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Renal disease in a retroperitoneal fibrosis

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ABSTRACT

Idiopathic retroperitoneal fibrosis (IRF) is characterized by the extensive development of inflammatory fibrotic tissue in the retroperitoneum, leading to the compression and obstruction of the ureters and other adjacent organs. Several observations suggest that the immune system has an important role in the pathogenesis of IRF. Steroids are normally used to treat idiopathic retroperitoneal fibrosis, several studies have demonstrated an excellent benefit of immunosuppressive treatment with and without surgical intervention. However, the optimal immunosuppressive agent, the length of treatment needed, and the best approach to monitoring disease activity have not been analysed. We report a case of IRF associated to bilateral ureterohydronephrosis and chronic renal insufficiency, the diagnosis was based on the typical clinical and biological presentation; with high Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) computed tomography (CT), magnetic resonance imaging (MRI) of the abdomen, showed typically a fibrotic mass and retroperitoneal histological findings confirms the diagnosis. Treatment consisted of left nephrectomy (because of pyonephrosis) with mass biopsy for histological analysis, right double-J ureteral stenting. The patient received oral prednisone at a dose of 1 to 1 mg/kg/d for 3 weeks, and then tapered until discontinuation within 12 months. Prednisone was combined with mycophenolate mofetil (2mg/d for 16months). Our patient reported substantial resolution of symptoms after a median treatment duration of 3 weeks. ESR and CRP also decreased. Repeated CT scanning showed slow but steady mass regression and partial improvement then a stabilisation of the renal function. After a follow up of 16 months, we observed a normal ESR and CRP; patient's clinical condition remained good. Side effects from corticosteroids were transient and not clinically important. No malignancies were observed during the follow-up period. Treatment was changed to tamoxifene and prednisone for 18 months, but with no change of the volume of the fibrosis, and a progression renal failure to end stage renal disease. Immunosuppressive treatment in combination with temporary ureteral stenting is a safe and effective treatment. Mycophenolate mofetil with steroids may be a viable therapeutic option in the

treatment of IRF. Response to treatment may be protracted and should be monitored with imaging studies.

INTRODUCTION

Retroperitoneal fibrosis (PIF) or Ormond's disease (1) is a rare, inflammatory and fibrotic disease of the retroperitoneal periaortic tissue, considered one of the causes of obstructive uropathy seen the sheathing of the ureters, and whose pathogenesis is a response systemic the plaque (2). Incidence, largely underestimated, is 1: 200 000 if limited only to clinical manifestations. However, 4.5% of patients undergoing abdominal aortic aneurysm, have a few symptoms periaortic fibrous sleeve (3).

MATERIALS AND METHOD

observation

We report the story of a woman of 55, known hypertensive for 5 years under enzyme inhibitor conversion related pain abdominopelvic and flanks, lasting for two years in spurts, in the context of conservation general condition. Its history goes back to current one month before admission to worsening pain, and fever, without digestive or urinary problems. Clinical examination revealed a patient weighing 60kg, asthenic, afebrile, hypertensive to 150/100 mmHg, with a sensitivity on the left flank, with no signs of systemic disease associated or obvious tumor syndrome. Laboratory tests revealed an inflammatory syndrome with ESR and CRP 82mm to 46mg / l, renal failure with creatinine 27 mg / l, a non-regenerative anemia normochromic 9.5 g / dl, proteinuria 1 g / 24 hours with a sterile urine culture. Renal ultrasound showed a right kidney of normal size with a discrete expansion pyelocaliceal and a left kidney with hydronephrosis pyélonéphritique associated without gallstone picture.

An MRI revealed Uro-tissue formation depends on the external face of the left kidney extending downwardly into contact with the psoas including the ureter. The appearance was suggestive of retroperitoneal fibrosis (Figure 1).

