Generalities About Idiopathic Retro Peritoneal Fibrosis: A Case Series and Review from the Literature

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Abstract

Retroperitoneal fibrosis is a rare disease that is often discovered late, and its most frequent complication is a non-functional kidney. Intravenous urography and ultrasound are essential for the diagnosis of complications. CT and MRI are the key examinations for a positive diagnosis.

The objectives of our study are knowing the radiological aspects of idiopathic retroperitoneal fibrosis, Learning how to identify the principal signs of retroperitoneal fibrosis, Search for the main associated signs that can indicate a multi-systemic pathology.

Keywords: Traumatic optic neuropathy • Visual evoked potential • Traumatic brain injury • Concussion • Disability

Introduction

Retroperitoneal fibrosis i s a rare d isease o ften discovered l ate, its most frequent complication is the non-functional kidney.

Studying radiological aspects of idiopathic retroperitoneal fibrosis, and signs allowing its positive diagnosis. The main associated signs that may point to a multisystemic pathology such as fibrosis

Our series of eight patients focus on multiple forms of retroperitoneal fibrosis, their clinical features, and complications, to bring the clinician closer to thinking about this complication.

Methods

This study was carried out in the regional university surgical and medical imaging department with approximately 15,000 consultations per year. It is a retrospective study based on the analysis of eight cases of patients who had their radiological check-ups at the radiology department of the Hassan II University Hospital in Fez. The average age of discovery was 53 years with a female predominance. All patients who received a medical visit by a surgeon were identified by a review of visit records using an electronic patient medical record system (SIVSA/Hosix.NET, Core).

All subjects between January 2018 and July 2020 were included in the study with their written and informed consent. The study was approved by the respective local ethics committee, which for the most part complied with national regulations and the standards of the National Institute of Health.

The retrospective examination of the files was conducted by two authors (CY, KS). The observations of the surgical intern and the senior urology surgeon caring for the patient were analyzed for significant signs of lumbar pain, vomiting, or alterations in general condition, laboratory results, and ultrasound findings. Our eight patients with confirmed retroperitoneal fibrosis were examined between January 2018 and July 2020. These cases were confirmed by an abdominal CT scan after 24 hours of hospitalization on average.

The CT scans were performed using a 16-section scanning system (GE medical system). The acquisitions were made in a supine position at the end of inspiration. The scanner parameters were as follows: 5mm section thickness with 1.25 mm reconstruction; tube voltage 120kV; tube current 100 mAs -200 mAs; collimation 3 mm; pitch 1-1.5; matrix size 512x 512, FOV 35 cm. 2/3 of our patients were injected with an iodinated contrast agent.

All CT images were sent to the post-treatment workstation, reviewed by two radiologists, and then approved by a qualified radiology specialist with approximately 7 years of experience.

A nephrological survey of the patients was done after 03 months regarding the functional complications. The data collected was compiled in an electronic database (microsoft excel for windows, microsoft corp., Redmond, WA), the mean values of the numerical elements were calculated and the data were evaluated.

Result

Concerning demographic and clinical characteristics, of the 8 patients included in the current study, 3 were men and 5 were women. The median age of the patients was 53 years, and half of the patients haven't had a history of illness or drug use.

Clinically, ureteral colic was the most common with 6 patients, mostly localized to the right flank (in), left flank (2), or bilateral (2). Most patients, except two, were non-febrile, with only one patient presenting with a pseudo-surgical abdomen.

