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Retroperitoneal Fibrosis: Case Series of 20 Patients

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Abstract: Retroperitoneal fibrosis (RPF) is a rare disease. The objective of this paper is to study the prevalence, clinical, biological and radiological aspects of the RPFand to specify its therapeutic modalities. This study provides descriptive evidence for a series of 20 patients with RPF. We conducted a retrospective study of cases of RPF diagnosed in the Urology and Internal Medicine departments of the Ibn Sina Military Hospital in Marrakech over a five-year period from July 2010 to December 2015. The positive diagnosis of RPF was retained either on the basis of a histological analysis from a biopsy fragment or, in the absence of histological evidence, by the detection on a scan of an infiltrate or "sleeve" Regular and homogeneous tissue density, perivascular topography, surrounding the abdominal aorta, more or less extended to the iliac vessels and able to take the contrast after injection. They were 15 men and 5 women with an average age of 53 years with extremes of 41 to 74 years. Almost all patients had mainly lumbar and abdominal pain. An inflammatory syndrome existed in all cases and renal insufficiency in 15 cases. Radiological investigations showed unilateral or bilateral hydronephrosis in 16 cases. RPF was idiopathic in 14 patients. Secondary forms were present in 6 patients (three atheromatous diseases, one gastric tumor, one case associated with Riedel's thyroiditis and one case of systemic fibrosis associating RPF, Riedel's thyroiditis and liver fibrosis). The treatment consisted of corticosteroids in 19 cases, surgery in 17 cases associated with corticosteroid therapy or even immunosuppressant in the case of steroid-dependence. Two deaths were observed in this series; one of these two patients had developed cervix carcinoma with pleural and peritoneal metastases after 5 years of immunosuppressive therapy. The second died due to the complications of his gastric neoplasm. The general characteristics of patients included are similar to the other series of the literature. The frequency of secondary forms seems to be underestimated, hence the value of a rigorous etiological investigation. The therapeutic strategy and the use of the cortisone saving treatments should be specified by randomized therapeutic trials. The prognosis is generally good; however, the possibility of recurrences, the incidence of which is greater during the first five years, requires prolonged clinical, biological and radiological monitoring.

Keywords: Retroperitoneal Fibrosis, Atheroma, Neoplasm, Vasculitis, Riedel's Thyroiditis, Corticosteroids

1. Introduction

Retroperitoneal fibrosis (RPF), first described in 1948 by Ormond, is a rare disease characterized by the presence of a fibro-inflammatory tissue, which usually surrounds the abdominal aorta and the iliac arteries and extends into the retroperitoneum to envolop neighboring structures-ureters [1, 5].

It is endowed with a great clinical polymorphism, a considerable etiological disparity and is characterized by the absence of therapeutic consensus [2, 3].

Retroperitoneal fibrosis is often idiopathic. It may be secondary in about one third of the cases to many underlying diseases, including infections such as tuberculosis, autoimmune diseases, certain neoplasias, asbestosis, certain drugs, radiotherapy or previous heavy surgery or Even more

recently the fibro- sclerosing disease (IgG4 related-disease) [3] or Erdheim-Chester disease [4].

The diagnostic approach of retroperitoneal fibroses has been modified in recent years by the progress of imaging, allowing the visualization of the fibrosis plate directly, but its discovery is often delayed because of its great clinical latency and the aspecific character of the revealing symptoms [6].

The treatment of retroperitoneal fibrosis has long remained strictly surgical. More recently, other therapeutic means have been proposed. Thus, the use of immunosuppressive drugs, including corticosteroids in the first place, has been proposed. In advanced stages of the disease, surgical treatment may be associated with medical treatment. Urinary drainage in case of obstructive renal failure, followed by surgical release of ureters by ureterolysis [5, 7, 8].

The objective of this paper is to study the prevalence, clinical, biological and radiological aspects of the RPFand to specify its therapeutic modalities.

2. Materials and Methods

We conducted a retrospective study of cases of RPF diagnosed in the Urology and Internal Medicine departments of Ibn Sina Military Hospital in Marrakech over a five-year period from July 2010 to December 2015.

The positive diagnosis of RPF was retained either on the basis of a histological analysis from a biopsy fragment or, in the absence of histological evidence, by the detection on a scan of an infiltrate or "sleeve" Regular and homogeneous tissue density, perivascular topography, sheathing the abdominal aorta, more or less extended to the iliac vessels and able to take the contrast after injection.

