective, multi-center studies and longer follow-up are required.

In conclusion, clinical characteristics of RPF patients in our study are similar to those previously reported. It is of great significance to recognize RPF as early diagnosis and treatment could alleviate symptoms, reduce acute-phase reactants, preserve renal function, and improve prognosis. Steroids and immunosuppressive therapy combined with ureterolysis could be a viable choice of treatment.

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