Biological tests reveal that most cases (6) had a renal failure, with a

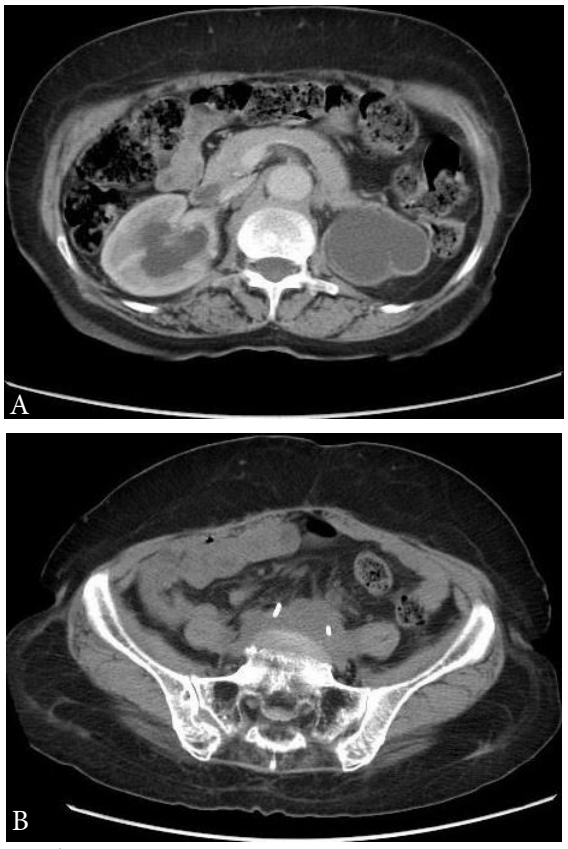

Figure 1. 1a) Contrast CT scan shows a severe bilateral ureteral obstruction proximal to a retractile fibrous mass **1b)** Placement of a double J catheter (arrow)

variable severity rate among patients; inflammatory results show CRP was averaging 25 mg/Dl.

Diagnostic findings on clinical and radiological examination were unilateral or bilateral ureteral obstruction and renal artery stenosis (Figures 1 and 2).

All of these patients underwent an abdominal ultrasound examination. The most specific aspect was found in four of our patients and revealed infiltration of the retroperitoneum leading to bilateral ureteral obstruction.

In addition, we found other signs of accompaniment, such as lymphadenopathies, atherosclerosis and aortitis.

Our patients benefited from an abdominal CT end or MRI which was specific in all cases and was able to reveal complicated forms such as a non-functional kidney.

The first line of therapy was medical and we used glucocorticoids in all cases. It was followed by double J stent placement (6 cases) or nephrostomy (2 cases).

Treatment using Tamoxifen was also proposed in one patient who experienced steroidal toxicity After 12 months of follow-up, four patients had developed chronic renal failure due to obstructive uropathy (Table 1).

Discussion

Idiopathic Retroperitoneal Fibrosis (RPF), is a rare fibro-inflammatory disease that develops around the abdominal aorta and the iliac arteries and spreads into the adjacent retroperitoneum, where it frequently causes ureteral obstruction and renal failure. The clinical phenotype of RPF is complex, because it can be associated with fibro-inflammatory disorders involving other organs, is considered part of the spectrum of IgG4-related disease, and often arises in patients with other autoimmune conditions [1].

Clinically Systemic symptoms (e.g., fatigue, anorexia, weight loss), and possible expression of inflammatory status, often herald the disease onset (Table 1). They usually coexist with back, flank, or abdominal pain. Pain is generally dull, does not modify with position, and transiently responds to nonsteroidal anti-inflammatory drugs; in cases of ureteral involvement, it

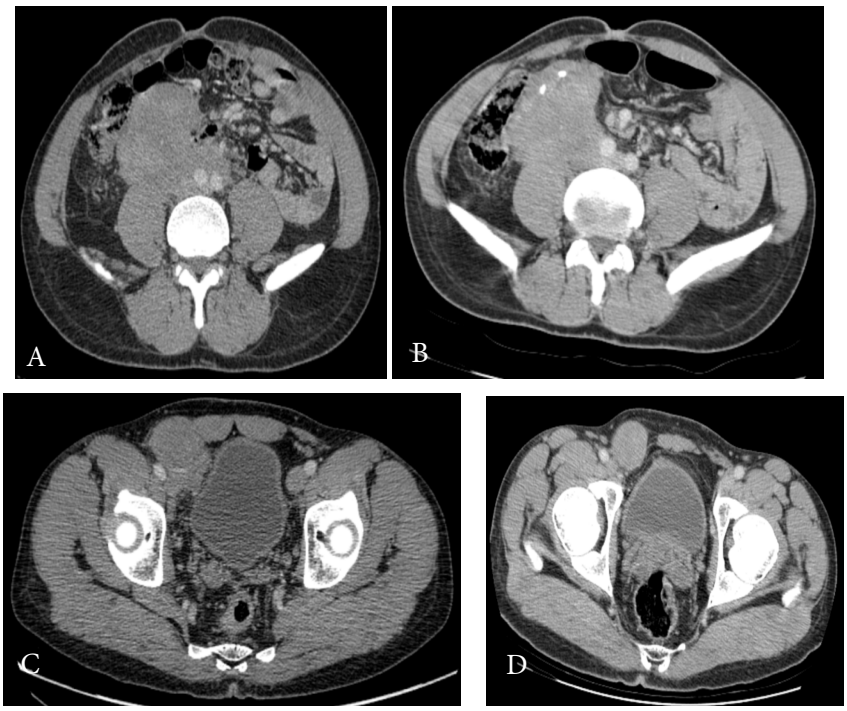

Figure 2. 2a,b) Contrast CT scan shows a retro and intra peritoneal fibrous mass which is enhanced after contrast **2c,d)** The mass is extending along the spermatic canal.