When a histological sample was taken, the diagnosis was retained in the presence of polymorphic fibro-inflammatory tissue composed of an infiltrate more or less dense in lymphoid, plasmocytoid, macrophage and eosinophilic polynuclear cells.

No immunohistochemical markers were specifically required.

The collection of data was carried out on the basis of the clinical records using a standardized collection sheet including clinical, paraclinical, therapeutic and evolutive data.

3. Results

3.1. Epidemiological, Clinical and Biological Characteristics

We collected 20 patients in whom the diagnosis of RPF was retained, 15 men (75%) and 5 women (25%), with a male/female ratio of 3. The average age of patients at diagnosis was 53 years with extremes ranging from 41 to 74 years. Almost all patients had mainly lumbar and abdominal pain. An inflammatory syndrome existed in all cases and renal insufficiency in 15 cases.

Table 1 summarizes the main symptoms and clinical signs of the patients as well as their biological characteristics.

Table 1. *Clinical and Biological Characteristics.*

Patients	n (%)
Initial Clinical Manifestations	
Pain (abdominal or flank or lumbar)	18 (90%)
Slimming	8 (40%)
Hypertension	6 (30%)
Fever	3 (15%)
Deep vein thrombosis	2 (10%)
Edema of the lower limbs	3 (15%)
Hydrocele	1 (5%)
Vomiting	2 (10%)
Diarrhea	2 (10%)
Constipation	1 (5%)
Dysuria, anuria	7 (35%)
Macroscopic haematuria	1 (5%)
Ascite	1 (5%)
Pleurisy	0
Portal hypertension	0
Ictere	0
Biological examinations	
Increase in SV> 30 mm / 1 h	20 (100%)
C-reactive protein (CRP)> 10 mg / L	17 (85%)
Leucocytosis (leukocytes> 10 × G / L)	4 (20%)
Thrombopenia	1 (5%)
Anemia (hemoglobin <11 g / dL)	5 (25%)
Serum creatinine> 110µmol / L	15 (75%)
LDH increased	4 (20%)
Proteinuria	4 (20%)
Hyperkalemia	3 (15%)
Antinuclear antibodies	0 (0)
Anti-cytoplasmic anticytoplasm antibodies of neutrophils	0 (0)
High serum tumor markers (ACE, CA15-3, CA19-9, CA125, , FP, HCG and PSA)	0 (0)

3.2. Radiological Examination Results

Abdominal ultrasound was performed in 17 patients (85% of cases). This examination had found an ureterhydronéphrosis (UHN) in 16 cases, of which 11 were bilateral and 5 were unilateral. However, the presence of a hypoechoic retroperitoneal mass surrounding the aorta and the inferior vena cava was found only in 3 cases (15% of cases).

Abdominal scannographic imaging was performed in all the patients. Magnetic resonance imaging (MRI) was performed in 3 patients.

The extension of fibrosis has been classified according to the Scheel classification [9]:

- 02 patients (10% of cases) have fibrosis of tissue density surrounding the renal aorta and / or iliac vessels (Scheel class I).
- 03 patients (15% of cases) had fibrosis of tissue density surrounding the inferior vena cava (IVC)(class II).
- 15 patients (75% of the cases) have lateral extension of fibrosis with compression of one or two ureters (class III).
- No case of extension of fibrosis including the renal hilum with compression of the renal artery or the renal vein (class IV).

The angio-MRI was performed in a single patient who had RPF complicated of renal insufficiency and IVC thrombosis.

Angio-MRI showed the presence of a retroperitoneal tissue in hyposignal T1, surrounding and compressing the aorta, primary iliac arteries and IVC. It had also objectified partial thrombosis of the inferior vena cava under renal, of the primitive iliac veins, and of the right external iliac vein.

Three patients had intravenous urography. The results were as follows:

- A case of bilateral hydronephrosis with high obstruction of the ureters.
- A case of unilateral hydronephrosis with attraction of the ureter towards the median line.
- A silent kidney on the right with a left ureter attracted towards the median line was found in a single patient.

3.3. Histological Study

Different techniques were used to obtain histological evidence of fibrosis in 15 patients (75% of cases). The most frequent was the radio-guided percutaneous route, performed in 10 cases (50%).

Laparoscopic biopsy was only performed in 5 cases (25%).

In all 15 cases, histological examination of the fibrosis plate showed that it was a dense fibrous tissue rich in fibroblasts and congestive vessels, suggesting non-specific fibro-inflammatory changes.

