

Retroperitoneal Fibrosis During Systemic Lupus

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Citation

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Abstract

Retroperitoneal fibrosis is a rare condition. It may be of idiopathic origin or secondary to the taking of certain drugs, to infectious, tumoral or autoimmune pathologies. We report here a case of retroperitoneal fibrosis that occurred with systemic lupus.

A 37-year-old female was seen for a right renal colic evolving for a week associated with a urinary irritative syndrome and a febrile dysenteric syndrome. The patient was followed for the past 4 months in the Dermatology department for systemic lupus. The examination noted diffuse abdominal tenderness, predominantly rhizomelic arthralgia, and chronic lupus lesions in the exposed areas. The biological assessment showed an inflammatory syndrome. Examination of urine on a strip showed the presence of red blood cells and proteins. Abdominal ultrasound showed bilateral dilatation of the pyelocalicielles cavities without individualized barriers. The uroscanner revealed a bilateral pyelo-ureteritis predominant on the right with important perireteral fibrous gangue.

The patient was placed on steroids (1 mg / kg / day of prednisone) associated with adjuvant measures and on hydroxychloroquine (400 mg / day). This treatment made it possible to eliminate the pain after 7 days. The abdominal ultrasound performed 20 days after the beginning of the corticotherapy showed a disappearance of the dilation of the pyelocalicielles cavities.

Conclusion: The occurrence of retroperitoneal fibrosis during systemic lupus is a rarely reported entity. In our observation, the medical treatment based solely on corticosteroids allowed an improvement of the clinical picture.

INTRODUCTION

Retroperitoneal fibrosis (RPF) is a rare disease characterized by the presence of fibro-inflammatory tissue which usually surrounds large vessels and organs in the peritoneum [1,2]. It may be idiopathic or secondary to infection, abdominal surgery, certain drugs for malignant tumors, or inflammatory or autoimmune diseases such as systemic lupus [1, 3,4]. According to our review of the literature, the occurrence of retro-peritoneal fibrosis during systemic lupus is a rarely reported entity [2]. We will describe one case.

CASE REPORT

A 37-year-old woman was hospitalized for a right renal colic evolving for a week associated with urinary irritative syndrome and a febrile dysenteric syndrome. In her antecedents it was noted that the patient was followed for the past 4 months by the Dermatology department for a systemic lupus.

The diagnosis was based on lesions of chronic lupus sitting on the exposed photo-zones associated with symmetric and bilateral inflammatory rhizomelic arthralgia. Laboratory results revealed cytopenia with normochromic anemia at 7.3g / dl and thrombocytopenia at 59000 / mm³. Serum creatinine and proteinuria were normal. In immunology, the anti-nuclear Ab were positive as well as the anti SSA / Ro Ab. Initial corticosteroid therapy with 40 mg / day of prednisone combined with synthetic antimalarials was instituted. However, the treatment was badly followed for lack of means. During the examination, the general condition was altered, the mucous membranes were pale anicteric, the blood pressure was 120/70 mm Hg, the weight was 39 kg, the pulse was 115 beats / min and the temperature was 38 ° C. There was no edema in the lower limbs.

Physical examination revealed:

- Diffuse abdominal tenderness without contracture or defense.
- The upper and middle urethral points were painful.
- The mobilization of large and small joints was painful without tenosynovitis.

The cutaneo Phanerian examination noted:

- Photosensitive erythematous lesions were found on the face in vesperilio, associated with achromic lesions sitting on the face, décolleté and forearms (figure 1) and a silky trichopathy.
- The blood count showed hypochromic normocytic 5.6 g / dl, leukocytosis at 10000 / mm³. The sedimentation rate was accelerated to 120 at the 1st hour and 190 at the 2nd hour.
- CRP was elevated to 12 mg / l. Examination of the urine in the strip showed the presence of red blood cells and proteins.
- There were no leucocytes or nitrite in the urine.

Cytobacteriological examination of urine, parasitological examination of stool and stool culture could not be performed. Because of the febrile dysentery syndrome, ciprofloxacin and metronidazole antibiotics were used to eliminate diarrhea, but the abdominal pain that prompted ultrasonography showed bilateral dilatation of the pyelocalic cavities with no individualized barrier. Uroscanning revealed a predominantly bilateral pyelo-ureteritis with significant periureteral fibrous gangue with moderate bilateral ureteropyelocalic dilatation without endoluminal urinary tract obstruction (Figure 2).

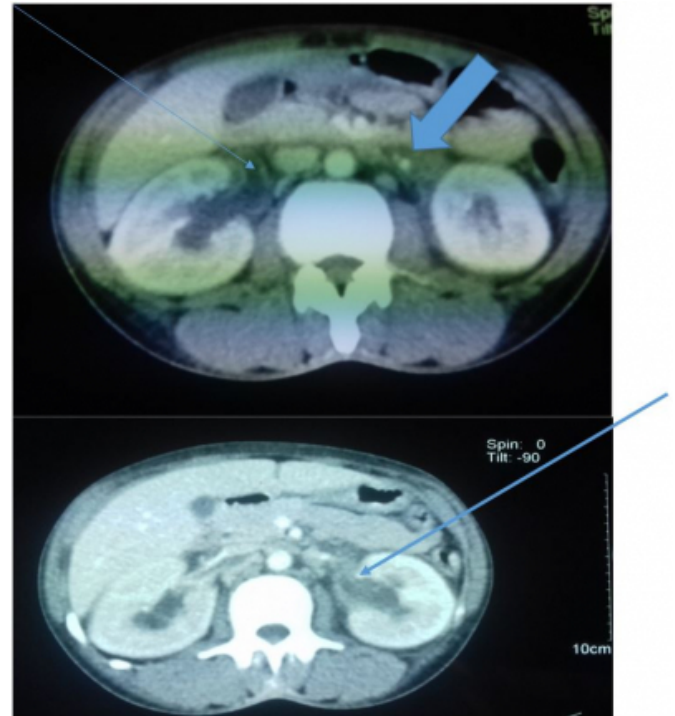
Figure 1

erythematous lesions on the face associated with achromic lesions sitting on the face, décolleté and forearms



