

# Tamoxifen Monotherapy in the Treatment of Retroperitoneal Fibrosis

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## Key Words

Retroperitoneal fibrosis · Ormond's disease · Tamoxifen

## Abstract

**Objective:** To evaluate the therapeutic effect of tamoxifen monotherapy in patients with retroperitoneal fibrosis (RPF). **Patients and Methods:** From 2007 on, 31 patients with idiopathic RPF were treated with tamoxifen monotherapy. Follow-up investigations included magnetic resonance imaging, laboratory measurements, registration of side effects and changes or removal of ureteral stents. Data were stored in the Else Kröner-Fresenius Registry of Retroperitoneal Fibrosis. **Results:** 25 men and 6 women with a mean age of 56.6 years were treated with tamoxifen monotherapy. Mean duration of treatment was 13.3 months, mean follow-up 26.8 months. A total of 44 renal units were affected by hydronephrosis and covered by DJ stents. Radiological regression of fibrosis was detected in 22 cases (71.0%); removal of ureteral stents was possible in 27/44 renal units (61.4%) and 17/29 patients (58.6%), respectively. Most patients showed only mild or no side effects of therapy. In 7 cases (22.3%) tamoxifen therapy had to be abandoned because of severe side effects, progression of fibrosis or persistent intolerance. **Conclusions:** Tamoxifen is an alternative in the medical treatment of RPF, especially if patients want to avoid gluco-

corticoids. The potential of regression of fibrosis seems to be slightly inferior and the relapse rate is higher compared to steroids, but the rate of successful DJ removals is comparable.

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## Introduction

Retroperitoneal fibrosis (RPF) in its typical form is characterized by fibroinflammatory tissue that spreads around the abdominal aorta, reaching from underneath the renal vessels to underneath the aortal bifurcation without displacing the abdominal aorta from the lumbar spine [1, 2]. Urologists are confronted with this disease because the most frequent complication is ureteral obstruction in 60–90% [3–7]. Other main clinical presentations are unspecific pain, fatigue, malaise, fever, night sweats and weight loss [3–7]. Idiopathic RPF is found in more than two-thirds of all cases with no reasonable cause of origin. Furthermore it occurs secondarily, e.g. after medical or surgical treatment, infection, neoplasm, trauma or radiotherapy [1, 3].

As the etiology of idiopathic RPF remains unclear and due to the lack of controlled trials, the medical treatment of RPF is still rather empirical. The goals of thera-

py are to remove the ureteral obstruction and to avert progression and recurrence of the fibrosis [1]. Traditionally, the relief of urinary tract obstruction has been surgical, but at present the primary approach is often medical, after initial relief with ureteral stenting or nephrostomy [8].

The classical approach to medical treatment is glucocorticoid monotherapy because of both the spectrum of reported immunologic findings and the spectrum of inflammatory syndromes, but the optimum dose and duration of medical treatment is still unknown, and glucocorticoids have several severe side effects [8–13]. Many practitioners use immunosuppressive drugs, e.g. azathioprine [14, 15], mycophenolate-mofetil [16, 17], cyclophosphamide [18] or colchicine [19] in addition to glucocorticoids to reduce the dose of steroids and to combine the anti-inflammatory and immunosuppressive effects, but the superiority of these combination therapies is still unproven [14].

Because of its anti-inflammatory and antifibroblastic effects, tamoxifen was used in the treatment of RPF and first described in several case reports before van Bommel et al. [20] presented the first series of 19 patients treated with tamoxifen monotherapy in 2006. The long-term safety of tamoxifen was proven by an extension of his series [21]. Furthermore, Vaglio et al. [19] treated 18 patients with tamoxifen monotherapy after induction therapy with prednisolone. We present our experiences of tamoxifen monotherapy in a series of 31 patients with RPF.

## Patients and Methods

From April 2007 to March 2012 a total of 31 patients with RPF were treated with tamoxifen monotherapy in our department. The diagnosis had been secured either by findings on computed tomography/magnetic resonance imaging (CT/MRI) or by histologic proof or both.