3.4. Etiologies

RPF was idiopathic in 14 patients (70% of cases). The secondary forms were present in 6 patients (30% of cases) distributed as follows:

- Three cases of RPF secondary to atherosclerotic disease. These patients had several cardiovascular risk factors; In addition to advanced age the three patients had chronic smoking and high blood pressure, two patients had diabetes and one patient had a history of atheromatous disease.
- A case of malignant retroperitoneal fibrosis secondary to a gastric adenocarcinoma.
- A case of systemic fibrosis associating RPF, Riedel thyroiditis and liver fibrosis.
- One case of RPF secondary to Riedel thyroiditis.

3.5. Treatment and Evolution

Initial medical treatment included corticosteroids in 19 patients (95% of cases). Two cases had a combination of corticosteroid therapy with anticoagulants including one for right lower limb thrombophlebitis and the other for IVC thrombosis.

Due to corticodependence, a second line treatment was proposed in a patient by Azathioprine.

17 patients (85% of the cases) had undergone surgical treatment ranging from endoscopic or percutaneous drainage to nephrectomy.

16 patients had double-J ureteral drainage (80% of cases): 12 bilateral and 4 unilateral. Only one patient received bilateral percutaneous nephrostomy under ultrasound scanning and local anesthesia after failure of the rising of the double-J probe. Another patient has benefited of a right nephrectomy after confirmation by the intravenous urography and the CT urography of the presence of a dumb right kidney.

After a average follow-up of 6 months and a half, clinical and biological improvement was observed in 18 patients (90% of cases) with disappearance of the pain and improvement of the general state. On the biological level, we observed in these patients an improvement of the inflammatory test and a normalization of the renal function.

A control with CT scan was performed in 9 patients (45% of cases) and showed:

- Stabilization of the fibrous mass in 4 patients (20%).
- Reduction of fibrosis plaque in 5 patients (25%).

No complications secondary to corticosteroid therapy were observed in patients during follow-up.

Two deaths were observed in this series; one of these two patients had developed cervix carcinoma with pleural and peritoneal metastases after 5 years of immunosuppressive therapy (Azathioprine). The second died due to the complications of his gastric neoplasm.

4. Discussion

RPF is a rare disease. Its incidence is estimated at 0.1 to 1.3 per 100,000 inhabitants per year [10, 11]. It occurs predominantly in the 4th, 5th and 6th decades of life. It is very rare after 70 years or before 20 years. However, cases of RPF have been reported in children [12]. The average age of discovery is 53 years with extremes ranging from 14 to 85 years.

In this series, the average age was estimated at 53 years for extremes ranging from 41 years to 74 years, with a predominance of cases in the age group of 50 to 74 years, which is consistent with data from the literature.

The pathogenesis of retroperitoneal fibrosis is not fully elucidated. Several mechanisms have been proposed to explain the evolution of this disease process. Unfortunately there is no animal model that has been developed to further study this disease, thus making understanding of pathogenesis difficult [14, 15].

Nevertheless, it seems that the combination of immunological factors, genetics and factors related to atherosclerosis is responsible for the retroperitoneal inflammatory and fibrous process [13]. It may have a starting point:

a) Endovascular, in the atheroma plate, whose cracking leads to the release of oxidized LDL and ceroids, and a strong local inflammatory reaction, causing fibrosis.
b) Or in the adventitia involving a vasculitis of vasavasarum, at the origin of both atherosclerosis with formation of aneurysm and peripheral perivascular fibrosis.

A rise in blood levels of IgG4 or plasmocyte overexpression of IgG4 has recently been described in RPF [13, 16]. As well as an association between RPF and other diseases with excess IgG4, such as Riedel's thyroiditis, autoimmune pancreatitis and retro-orbital inflammatory

pseudotumors [17].

The possibility of a genetic predisposition was studied by De Luca [18, 19] who showed the presence of the histocompatibility antigen HLA B27 in 44% of the cases. Thus, the existence of cases of familial RPF is in favor of this genetic predisposition [20, 21].

RPF is idiopathic in 70% of cases [4, 19], when no etiology is found. Secondary forms account for only about 30% of cases. It may be secondary to drugs, neoplasias, infections, radiotherapy, trauma or abdominal surgery, retroperitoneal haemorrhage, local or systemic inflammation or vasculitis (Table 2 and 3).

