REVISTA BRASILEIRA DE REUMATOLOGIA

www.reumatologia.com.br

SOCIEDADE BRASILEIRA DE REUMATOLOGIA

Original article

Retroperitoneal fibrosis: case series of five patients and review of the literature

Shacahf Shiber[a,b,*], *Noa Eliakim-Raz*[b,c], *Molad Yair*[a,b]

[a] *Rheumatology Unit, Rabin Medical Center, Petach Tikva, Israel*
[b] *Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel*
[c] *Department of Medicine, Rabin Medical Center, Beilinson Hospital, Petah Tikva, Israel*

ARTICLE INFO

Article history:
Received 5 April 2014
Accepted 14 September 2014
Available online 6 January 2015

Keywords:
Retroperitoneal fibrosis
Prednisone
Renal failure
Tamoxifen
IgG4

ABSTRACT

Chronic periaortitis (CP) is an umbrella term used to describe a group of nosologically allied conditions that include idiopathic retroperitoneal fibrosis (Ormond's disease), inflammatory abdominal aortic aneurysm, and perianeurysmal retroperitoneal fibrosis. Retroperitoneal fibrosis encompasses a range of diseases 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 envelop neighboring structures-ureters. Retroperitoneal fibrosis is generally idiopathic, but can also be secondary to the use of certain drugs, malignant diseases, infections, and surgery. Here we describe a 5 years follow-up (2006–2011) of 5 patients admitted to our hospital with symptoms, laboratory, imaging and pathologic finding compatible with retroperitoneal fibrosis. We review our clinical course of our patient with respect to the literature.

Fibrose retroperitoneal: série de cinco casos e revisão da literatura

Palavras-chave:
Fibrose retroperitoneal
Prednisona
Insuficiência renal
Tamoxifeno
IgG4

RESUMO

Periaortite crônica (PC) é um termo genérico usado para descrever um grupo de condições nosologicamente ligadas que incluem a fibrose idiopática retroperitoneal (doença de Ormond), o aneurisma da aorta abdominal inflamatório e a fibrose retroperitoneal perianeurismática. O termo fibrose retroperitoneal engloba uma gama de doenças que se caracterizam pela presença de um tecido fibroinflamatório que geralmente envolve a aorta abdominal e as artérias ilíacas, se estende ao retroperitôneo e envolve estruturas ureterais vizinhas. A fibrose retroperitoneal geralmente é idiopática, mas pode também ser secundária ao uso de determinados fármacos, doenças malignas, infecções e cirurgia. Este

* *Corresponding author.*
E-mail: sofereret@gmail.com (S. Shiber).
http://dx.doi.org/10.1016/j.rbre.2014.09.006

estudo descreve o seguimento por cinco anos (2006–2011) de cinco pacientes internados em nosso hospital que apresentavam sintomas e achados laboratoriais, de imagem e patológicos compatíveis com a fibrose retroperitoneal. Revisou-se a evolução clínica dos pacientes, que foi comparada com os achados da literatura.

Introduction

Chronic periaortitis encompasses a group of rare abdominal aorta diseases, including idiopathic (nonaneurysmal) retroperitoneal fibrosis, inflammatory abdominal aortic aneurysm (IAAA), and perianeurysmal retroperitoneal fibrosis.[1] Some cases involve the thoracic aorta, leading to mediastinal fibrosis. The idiopathic form accounts for over 70% of cases of retroperitoneal fibrosis.[2] According to a Finnish study, the estimated incidence of idiopathic retroperitoneal fibrosis is 0.1 per 100,000 person-years.[3] Men are affected twice to three times more often than women, the mean age at presentation is 50–60 years, although reports of the condition in children and older adults are not uncommon.

This report reviews the literature and describes clinical and laboratory characteristics, treatment, and outcome of a series of patients with retroperitoneal fibrosis.

Results

Five patients with retroperitoneal fibrosis were identified reviewing all rheumatology clinic inpatients records (2006–2011). The mean (±SD) age was 62.6 ± 5.7 years and mean (±SD) duration of disease at diagnosis was 3.2 ± 2.16 years. Abdominal and groin pain were the most common clinical manifestations (all patients).

All patients had normocytic anemia and a high C-reactive protein (CRP) and ESR levels. Three patients were found to have renal failure. Treatment in all cases consisted of steroids and tamoxifen. One patient did not improve after treatment, two patients had partial response, and two patients had full response (complete remission).

Case reports

Patient 1

A 64-year-old man with a history of dyslipidemia presented with chief complaint of groin pain during the preceding two months. Physical examination was normal, laboratory tests revealed elevated ESR (101 mm/h), mild anemia with hemoglobin of 11.6 g/dL, and mildly elevated creatinine (1.29 mg/dL). Anti-nuclear antibody (ANA) titer was negative. Imaging included a computed tomography (CT) scan, which showed bilateral moderate hydronephrosis and a mass surrounding the abdominal aorta from the level of the renal arteries through the iliac bifurcation. The patient underwent bilateral ureterolysis and omentopexy. Biopsy of the retroperitoneal mass disclosed collagenous tissue with inflammatory cells, compatible with retroperitoneal fibrosis. The patient was first prescribed prednisone 1 mg/kg/day with tapering therapy down for three months then maintenance therapy with azathioprine 2 mg/kg/day and tamoxifen 20 mg bid. At a 6-month follow-up, an increment in the retroperitoneal process was noted and the dose of azathioprine was increased. Repeated CT scan one year later revealed a stable retroperitoneal lesion. The patient continued tamoxifen and azathioprine.

Patient 2

A 55-year-old man, a smoker and with a history of diabetes mellitus type 2, presented with abdominal pain and weight loss during the preceding 6 months. Physical examination was remarkable for abdominal tenderness. Laboratory tests revealed elevated inflammatory markers (CRP 138 mg/L, ESR 77 mm/h), mild anemia and normal creatinine. A CT scan, revealed hard tissue surrounding the abdominal aorta with hydronephrosis, and signs of atherosclerosis within the large arteries. The biopsy revealed fibrous tissue and inflammatory cells.

Treatment with high-dose steroids (1 g methylprednisolone for 3 days) and cyclophosphamide (500–1000 mg/m^2 IV monthly for 6 doses) led to resolution of the symptoms. Thereafter, the patient was prescribed tamoxifen and azathioprine 2 mg/kg/day. Under treatment, repeated imaging of magnetic resonance (MR) angiography study, one year later at follow up, disclosed minor tissue surrounding the aorta and inflammatory markers normalized. Treatment with cyclophosphamide was stopped after 6 months and the patient continued tamoxifen and azathioprine.

Patient 3

A 57 year-old man with a 3 year history of Etanercept treated sacroiliitis presented with pelvic pain and burning sensation in the groin. Physical examination was positive for bilateral flank tenderness, laboratory tests revealed elevated inflammatory markers (CRP 126 mg/L, ESR 113 mm/h), mild anemia with hemoglobin of 11.5 g/dL, and mildly elevated creatinine of 1.28 mg/dL. ANA was negative. IgG4 isotype level was elevated (243 mg/dL; normal 1–112). A CT scan, revealed a retroperitoneal mass encompassing the inferior vena cava and abdominal aorta with hydronephrosis. The sacroiliac joints appeared normal. Laparoscopic biopsy disclosed fibrotic tissue with chronic inflammatory aggregation and a few macrophages. Treatment consisted of prednisone 60 mg qd and tamoxifen 20 mg bid. Etanercept was discontinued. Repeated CT scan at follow up, one year later, showed marked regression of the retroperitoneal mass. Prednisone was tapered down and the patient continues tamoxifen only.