Abstract

The term retroperitoneal fibrosis (Ormond’s disease) encompasses a group of diseases characterized by fibrosis and inflammation of the tissue in the retroperitoneal space, which may surround abdominal and pelvic structures, including the aorta, iliac arteries, and ureters. The origin is idiopathic in about 70% of the cases.

There is controversy regarding the best treatment and management for idiopathic retroperitoneal fibrosis (IRPF), and clinical strategies vary widely, from treatment with corticosteroids and immunosuppressant’s to surgery. We present the case of a patient with IRPF associated with bilateral ureteral obstruction and periaortitis, receiving treatment with tamoxifen.

Keywords: Tamoxifen; Retroperitoneal fibrosis

Case Report

Man of 70 years age, referred to rheumatology outpatient services in November 2009 for chronic lumbago (low back pain) over the previous three months, radiating to both flanks and impeding sleep. Relevant medical history included hypertension, non-insulin-dependent diabetes mellitus, and dyslipidaemia. Complementary clinical examination revealed a notable increase in acute phase reactants erythrocyte sedimentation rate (ESR): 70 mm/h; C-reactive protein [CRP]: 17.4 mg/dL), and thoracic computed tomography (CT) results showed retroperitoneal soft tissue invasion of the periaortie space, iliac arteries, and ureters. A positon emission tomography (PET) with fludeoxyglucose-18F (18F-FDG) completed the study, characterizing the findings as active retroperitoneal fibrosis [1].

Initial treatment was prednisone plus methotrexate with subsequent introduction of mycophenolate; although there was a small clinical improvement, radiographic follow-up showed progression of lesions and continued elevation of acute phase reactants. During clinical evolution, the patient experienced an episode of obstructive acute kidney failure due to bilateral compression of the ureters, prompting the insertion of a bilateral double J stent to control the condition. Upon detecting further disease progression, in 2011 treating physicians substituted mycophenalate with tamoxifen, observing clinical and radiological improvement, with normalisation of ESR and CRP [2].

In 2014, tamoxifen treatment was suspended in order to intervene with bilateral ureterolysis; subsequent clinical recurrence and elevation of acute phase reactants led to resuming tamoxifen therapy with gradually decreasing dosis of corticosteroids. From that time until the most recent follow-up in October 2015, the patient has been clinically, analytically, and radiographically stable, with tamoxifen 20 mg daily and no associated corticosteroids [3].

A rare disease, IRPF most frequently affects men aged 40-60. The physiopathological mechanisms of the disease are unknown, although investigators have proposed various possibilities, ranging from the existence of a local reaction mediated by cytokines against the atherosclerotic arterial wall, to secondary reactions stemming from the attack of plasma cells producing IgG-4 in the context of systemic disease.

Clinically, IRPF is characterised by low back pain radiating to both flanks and intermittent claudication of lower limbs; advanced stages are associated with acute renal failure secondary to bilateral ureteral obstruction. Increased levels of acute phase reactants such as ESR and CRP are common, as are positive antinuclear antibody results (in up to 60% of cases). Diagnosis is traditionally performed via CT of the abdomen and pelvis, together with biopsy, although some authors do not consider the latter to be necessary. However, to assess metabolic activity it is preferable to run a PET scan with 18F-FDG [4].

With regard to the best disease treatment and management, there is still considerable debate, as there are no clinical trials that support a specific treatment. The most common first-line therapy consists of glucocorticosteroids for six months, followed by immunosuppressants such as mycophenolate and methotrexate. In refractory cases such as ours, a few series describe tamoxifen as efficacious. The mechanism of action is
unknown, but its use is justified due to the good results obtained as an adjuvant therapy in dermoid tumours with retroperitoneal invasion, and in cases of Peyronie’s disease. The anti-estrogenic effects of tamoxifen have been associated with the inhibition of the tumour necrosis factor-beta (TNF-β) of the fibroblasts, impeding their growth. Obstructive complications are not uncommon in IRPF, making vascular and urological intervention necessary, as in the present case [5].

In short, and consistent with current literature, the use of tamoxifen appears to be effective as a therapeutic measure for refractory disease in patients receiving treatment with immunosuppressants and corticosteroids.

References