Table 2. *Main etiologies of retroperitoneal fibrosis [4].*

Idiopathic:	**70% of cases**
Secondary:	**30% of cases**
Neoplasia:	• Retroperitoneal tumors(Urinary tract, lymphoma....)
	• Carcinoid tumor
	• Retroperitoneal metastases
	-Lobular infiltrating breast carcinoma ++
	- Prostate carcinoma +++
	- Gastric carcinoma, colon
	-Cervix Cancer
Retroperitoneal trauma:	
	• Hemorrhage
	• Regional enteritis
	• Perforated Diverticulitis
	• Appendicitis
	• Urinary Extravasation
	• Irradiation
	• Surgery
	• Iatrogenic
Infectious agents:	
	• Urogenital tract infection
	• Histoplasmosis
	• Tuberculosis
Drugs:	
	• Methysergide
	• Ergotamine
	• Methyldopa
	• Hydralazine
	• Beta blockers
Others:	
	• Vasculitis
	• Other systemic diseases
	• Panniculitis of Weber-Christian
	• Mesenteric panniculitis
	• Exposure to asbestos
	• IgG4-related disease

Table 3. *Distribution of RPF in the literature.*

	Ormond 1948	**Koep et Zuidema 1977**	**Wagenknecht et Hardy 1981**	**BAKER 1988**	**LEGUYADER 1992**	**ERIC ET VAN BOMMEL 2007**	**Garrostre 2012**	**LUGOSI et SACRE 2013**
	n =491 [5]	**n =481 [23]**	**n = 430 [25]**	**n =60 [22]**	**n=46 [24]**	**n=24 [26]**	**n=14 [27]**	**n=18 [10]**
Idiopathic	67,80 %	69 %	43 %	86,60 %	67 %	60 %	78,50 %	39 %
Medication	12,40 %	12 %	3 %	6,70 %	10 %	_____	_____	_____
Malignant	7,90 %	11 %	11 %	6,70 %	14 %	_____	14,30 %	_____
Post-traumatic	_____	8 %	15,50 %	_____	4,50 %	_____	_____	11,10 %
Inflammatory or Infectious Diseases	1,20 %	_____	7 %	_____	4,50 %	20 %	7,20 %	16,7 %
Aneurysms of the abdominal aorta	1,80 %	_____	8,5 %	_____	_____	20 %	_____	33,20 %
Post Radics	_____	_____	12 %	_____	_____	_____	_____	_____

(N = number of cases of fibrosis reported in the series).

The results of this study concerning the etiological distribution of RPF remain very close to those reported in the literature.

The progression of fibrosis is slow. Thus, in the absence of compression of adjacent structures, some RPF remain asymptomatic and are discovered accidentally during a prescribed imaging examination for another reason [31].

The symptomatology of RPF is usually insidious, non-specific [28, 29,30], and highly variable; Pain is the most frequently found sign (80 to 92% of cases). Frequent gastrointestinal disorders usually reflect renal insufficiency, but they can exist even outside it and are then explained by the progressive invasion of the autonomic nervous system of the digestive tract and its lymph nodes. Urinary disorders may direct the diagnosis towards a pathology of the lower urinary tract. Vascular disorders type intermittent arterial claudications of the lower limbs by compressing the iliac arteries and the unilateral or bilateral varicocele by spermatic venous compression, frequent during the RPF [28].

The results of this clinical study remain very close to those described in the literature.

Table 4 compares the frequency of clinical signs of RPF in this series and in the literature.

Table 4. *Clinical signs of RPF in this series and in the literature.*

	Lepor	**Baker**	**Le Guyader**	**Deluca**	**Vivas**	**Kaaroud**	**Garroustre**	**Lugosi**	**Our series**
Clinical study	**1979 [32]**	**1988 [22]**	**1992 [24]**	**1998 [18]**	**2000 [33]**	**2005 [28]**	**2012 [27]**	**2013 [10]**	
Number of patients	70	60	31	13	30	15	14	18	20
Pain	81%	68%	71%	85%	100%	100%	78.5%	93%	90%
Slimming	39%	38%	29%	15%	------	60%	42.8%	55.5%	40%

	Lepor	Baker	Le Guyader	Deluca	Vivas	Kaaroud	Garroustre	Lugosi	Our series
Clinical study	**1979 [32]**	**1988 [22]**	**1992 [24]**	**1998 [18]**	**2000 [33]**	**2005 [28]**	**2012 [27]**	**2013 [10]**	
Oligo-anuria	10%	16%	19%	15%	------	6.7%	------	66.6%	10%
Hematuria	------	2%	------	------	3%	------	------	------	5%
Hypertension	47%	------	45%	31%	------	33%	------	------	30%
Edema of the lower limbs	9%	10%	32%	15%	------	6.7%	21.4%	16.6%	15%

Imaging is currently emerging as an essential element in the diagnosis of RPF. In contrast to the historical intravenous urography and abdominal ultrasound with low sensitivity and specificity. The scanner and the MRI make it possible to make a diagnosis in a reliable way. Although histology remains the "absolute" examination for positive and differential diagnosis, biopsies are currently used only to eliminate specific secondary forms (neoplastic or infectious). Indeed CT and MRI allow the diagnosis, the evaluation of the obstructive complications and the appreciation of the evolution. The use of one or the other will depend on the availability, the irradiation and the iatrogenic risk associated with the injection of iodinated contrast agent [4, 34,35].