Patients' data were recorded in the Else Kröner-Fresenius Registry of Retroperitoneal Fibrosis in Germany, a nationwide registry for RPF headquartered in our department [3]. The data sheet for registration consisted of questions concerning demographic data, date of diagnosis, pre-existing and accompanying diseases, previous and current medical and surgical therapies, ureteral stenting and symptoms at the beginning and over the course of the disease. Additionally laboratory values and clinical reports of CT and MRI were recorded in the registry.

Before the beginning of treatment all patients received laboratory examinations and baseline MRI to record the expansion of fibrosis. If necessary, renal drainage was done by ureteral stenting. After careful exclusion of malignant disease and contraindications, medical therapy was started with tamoxifen 20 mg twice a day. All patients gave informed consent to treatment.

Follow-up examinations were performed after 3, 6, 12, 18 and 24 months. In every follow-up all patients received MRI, clinical and laboratory examinations. Patients were asked about side effects of the medical therapy in a questionnaire consisting of 36 questions concerning headache, nausea, mood changes, sleeping disorders, allergic reactions, flushes or sweats, weight changes, visual problems, infections, pain, loss of libido, vaginal bleeding and signs of thrombosis. All female patients receiving tamoxifen were consulted by a gynecologist twice a year.

In every follow-up MRI, response to treatment was evaluated by 3 independent observers (2 radiologists, 1 urologist) and categorized into one of five categories: (0) progression of disease; (I) stable disease, size reduction <20%; (II) mild regression of fibrosis, reduction 20–50%; (III) significant regression, reduction >50%; (IV) complete regression, i.e. no further delineable fibrosis in all examinations.

In cases of fibrosis regression and in accordance with the patient's wish, DJ stents were removed and success was evaluated by intravenous pyelogram and/or MAG3 scan. In every follow-up examination each case was re-evaluated to decide whether to resume or change medical therapy or to perform surgery for ureteral obstruction. After successful medical or surgical therapy patients were followed up by MRI twice a year for the first year and once a year afterwards.

### *Data Storage and Statistical Analyses*

All patients gave written consent to storage and analysis of their personal and disease-related data in the Else Kröner-Fresenius Registry of Retroperitoneal Fibrosis. For data storage we used an SQL database in pseudo-anonymous form, conforming to the standards of the ethics committee of the University Witten/Herdecke.

Statistical analyses were performed with the Wilcoxon rank-sum test for two groups. Fisher's exact test was used for contingency tables. For all tests  $p < 0.05$  was considered statistically significant. All tests were performed with commercial software (Microsoft Excel, XLSTAT).

## Results

We treated 31 patients with RPF with tamoxifen monotherapy in our department. Mean age at diagnosis was  $56.6 \pm 12.1$  (35–80) years; 6 patients were women (19.4%) and 25 men (80.6%). Idiopathic RPF was histologically confirmed in 22 cases (71.0%); of these, 18 cases were confirmed before onset of therapy (CT-guided biopsy in 8, laparoscopic biopsy in 7 and open biopsy in 3) and 4 during or after medical therapy of idiopathic RPF in the course of operative treatment. In 9 patients idiopathic RPF was assumed from typical formation of fibrosis in CT diagnosis. The demographic and clinical data of patients are presented in table 1.

Bilateral hydronephrosis necessitating urinary diversion was present in 15 patients (48.4%), unilateral hydronephrosis owing to idiopathic RPF in 14 patients (45.2%)

(right: 8, left: 6), and a retroperitoneal mass without ureteral involvement was found in 2 (6.5%). All patients with ureteral obstruction received primary renal drainage by DJ stenting.

All 31 patients started with tamoxifen monotherapy 20 mg twice a day. Medication was discontinued in 7 patients (22.6%) after a mean duration of  $5.1 \pm 2.9$  (3–11) months because of side effects in 3, progression of fibrosis in 3 and persistent intolerance in 1 case, who suffered from vomiting and malaise after every intake of tamoxifen. Mean duration of medical therapy without dropouts was  $13.3 \pm 4.9$  (8–27) months.