Table 1. Clinical data of 8 patients with idiopathic retroperitoneal fibrosis.

Patient Nor	1	2	3	4	5	6	7	8
Sex Age Temperature	Women 39 37°	Women 58 38	Male 55 38,5	Male 38 36,5°	Women 67 37°	Women 60 37°	Male 52 37,5°	Women 50 37°
OMS Scale	2	1	4	3	4	2	1	2
Radiologic Diagnosis CT-Scan	Unilateral ureteral obstruction	Unilateral ureteral obstruction	Bilateral ureteral obstruction	Bilateral ureteral obstruction	Bilateral ureteral obstruction	Unilateral ureteral obstruction	Unilateral ureteral obstruction	Unilateral ureteral obstruction
Radiologic Duration of Pain Diagnosis (days)	+ Renal artery stenosis 7	11	20	15	+ Renal arteryste nosis 8	7	6	10
First symptoms to Note	Right lumbar pain Fever	Left lumbar Pain	Advanced Rena Failure	Bilateral lumbar Pain	Advanced Renal Failure	Right lumbar pain	Right lumbar pain	Left lumbar pain
Medical Treatment	Glucocorticoïds	Glucocorticoïds	Glucocorticoïds	Glucocorticoïds	Glucocorticoïds	Glucocorticoïds	Glucocorticoïds	Glucocorticoïds
Surgical Treatment	Double J stent	Double J stent	Double J stent + Nephrostomy	Double J stent	Double J stent + Nephrostomy	Double J stent	Double J stent	Double J stent
Follow up Duration and Outcomes	5 years Stability	2 years Stability	3 years Worsening of renal failure	2 years Stability	6 years Worsening of renal failure	3 Years Stability	1 Year Stability	2 Years Stability

may mimic ureteral colic. Constipation may be another dis- ease-related manifestation, although it is rarely severe. Other urologic manifestations are frequent: they range from testicular pain, often accompanied by hydrocele and/or varicocele due to spermatic vein encasement by RPF– to retrograde ejaculation and erectile dysfunction [2-4] (Figure 2). Other less common manifestations include frequency, hematuria, and dysuria [5,6].

In pathophysiology, its association with autoimmune disorders highlights the pathogenic relevance of autoimmune mechanisms. Autoimmune thyroiditis is the most frequently associated autoimmune condition: in a recent case-control study, idiopathic RPF patients had a prevalence of anti-thyroperoxidase antibodies of 24.7% (versus 10.6% in healthy controls) and ultrasound evidence of thyroiditis; after a median follow-up of 45 months, 25% of RPF patients developed hypothyroidism requiring Lthyroxine. Where available, histology showed typical Hashimoto thyroiditis or its fibrous variant. However, cases of Riedel thyroiditis were also described [7,8].

Other associations include rheumatoid arthritis, ankylosing spondylitis, ANCA-associated vasculitis, systemic lupus erythematosus, and psoriasis [9,10].

The positive diagnosis of idiopathic RPI is made by Computed Tomography (CT) or Magnetic Resonance Imaging (MRI). On CT, it appears as a homogeneous plaque surrounding the anterolateral sides of the abdominal aorta and encircling the common iliac arteries. Medial ureteral deviation and/or obstruction and encasement of the inferior vena cava are common. The tissue is more isodense than that of muscle with varying degrees of contrast enhancement (Figure 1).

The intensity of fibrosis is low on T1-weighted images and variable (high in active stages) on T2-weighted images. Contrast enhancement and diffusion coefficient values are useful in differentiating active and inactive lesions.

Fluorodeoxyglucose (18FFDG) Positron Emission Tomography (PET) is a helpful tool for the assessment of RPF activity; this technique also detects the metabolic activity of post-treatment residual disease and thus guides subsequent therapy. However, 18 FDG PET has little diagnosis utility because many infectious, inflammatory, or neoplastic lesions also accumulate 18FFDG.

