

Idiopathic Retroperitoneal Fibrosis Associated with Biermer's Disease

Hassan Kassem*, Kamal Khamadi and Luc Turner

Departement of Internal Medecine, CHSE-Dourdan Site 2, rue du potelet, 91410, Dourdan, France

*Corresponding author: Hassan Kassem, Departement of Internal Medecine, CHSE-Dourdan Site 2, rue du potelet, 91410, Dourdan, France, Tel: 0033160815942; E-mail: hkassem@hotmail.fr

Received date: September 25, 2017; Accepted date: October 07, 2017; Published date: October 10, 2017

Citation: Kassem H, Khamadi K, Turner L (2017) Idiopathic Retroperitoneal Fibrosis Associated with Biermer's Disease. J Autoimmune Disord Vol 3 No 4: 44.

Copyright: © 2017 Kassem H, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Introduction

In December 2008, a 70-year old woman presented to the emergency department for diffuse abdominal pain and denied any fever, diarrhea, or vomiting; she had no urinary symptoms. She had a history of appendectomy, cholecystectomy, and of bilateral breast prosthesis. She was not taking any medications. Abdominal examination and gynecological examination were normal. Lab results showed an initial sedimentation rate of 46 mm (normal<10), and pancytopenia: hemoglobin at 11.7 g/dl (normal: 12.5-16 g/dl), mean corpuscular volume (MCV) at 88 (normal: 80-95), reticulocytes at 86400/mm³ normal>35000, the leukocytes were at 3500/mm³ (normal: 4000-10000) and platelets at 127000/mm³ (normal: 150000-400000). C-reactive protein (CRP), blood ionogram, blood glucose, serum calcium, creatinine, lactate dehydrogenase (LDH), complete liver function and lipase were normal. The serum protein electrophoresis did not show any monoclonal peak. Urine count was sterile; red blood cell and proteinuria counts were negative.

A CT scan of the abdomen and pelvis showed a promontory obstruction at the level of the right ureter, associated with retroperitoneal infiltration, which led to the initial diagnosis of retroperitoneal fibrosis (RPF). It showed no other abnormalities at the abdomino-pelvic levels.

Our etiological diagnosis of RPF included: antinuclear antibodies (ANA), anti-neutrophil-cytoplasmic antibodies (ANCA), and cryoglobulinemia were negative, tumor markers were negative: carcinoembryonic antigen (ACE), neuron specific enolase (NSE), carbohydrate antigen 15-3 (CA15-3), carbohydrate antigen 125 (CA125), carbohydrate antigen 19-9 (CA19-9). Thyroid-stimulating hormone (TSH) was normal and antithyroid antibodies were negative. The IgG 4 was normal. Thoracic CT scan showed no tumors or infectious lesions. Mammography showed no abnormality.

The pancytopenia was found to be associated with vitamin B12 deficiency at 178 ng/l (normal: 210-910) and iron deficiency with ferritin at 14 µg/l (normal: 20-190) and transferrin at 3.4 g/l (normal: 1.60-2.60). Folic acid was normal. This evaluation was completed by an assay of the gastric parietal cell antibodies which returned positive at 1280 (positive value: greater than or equal to 80). Anti-intrinsic factor antibodies were negative.

Gastroscopy showed moderate atrophy of the fundus with histology in favor of atrophic gastritis.

Our final diagnosis was idiopathic RPF associated with Biermer's disease. Our treatment was double J stent for the right ureter obstruction, intramuscular vitamin B12 administration and oral Tardyferon. The patient had refused the corticotherapy. At a three month-control, an abdomino-pelvic CT scan showed a complete disappearance of the retroperitoneal fibrosis as well as the absence of dilation of the pyelocalicious cavities and the right ureter. The double J stent was removed in May 2009.

Several abdominal-pelvic scans were performed between 2009 and 2016, always showing the absence of retroperitoneal fibrosis.

RPF is an uncommon disease characterized by a fibrous reaction that takes place in the peri-aortic retroperitoneum and often entraps the ureters causing obstructive uropathy. RPF is idiopathic in more than two third of the cases, the remaining third being secondary to other causes such as neoplasms, infections, trauma, radiotherapy, major surgery, asbestosis, Erdheim-Chester disease, and intake of drugs [1]. Idiopathic RPF is an immune-mediated disease, which can either be isolated, associated with other autoimmune diseases, or arise in the context of a multifocal fibro-inflammatory disorder recently renamed as IgG4-related disease. Clinical symptoms are not specific, mostly abdominal or lumbar pains sometimes associated with altered general health conditions [2]. The differential diagnosis between idiopathic, IgG4-related and secondary RPF is crucial, essentially because the therapeutic approaches which can be dramatically different [3]. Currently, abdominal computed tomography (CT) and magnetic resonance imaging (MRI) are considered the imaging studies of choice to diagnose idiopathic RPF [4,5]. On CT, it appears as a homogeneous tissue, isodense to muscle, that surrounds the lower abdominal aorta and the iliac arteries, and often enveloping neighbouring structures such as the ureters that generally are displaced medially.

In a retrospective multicentric French study, all patients with fibrosis atypical extension (above the renal artery or to the thorax) had a secondary RPF [2].

First-line medical treatment of idiopathic RPF is Prednisone at a dose of 1 mg/kg/day. After 1 month of corticosteroid therapy, the status of the disease is assessed: clinical signs, VS, CRP and an imaging for fibrosis [3]. If remission is obtained, the corticosteroid is progressively reduced until the dose of 5 mg per day is reached in 3 to 4 months, which dose must be maintained for 6 to 9 months. Several immunosuppressants may be used in patients with corticosteroid resistance or dependence or in the case of relapse.

In our patient, the etiological assessment did not found any evidence for a secondary RPF.

To our knowledge, the association of idiopathic RPF with Biermer's disease has not been published. Our observation may be a first. The RPF disappeared 3 months after the start of symptomatic treatment of Biermer's disease without requiring corticosteroid therapy.

In our observation, we do not have a physiopathological explanation of the regression of RPF without corticosteroid therapy. Could the symptomatic treatment of Biermer's disease be responsible of such evolution? Other similar cases are needed to support this hypothesis.

Conflict of Interest

The authors declare that they have no competing interests.

References

1. Palmisano A, Vaglio A (2009) Chronic periaortitis: A fibro-inflammatory disorder. *Best Pract Res Clin Rheumatol* 23: 339-353.
2. Lioger B, Yahiaoui Y, Kahn JE, Fakhouri F, Belenfant X, et al. (2016) Fibrose rétro-péritonéale de l'adulte: analyse descriptive et évaluation de la pertinence des examens complémentaires réalisés à visée diagnostique à partir d'une série rétrospective multicentrique de 77 cas. *La Revue De Medecine Interne* 37: 387-393.
3. Urban ML, Palmisano A, Nicastro M, Corradi D, Buzio C (2015) Idiopathic and secondary forms of retroperitoneal fibrosis: A diagnostic approach. *La Revue De Medecine Interne* 36: 15-21.
4. Vaglio A, Salvarani C, Buzio C (2006) Retroperitoneal fibrosis. *Lancet* 367: 241-251.
5. Salvarani C, Pipitone N, Versari A, Vaglio A, Serafini D, et al. (2005) Positron emission tomography (PET): evaluation of chronic periaortitis. *Arthritis Rheum* 53: 298-303.