The analysis of follow-up MRI examination showed 3 cases of disease progression; RPF remained stable without any regression in 6, regressed mildly in 7, significantly in 11 and completely in 4. There were no statistically significant differences between patients without (categories 0 and I) and with (categories II–IV) regression of fibrosis according to age ( $p = 0.144$ ), sex ( $p = 0.176$ ), immunoglobulin G4 (IgG4) serum value ( $p = 0.688$ ), erythrocyte sedimentation rate (ESR) ( $p = 0.593$ ) and C-reactive protein (CRP) ( $p = 0.683$ ).

DJ stents could be removed after medical therapy in 17 of 29 patients (58.6%) in a total of 27 of 44 renal units (61.4%), and all 17 patients were considered 'free of stents' after successful tamoxifen monotherapy. As to regression of fibrosis there were no statistically significant differences between patients with and without successful DJ removal according to age ( $p = 0.064$ ), sex ( $p = 0.669$ ), IgG4 serum value ( $p = 0.831$ ), ESR ( $p = 0.825$ ) and CRP ( $p = 0.713$ ). Furthermore no single or combination of laboratory values could serve as a predictive factor for successful medical therapy. The mean duration until DJ removal was  $8.8 \pm 7.0$  (3–27) months.

In 12 patients either renogram or MAG3 scan or both showed ureteral obstruction after stent removal, necessitating reinsertion. Of these 12 patients 1 was lost to follow-up and 7 received final operative therapy: ureterolysis in 3, psoas-hitch ureteroneocystostomy in 1 and ureteral reconstruction with ileum segments in 2. In 1 patient nephrectomy was performed due to loss of function. In 4 patients medical therapy was changed to a combination of prednisolone and azathioprine which led to a regression of fibrosis in all 4 cases. Nevertheless, DJ stents could only successfully be removed in 1 case after another 9 months of second-line medical therapy. One patient received ureterolysis and 2 patients were permanently covered with DJ stents.

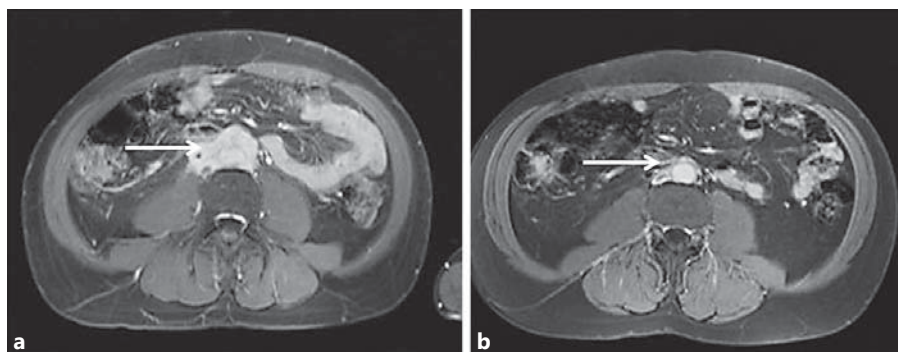
Two patients had side effects of tamoxifen in the form of pulmonary embolism and deep vein thrombosis, re-