In general, treatment immunosuppressants have been used in combination with glucocorticoids; however, it remains unclear whether they potentiate the efficacy of glucocorticoids or function as steroid-sparing agents. Mycophenolate mofetil is widely used, in part because of its good tolerability and lack of contraindications in patients with renal failure [11-13]. Cyclophosphamide has also been used effectively as initial therapy followed by maintenance with other immunosuppressants but is not currently recommended as first-line therapy [14,15].

Patients who achieve remission should be carefully monitored with laboratory tests, periodic ultrasound (to monitor hydronephrosis and aneurysmal dilatation), and CT/MRI studies (capable of accurately defining the size and morphological changes of the RPI) to allow for early detection of relapse. Long-term maintenance therapy should be considered, especially in patients with aggressive disease. Idiopathic RPI is indeed a chronic relapsing disease, with relapse rates of up to 72%. Importantly, patients who relapse often experience multiple relapses and are therefore exposed to high cumulative doses of glucocorticoids. In such cases, methotrexate has been used successfully as a steroid-sparing agent.

Conclusion

In conclusion, ultrasound is the paraclinical imaging examination that is often performed first. As for CT scan and MRI, they remain the reference means of exploration for the diagnosis of the disease. Monitoring the evolution of the fibrous plaque is done by CT. The etiological diagnosis is based on histology.

The treatment is usually palliative, surgical intervention helped by medical treatment is necessary to stabilize the evolution of this process.

References

1. Vaglio, A., & Federica M.. "Idiopathic retroperitoneal fibrosis." *J Am Soc Nephrol.* 27.7 (2016): 1880-1889.
2. Van Bommel, E. F. H., et al. "Idiopathic retroperitoneal fibrosis: prospective evaluation of incidence and clinicoradiologic presentation." *Medicine* 88.4 (2009): 193-201.
3. Kermani, T. A., et al. "Idiopathic retroperitoneal fibrosis: a retrospective review of clinical presentation, treatment, and outcomes." *Mayo Clin Proc* Vol. 86. No. 4. Elsevier, 2011.
4. Vaglio, A., et al. "Prednisone versus tamoxifen in patients with idiopathic retroperitoneal fibrosis: an open-label randomised controlled trial." *The Lancet* 378.9788 (2011): 338-346.
5. Scheel J., P. J., & Nancy F. "Retroperitoneal fibrosis: the clinical, laboratory, and radiographic presentation." *Medicine* 88.4 (2009): 202-207.
6. Ceresini, G., et al. "Association between idiopathic retroperitoneal fibrosis and autoimmune thyroiditis: a case–control study." *Autoimmun* rev 14.1 (2015): 16-22.
7. Fatourechi, M.M., et al. "Invasive fibrous thyroiditis (Riedel thyroiditis): the Mayo Clinic experience, 1976–2008." *Thyroid* 21.7 (2011): 765-772.
8. Afeltra, A., et al. "Retroperitoneal fibrosis and ankylosing spondylitis: which links?" *Semin arthritis rheum* Vol. 35. No. 1. WB Saunders, 2005.
9. Vaglio, A., et al. "ANCApositive periaortic vasculitis: does it fall within the spectrum of vasculitis?." *J intern med* 251.3 (2002): 268-271.
10. Vaglio, A., et al. "18Ffluorodeoxyglucose positron emission tomography in the diagnosis and followup of idiopathic retroperitoneal fibrosis." *Arthritis Care Res* 53.1 (2005): 122-125.
11. Marcolongo, R., et al. "Immunosuppressive therapy for idiopathic retroperitoneal fibrosis: a retrospective analysis of 26 cases." *Am j med* 116.3 (2004): 194-197.
12. Jois, R. N., et al. "Mycophenolate mofetil for maintenance of remission in idiopathic retroperitoneal fibrosis." *Rheumatology* 46.4 (2007): 717-718.
13. Scheel Jr, Paul J., et al. "Combined prednisone and mycophenolate mofetil treatment for retroperitoneal fibrosis: a case series." *Ann. intern. med*. 154.1 (2011): 31-36.
14. Binder, M., et al. "Cyclophosphamide is a highly effective and safe induction therapy in chronic periaortitis: a long-term follow-up of 35 patients with chronic periaortitis." *Ann Rheum dis* 71.2 (2012): 311-312.
15. Alberici, F., et al. "Methotrexate plus prednisone in patients with relapsing idiopathic retroperitoneal fibrosis." *Ann rheum dis* 72.9 (2013): 1584-1586.

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