Functional imaging (gallium scintigraphy and 18-FDG PET) seems useful in the initial assessment. Although the fixation does not systematically cross morphological data visualized by the scanner and the MRI, nor the presence of a biological inflammatory syndrome. Functional imaging allows evaluation of the extent of systemic fibrosis, vascular and perivascular lesions [10, 27, 36].

The treatment of PFR is primarily an etiological treatment (chemotherapy, surgery, antibiotic, stop of the drug in question...).

In idiopathic forms, analysis of the literature shows that treatment remains empirical. There are no publications of a sufficient level of evidence to define the optimal therapeutic strategy for the use of surgery, corticosteroids, immunosuppressants and tamoxifen.

Therapeutic habits, mostly not validated by prospective studies, are based on corticosteroids associated with drainage of the urinary tract.

The use of corticosteroids in the treatment of RPF is important through the results and benefits that it brings. The general therapeutic regimen includes a dose of 0.5 to 1.0 mg / kg / day of prednisone for two to four weeks followed by a gradual decrease over two to three months to 10 mg / d (maintenance treatment). The rapid decrease of doses would allow a balance between iatrogenic complications and fibrosis control. In general, treatment is continued for six to twelve months, but may extend for up to two years [4, 38, 39].

Resistances and corticosteroid addictions although less frequent exist. In these forms, the combination of immunosuppressants such as azathioprine, mycophenolate mofetil, methotrexate or cyclophosphamide has been proposed with some success [4, 40, 41].

Some authors have used Tamoxifene for its antifibrotic properties in the treatment of RPF and have demonstrated the clinical benefit of this molecule in addition to its good tolerance and the absence of major adverse effects [4, 42, 43].

Surgery, the historical treatment of RPF, has become an adjuvant treatment, since the use of corticosteroids. It allows the management of obstructive complications including urological, usually endoluminal (Installation of double-J probe) or percutaneous (nephrostomy). More complex surgical management, such as the classic ureterolysis with omentoplasty, is now performed exceptionally in cases of persistence or aggravation of obstruction despite medical treatment [44].

Patients with RPF associated with an abdominal aortic aneurysm should be considered separately. Their treatment is initially based on the placement of a vascular prosthesis, associated in case of persistence of fibrous plate to corticosteroid therapy [4].

The evolution of RPF is unpredictable. In the absence of treatment, most of the time fibrosis progresses progressively. When the RPF is treated in a timely manner, its evolution is usually favorable. However, relapse is possible when the corticosteroid treatment is discontinued.

The prognosis for RPF is generally good [1], with a ten-year survival rate of more than 70% [45, 46, 47]. Most deaths are attributed to neoplastic etiologies and atherosclerotic complications rather than to RPF.

Late recurrences are possible and unpredictable and can occur within three months to more than 9 years regardless of the treatment, and are more frequent in the first 5 years thus requiring close and prolonged clinical, biological and radiological monitoring [25, 31, 37, 48].

5. Conclusion

This study provides descriptive evidence for a series of 20 patients with retroperitoneal fibrosis. The general characteristics of the patients included are similar to the other series of the literature.

The clinical manifestations of the RPF are very variable, but not specific, often misleading the diagnosis.

Computed tomography and magnetic resonance imaging (MRI) remain the means of reference exploration for the diagnosis of the disease. Retroperitoneal fibrosis is in most cases idiopathic. The secondary forms represent only about 30% of cases dominated by abdominal aortic aneurysm and neoplasia. This frequency appears to be underestimated, Hence the interest of a rigorous etiological investigation. The treatment should be adapted to the etiology and mainly involves corticosteroid therapy and drainage of the urinary tract. The use of immunosuppression or more recently Tamoxifen may in some situations be necessary. The prognosis is generally good, however, with the possibility of recurrences whose incidence is greater during the first five years requiring prolonged clinical, biological and

radiological monitoring.

These results deserve to be confirmed by a prospective multicenter study involving a higher number of patients. The therapeutic strategy and in particular the use of the cortisone saving treatments should be specified by randomized therapeutic trials.

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