**Table 1.** Demographic and clinical data of patients treated with tamoxifen

Characteristic	%	
Age, years	56.6±12.1	
Male	25	80.6
Female	6	19.4
Smoking	23	74.2
Hydronephrosis	29	93.5
Bilateral	15	48.4
Unilateral	14	45.2
Pre-existing disease*		
Hypertension	19	61.3
Diabetes mellitus	5	16.1
Arteriosclerosis	5	16.1
Immune-mediated disease	3	9.7
Hashimoto's thyroiditis	1	3.2
Riedel thyroiditis	1	3.2
Unspecified vasculitis	1	3.2
Hyperthyroidism	1	3.2
Hypothyroidism	2	6.5
Dupuytren's disease	1	3.2
Crohn's disease	1	3.2
Rheumatism	1	3.2
Malignancies	0	0
Symptoms at onset*		
Back pain	25	80.6
Flank pain	23	74.2
Upper abdominal pain	6	19.4
Lower abdominal pain	13	41.9
Testicular pain/hydrocele	6	19.4
Leg pain	3	9.7
Fatigue	18	58.1
Malaise/vomiting	5	16.1
Fever	6	19.4
Night sweats	9	29.0
Weight loss	10	32.3
Laboratory values		
Increased CRP	22	71.0
Increased ESR	23	74.2
Increased IgG4	6	19.4

\* Multiples possible.

spectively. One woman developed ovarian cysts leading to pain and ureteral obstruction of the contralateral ureter. In all cases medical therapy with tamoxifen was interrupted at the time side effects were noticed. Minor side effects of therapy occurred in a total of 16 patients (51.6%) at the onset of therapy. The most frequent side effect was malaise, which mostly dissolved after the first weeks. In the course of medical therapy mild adverse effects occurred or persisted in 7 patients (22.5%) (table 2).



**Fig. 1.** Extent of fibrosis in a patient before (a) and after (b) 6 months of treatment with tamoxifen monotherapy.

**Table 2.** Side effects of medical therapy with tamoxifen (multiples possible)

Side effect	At beginning of medication	Over the course of medication
Progression of fibrosis*	0 (0%)	2 (6.5%)
Pulmonary embolism*	0 (0%)	1 (3.2%)
Deep vein thrombosis*	0 (0%)	1 (3.2%)
Ovarian cyst*	0 (0%)	1 (3.2%)
Malaise/vomiting	5 (16.1%)	2 (6.5%)
Hot flush	4 (12.9%)	1 (3.2%)
Headache	4 (12.9%)	3 (9.7%)
Skin eruption	2 (6.5%)	0 (0%)
Fatigue	2 (6.5%)	1 (3.2%)
Depressive episodes	1 (3.2%)	1 (3.2%)

\* Severe side effects leading to interruption of treatment.

The mean follow-up after medical therapy with tamoxifen was  $26.8 \pm 15.1$  (0–55) months. No recurrences after successful therapy have been recorded until now. An example of the regression of fibrosis is presented in figure 1.

## Discussion

Idiopathic RPF remains a disease rather seldom treated in urologic practice. Several approaches to medical treatment have been described in the literature, but the lack of controlled trials has meant that treatment has not been standardized and is still largely empirical [4]. Because of the nonspecific inflammatory nature of idiopathic RPF, corticosteroids are often used at onset [19]. The main problem of these therapy regimes are side effects such as cushingoid changes, weight gain plus changes in

glucose tolerance and bone metabolism that are both severe and extremely wearing for the patient and often lead to decreased patient compliance and to therapy interruptions. As many patients want to avoid glucocorticoids, tamoxifen seems to be a promising alternative. Especially patients at risk of developing diabetes or osteoporosis and patients with obesity could benefit from avoiding glucocorticoids. As until now there are only few series about treatment of RPF with tamoxifen monotherapy, we believe that our series is able to support the insight into the medical treatment of patients suffering from RPF.

The exact mechanism of tamoxifen on fibrosis is still unknown, but it seems to have anti-inflammatory or antifibroblastic activity in addition to its antiestrogenic effects [4]. Besides the classical treatment in breast cancer its antifibroblastic effect is also used in the treatment of Peyronie's disease and pelvic fibromatosis [22–24].

Tamoxifen monotherapy as a possible treatment of RPF has been described in several anecdotal case reports before van Bommel et al. [20] published the first extensive series of 19 patients in 2006. In their series 14 clinical responders (73.7%) showed a slow but steady regression of the retroperitoneal mass with almost no side effects. In the extension of their series they documented 85.5% of regression under tamoxifen monotherapy. This compares well to our series, which showed regression of fibrosis in 22/31 patients (71.0%). Regression rates for steroid therapy alone are reported between 79.2% [10] and 91.6% [11].