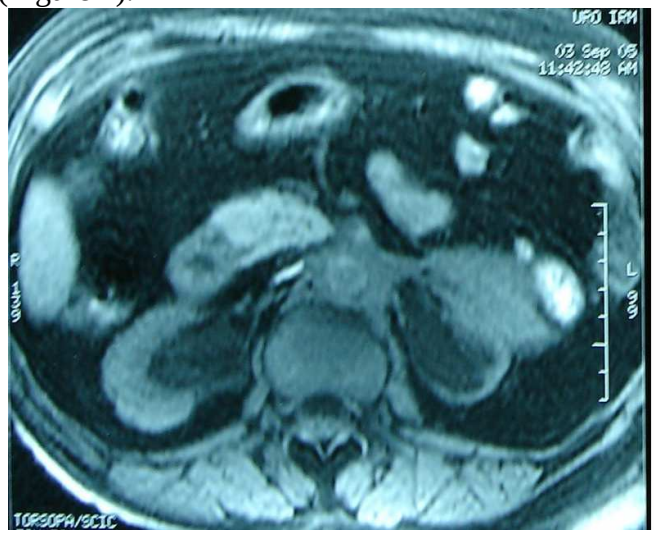

Figure 1: shows a pelvic MRI retroperitoneal fibrosis extended to the outside of the kidney associated with hydronephrosis.

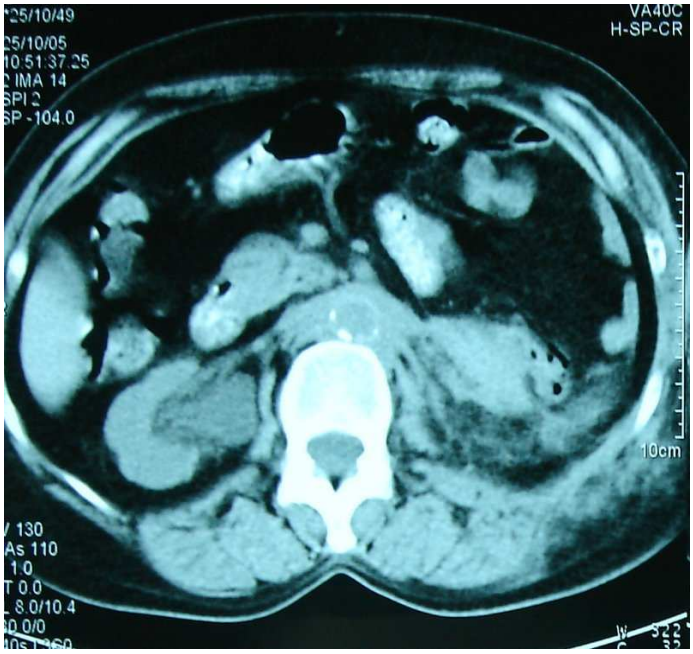

Figure 2 Abdominal CT post chirutgicale response showing the plate fibrosis.

Surgery was decided with a left ureterohydronephrosis discovery, the presence of a fibrous sheath gray adjoining the parietal peritoneum, surrounding the aorta and inferior vena cava, and pushed the left ureter with an extension to the psoas . The damage imposed a left nephrectomy (Figure 2). Pathological examination of the piece of nephrectomy and fibrous formations showed the presence of a non-specific tissue fibrosis associated with chronic pyelonephritis; absence of malignant cells or signs of chronic infection, all evoking a nonspecific retroperitoneal fibrosis. The search for possible secondary causes of this PIF was negative: normal chest radiograph with the BK search by gastric intubation and negative in urine, no concept of drug taking. Immunological investigations were without abnormalities, gynecological examination with bilateral mammography were normal, the thoracoabdominal scanner revealed no images or suspicious lesions, thyroid exploration was normal and infectious investigation. Following this investigation, the diagnosis of idiopathic retroperitoneal fibrosis was retained, allowing the start of immunosuppressive therapy with Mycophenolate Mofetil (CELLCEPT *) at a dose of 1mgx2 / day for nine months and Prednisone 1 mg / kg / day with degression gradually over a total period of 12 months. Rapid resolution of clinical symptoms associated with normalization of inflammatory markers were obtained after four weeks. On a 16-month follow-up, renal function remained stable, and inflammatory markers normalized. The scanner checks carried out after 6 months of starting treatment, and 12 months showed a stabilization of plaque progression without periphery (Figure 3).

