Best Practice & Research Clinical Rheumatology 26 (2012) 439–448

Contents lists available at SciVerse ScienceDirect

Best Practice & Research Clinical Rheumatology

journal homepage: www.elsevierhealth.com/berh

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Retroperitoneal fibrosis

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Keywords:
Chronic periaortitis
Retroperitoneal fibrosis
Computerized tomography
Magnetic resonance
PET
Glucocorticoids
Tamoxifen
Mycophenolate mofetil
Cyclophosphamide
Rituximab

Retroperitoneal fibrosis is a rare syndrome hallmarked by a fibrosclerotic tissue in the retroperitoneum, often leading to encasement of the ureters. About two-thirds of cases of retroperitoneal fibrosis are idiopathic, while the remaining cases are secondary to a variety of different causes, including drugs, tumors and infections. Idiopathic retroperitoneal fibrosis may be associated with abdominal aorta aneurysms, with vasculitis of the thoracic aorta and of epi-aortic vessels, or both. Most patients present with abdominal and/or low back pain. Serum markers of inflammation are usually, but not invariably, elevated. The diagnosis is secured by computed tomography or magnetic resonance imaging, which typically show an enhancing retroperitoneal mass medially dislodging the ureters. Positron emission tomography can be useful to document the extent and metabolic activity of the inflammatory process. Treatment rests on glucocorticoids with a tapering scheme variably combined with immunosuppressive agents. In cases of ureter obstruction, relief of obstruction by stenting or ureterolysis is required.

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1521-6942/$ – see front matter © 2012 Published by Elsevier Ltd.
http://dx.doi.org/10.1016/j.berh.2012.07.004

Practice points

- Idiopathic retroperitoneal fibrosis is a fibro-inflammatory disorder characterised by a retroperitoneal mass often encasing the ureters.
- Computed tomography or magnetic resonance imaging is required to secure the diagnosis.
- Glucocorticoid therapy is usually effective, but relapses are high upon glucocorticoid tapering.
- Immunosuppressive agents should be used in patients with frequent relapses.

Research agenda

- Classification criteria for idiopathic retroperitoneal fibrosis should be developed.
- The molecular mechanisms of retroperitoneal fibrosis need to be better understood.
- Controlled trials of synthetic and biological agents are warranted.

Introduction

Retroperitoneal fibrosis is a rare syndrome characterised by the development of fibrosclerotic tissue in the retroperitoneum, which often leads to encasement of the ureters and, less frequently, of blood and lymphatic vessels [1]. About two-thirds of cases of retroperitoneal fibrosis are idiopathic, while the remaining cases are secondary to intake of a number of drugs, infections, malignancies, surgery or exposure to radiation [1,2]. In this review, we will focus on the idiopathic form of retroperitoneal fibrosis, and discuss the latest developments in the understanding of its pathogenesis, imaging and treatment.

Classification of retroperitoneal fibrosis

There are no formal criteria to classify idiopathic retroperitoneal fibrosis (IRF). Currently, IRF is considered part of the spectrum of chronic periaortitis (CP), a large-vessel vasculitis [3]. Specifically, IRF is characterised by a retroperitonal fibro-inflammatory tissue in the absence of a dilated aorta, while in inflammatory abdominal aorta aneurysms (IAAAs) the fibro-inflammatory tissue develops around a dilated aorta, and in perianeurysmal retroperitoneal fibrosis the fibro-inflammatory tissue spreads from a dilated aorta into the retroperitoneum [3]. CP has been shown to be associated with large-vessel vasculitis in vessels other than the abdominal aorta, including the epiaortic and gastrointestinal vessels, as well as with various auto-immune disorders such as Hashimoto thyroiditis and primary Sjøgren's syndrome, in agreement with its auto-immune nature [1,3–5].

There is evidence that, in a number of patients, CP may be part of the so-called immunoglobulin G4 (IgG4)-related disease (IgG4-RD), an auto-immune disorder characterised by elevated serum levels of IgG4 and abundant infiltration of IgG4-positive plasma cells in different organs [6]. A study assessing 17 patients with retroperitoneal fibrosis demonstrated that 10 patients had elevated numbers of IgG4-positive plasma cells both in the serum and at tissue level [7]. Likewise, approximately 50% of patients with IAAA have evidence of IgG4-RD [8].

Epidemiology

CP is a rare disease with prevalence of 1.4/100 000 and a yearly incidence of 0.1–1.3/100,000 [9,10]. Mean age at onset is approximately 55–60 years [11–13]. Men are affected two to three times more