As Vaglio et al. [25] stated in their letter to the editor, the series of van Bommel was limited by the fact that only 47% respectively 54.5% of patients had hydronephrosis, which is a rather small number compared to other study groups. In our series hydronephrosis was found in 93.5% of patients so that the effect of tamoxifen on ureteral obstruction could be assessed more precisely.

The success rate of DJ removal should be a major end point of treatment, but several investigations refuse to mention it in their studies [7, 11]. Success rates vary in the literature, reaching from 50.0 to 90.9% [13, 17, 18], comparing well with our findings of 61.4% successful stent removals.

Another critical point is the relapse or rate of progression of fibrosis during the medical treatment of fibrosis. After successful induction therapy Vaglio et al. [19] found relapse of disease in 7/18 patients (38.9%) treated with tamoxifen, whereas the relapse rate was only 5.5% in patients treated with prednisone. In our series progression of fibrosis occurred in 3 cases (9.7%), leading to a modification of treatment, so the relapse rate in tamoxifen monotherapy seems to be higher than with glucocorticoids, a fact patients should be informed about before the beginning of treatment.

Similar to previous reports, most adverse effects of tamoxifen were only mild and dissolved regularly after induction therapy. Nevertheless, 3 patients had side effects that led to treatment interruption, emphasizing the importance of screening for thrombosis and gynecological attendance in women treated with tamoxifen. The frequency of pulmonary embolism amounts to 3.6% in the only comparable study of van Bommel et al. [21], comparing well to our findings. Vaglio et al. [19] found a significantly higher number of cushingoid changes and grade 2 hypercholesterolemia in patients treated with prednisolone in the only study so far comparing both therapy regimes. The frequency of high-grade adverse effects in patients treated with glucocorticoids or immunosuppressive drugs varies significantly in the literature as many studies refuse to report adverse effects and others report up to 20–25%. The most frequent reported side effects are leukopenia, anemia, hyperglycemia and diabetes mellitus, hypertension, recurrent pancreatitis, bone fracture and myopathy [8–21].

The impact of IgG4 is still unknown, but several authors recommend the classification of IgG4-related and non-IgG4-related disease [26, 27]. Furthermore some authors report better response rates in patients with IgG4-related disease. Marumo et al. [28] reported a patient with RPF and increased IgG4 serum values who responded extremely well to steroid therapy, so they assumed that elevated serum IgG4 may predict the sensitivity to steroid therapy in RPF. Vaglio et al. [19] found 4 cases with high IgG4+ plasma cell infiltration, and all 4 patients achieved remission with no case of relapse in the follow-up of 26 months. In contrast to their investigations we found no statistically significant difference in IgG4 serum levels in

patients with and without regression of fibrosis under medical treatment. As reported before [29, 30], acute-phase reactants such as ESR and CRP were also only poor predictors of differentiation between patients with and without treatment success in our series.

Our series is certainly limited by the still small number of patients and the lack of a control group. Furthermore, patients were not randomized to therapy; their preference influenced the agent used. Therefore, further studies of treatment outcomes with more patients and in comparison to patients treated with glucocorticoids or combination therapy from the Else Kröner-Fresenius Registry of Retroperitoneal Fibrosis are planned.

## Conclusion

Tamoxifen monotherapy is a promising alternative approach to the medical treatment of RPF, especially if patients want to avoid glucocorticoids. The potential of regression of fibrosis seems to be slightly inferior to steroids and/or immunosuppressive drugs, but the rate of successful DJ removals in case of ureteral affection is comparable and the recurrence rate after successful treatment seems to be small. The main advantage of tamoxifen monotherapy is that there are mostly mild side effects that often resolve after induction therapy. Nevertheless, patients must be followed up regularly to detect severe adverse effects at the right time. Patients should be informed that a progression of fibrosis under medical therapy with tamoxifen occurs more often than with steroids or immunosuppressive drugs.

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