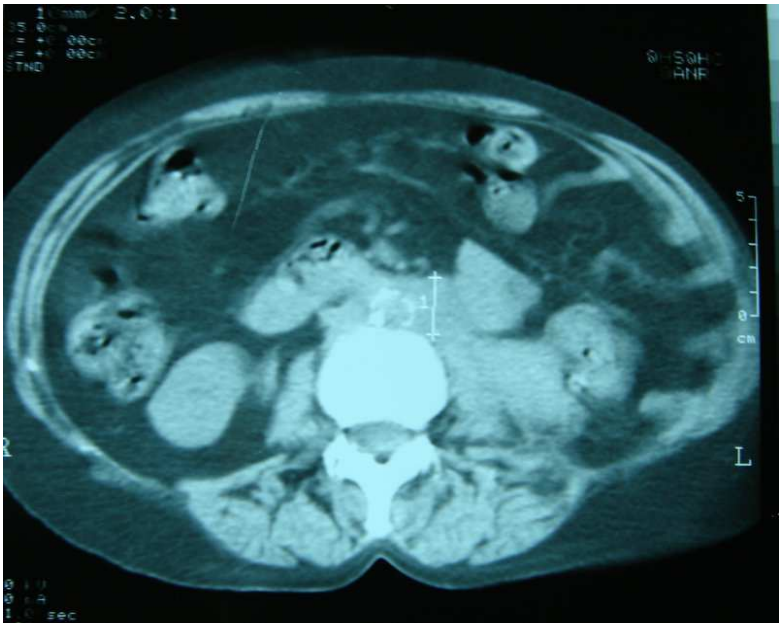

Figure 3: CT scan after 12 months of treatment combined MMF and corticosteroids showing non plaque progression.

The control CT scan showed virtually the same development of fibrosis around the large vessels up to the level of the left iliac region without significantly increasing the process. Biologically however, there was no improvement in renal function. On these data we opted for a change in the treatment and cure of tamoxifen was started at a rate of 20mgx2 / day associated with low corticosteroids: Prednisone 5 mg / day. The audit carried out after 8 months of treatment showed a worsening of renal function with a creatinine 34 mg / l, correction of anemia and inflammatory markers (CRP = 4.3mg / l and VS = 13mm). The control CT scan performed after 8 months showed stable plaque fibrosis, but with a more pronounced expansion ureteropelvic (Figure 4).

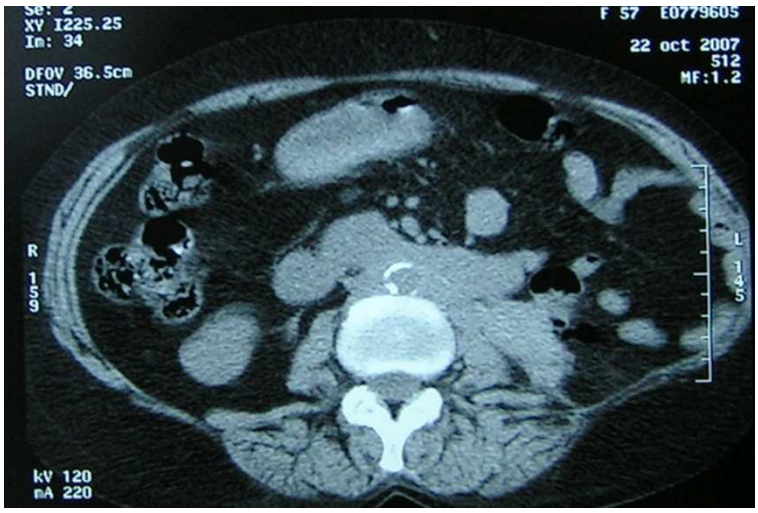

Figure 4: abdominal CT after 8 months of treatment with tamoxifen showing a stable aspect of FRP with greater expansion urétéropyélique