Figure 2

bilateral pyelo-ureteritis (blue arrow) with significant periureteral fibrous gangue (full blue arrow) with moderate bilateral ureteropyelocalic dilatation without endoluminal urinary tract obstruction



These results allowed us to diagnose a retro-peritoneal fibrosis. The patient was placed on steroids (1 mg / kg / day of prednisone) associated with adjuvant measures and on hydroxychloroquine (400 mg / day). This treatment made it possible to eliminate the pain after 7 days. Following a transfusion of 02 blood bags, the control count showed a level of Hg at 8.6 g / dl. The abdominal ultrasound performed 20 days after the beginning of the corticotherapy had shown a disappearance of the dilation of the pyelocalic cavities.

DISCUSSION

Retroperitoneal fibrosis is a rare condition. This disease was described for the first time by Albarran in 1905, but Ormond's description of two cases in 1948 made it a clinical entity [5,6]. Epidemiological data on this disease are not well established and only data on idiopathic forms are available. A Finnish study showed an incidence of 0.1 / 100,000 inhabitants per year and a prevalence of 1.4 / 100,000 inhabitants, while a more recent analysis in the Netherlands revealed a higher incidence of 1, 3 / 100,000 inhabitants / year [8].

The age range of onset is 40 to 60 years and there is a male

predominance [3, 8, 9, 14]. Retro-peritoneal fibrosis secondary to systemic lupus is an entity rarely reported in the literature [2, 7, 10]. In our observation it was a 37-year-old woman. This reveals a relatively young onset of age relative to the age group of onset according to literature data [2,3]. In addition, cases have been described in young patients, all 17 years old [8,10]. Several theories have been put forward to explain the pathogenesis of FRP.

A wide range of factors can cause secondary forms of FRP, including drugs, infections, external radiation and malignant tumors [1, 3, 8, 11]. An early theory, proposed in the mid-1980s by Parums and Mitchell, defined idiopathic RPF as a complication of advanced atherosclerosis of the aorta. However, patients with FRP often have systemic symptoms, high levels of markers of inflammation, positive autoantibodies, associated autoimmune diseases, involvement of other organs, and a good response to immunosuppressive therapies.

These results suggest that FRP is a manifestation of a systemic disease rather than an exaggerated local reaction to atherosclerosis [1, 12, 11]. This theory, in the absence of other risk factors, could be retained for our case. Recently, some authors have hypothesized that idiopathic RPF may be related to a systemic inflammatory disease known as IgG4 disease [1,12]. The clinical presentation of FRP is highly variable and often related to the mechanical effect of FRP on nearby structures:

- Abdominal pain
- Lower back pain
- Lower limb edema
- Testicular pain
- Hydrocele
- Intermittent limb claudication
- Renal colic are the clinical signs most frequently reported [3, 9,13].

The latter was the main clinical manifestation in our patient. On a biological level, classically during a FRP, there is an increase in markers of inflammation associated with inflammatory anemia and an impairment of renal function [1, 3, 8]. This was the case in our observation. Anti-nuclear antibodies (ANA) are positive in 5 to 60% of patients with FRP but other autoantibodies may also be positive, including rheumatoid factor, cytoplasmic anti-neutrophil antibodies, anti-smooth muscle cells and antithyroid antibodies. The presence of these autoantibodies, although specific to the organ and often positive in low titers, may suggest an autoimmune origin of the disease [1,8].

In our patient, the antinuclear antibodies were positive as

well as in the observation of Hu et al [2]. To confirm the diagnosis of FRP, CT and MRI are the exams of choice. They make it possible to highlight the fibrosis plaque, to specify its morphology, its location and its propagation to neighboring structures [1, 8, 9]. Biopsy is not always carried out, especially if the radiological characteristics are in favor of an RPF. However, if diagnostic dilemmas exist and an underlying malignancy is suspected or there is no response to initial treatment, the biopsy must be performed for the diagnosis to be confirmed [1].

However, according to the study by Khanc et al, in the absence of clinical or paraclinical guidance elements, the systematic realization of noninvasive (autoimmune markers, tumor markers and bone scintigraphy) or invasive (biopsy of fibrosis and osteomedullary biopsy) does not seem relevant because no cause and / or associated disease is found in the majority of cases of retroperitoneal fibrosis when the radiological presentation is typical [15]. The treatment of FRP was essentially based on surgery. In recent years, management has shifted from an essentially surgical approach to immunosuppressive therapy to modulate the immune system [1, 9, 16].

Our patient benefited from a corticosteroid treatment, which allowed a good evolution of the symptomatology. However, Heidenreich et al suggest that the combination of immunosuppressive therapy and surgical management results in an excellent long-term outcome in idiopathic retroperitoneal fibrosis with a recurrence rate of only 8%. Thus, combined therapy should be considered a therapeutic option in the absence of initial improvement under medical treatment [17].

CONCLUSION

Retroperitoneal fibrosis secondary to systemic lupus is an entity rarely reported in the literature. The diagnosis can be easily confirmed by computed tomography. If the evolution under medical treatment alone was favorable in our patient, it remains that the surgery constitutes a good alternative especially in case of failure of the medical treatment.

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