During these eight months of treatment, tamoxifen was well tolerated. Treatment was maintained for 18 months. After this period, renal function continued to deteriorate, and progresses to ESRD requiring starting a replacement therapy by hemodialysis.

RESULTS AND DISCUSSION

We reported the case of a patient with idiopathic retroperitoneal fibrosis, complicated by obstructive IRA, the anti-inflammatory treatment with immunosuppressive helped stabilize the plate with reducing inflammation over 2 years, but progression to ESRD was inevitable. FRP is an entity individualized in 1948 by Ormond, but previously described by Albarran in 1905 under the term peri-urethritis stenosis (3), a prevalence of 2-5 cases / million population (4), it preferentially affects the man with a sex ratio of 2 (5). It is characterized by the sheathing of retroperitoneal structures including the abdominal aorta by a fibro-inflammatory tissue devoid of special features as histology (6). There are twenty years the nosology of chronic périaortites included retroperitoneal fibrosis, inflammation of abdominal aortic aneurysms and peri-aneurysmal retroperitoneal fibrosis. Now these entities must be distinguished and FRP strict sense is the development of a fibro-inflammatory tissue without dilatation of the aorta for retroperitoneal fibrosis, unlike the two other entities (7). It can be isolated or associated with many diseases; In most cases, retroperitoneal fibrosis is primitive. Secondary forms which are many causes should be investigated. They represent, in series, 20% to 30% of cases. The main causes are the causes neoplastic, inflammatory diseases or autoimmune diseases, post radiotherapy and drug causes infections (8).

Pathophysiologically the pathogenesis of idiopathic form is unclear; two main theories (9.10) are now discussed:

1 A local inflammatory overreaction to aortic atherosclerosis, stimulated by oxidized LDL-cholesterol. This mechanism is found predominantly in patients with periaortitis where T and B lymphocytes observed in the media and adventitia, high levels of interleukins in aortic wall sections and circulating antibodies against oxidized LDL. (10)

2. Autoimmune manifestation of systemic disease. This hypothesis is supported by the frequent presence of autoantibodies.

Other potential pathogenetic processes have been reported in some carrying a large FRP with vasculitis patients large vessels, such as the presence of anti-fibroblast and IgG4 produced by plasma cells. (10)

There is probably a genetic component associated with the HLA-DRB1 * 03, which is associated with type I diabetes, systemic lupus erythematosus and myasthenia gravis. (10)

The analysis of the literature over the past 20 years shows that the treatment of idiopathic retroperitoneal fibrosis remains empirical. There are no publications with a sufficient level of evidence to define the optimal therapeutic strategy of using surgery, corticosteroids, immunosuppressants and tamoxifen (11), surgery is to create a urinary diversion in emergency (percutaneous nephrostomy, urethral probes, double probes J) or perform a ureterolysis (12). For symptomatic patients, corticosteroid therapy remains the first-line treatment, but the dose (0.5 to 1 mg / kg / day) and duration are not consensual (13) Other strategies corticosteroid sparing were tested with mycophenolate mofetil, cyclophosphamide and azathioprine (14-15).

CONCLUSION

The diagnosis of FRP purveyor of postrenal renal failure in 75% of cases, should not be ignored,. Treatment consists of surgical symptomatic component to release the bodies of the retroperitoneal fibrotic sleeve, and combined with immunosuppressive and antifibrotic agents to, however in the absence of an effective drug treatment aetiopathologic part by anti-inflammatory a progression to end stage renal disease due to the extension of the plate fibrosis is very common.

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