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Chronic periaortitis: a fibro-inflammatory disorder

Alessandra Palmisano, MD, Doctor, Augusto Vaglio, MD, Doctor *

Dipartimento di Clinica Medica, Nefrologia e Scienze della Prevenzione, Università degli Studi di Parma, Via Gramsci 14, 43100 Parma, Italy

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Chronic periaortitis includes a spectrum of rare conditions characterized by fibro-inflammatory tissue surrounding the abdominal aorta. Although it has been considered a localized inflammatory disease secondary to atherosclerosis, several clinico-laboratory findings suggest a systemic autoimmune origin; additionally, it may involve the thoracic aorta and the origin of its major branches, with a pattern similar to that of the large-vessel vasculitides. Its pathogenesis is still unclear. Computed tomography and magnetic resonance imaging are the modalities of choice for the diagnosis, whereas fluorodeoxyglucose/positron emission tomography emerges as a sensitive imaging modality to assess the inflammatory activity of the periaortic tissue. The treatment of chronic periaortitis is largely empirical, since no randomized trials have been carried out. Corticosteroids, immunosuppressants and endoscopic or surgical procedures must be appropriately combined for the correct management of chronic periaortitis patients.

Chronic periaortitis (CP) refers to a spectrum of idiopathic diseases whose common denominator is a fibro-inflammatory tissue developing in the periaortic retroperitoneum and frequently encasing neighbouring structures such as the ureters and the inferior vena cava [1,2]. CP includes idiopathic retroperitoneal fibrosis (IRF), inflammatory abdominal aortic aneurysms (IAAAs), and a combination of the two diseases called perianeurysmal retroperitoneal fibrosis [3].

IRF accounts for more than two thirds of the cases of retroperitoneal fibrosis, whereas the remaining third is secondary to several aetiologies such as neoplasms, drugs, trauma, radiotherapy and infections [4]. Histology of the retroperitoneal mass in IRF shows a collagen-rich tissue along with an

* Corresponding author. Tel.: +39 0521 702345; Fax: +39 0521 033185.
E-mail address: augusto.vaglio@virgilio.it (A. Vaglio).

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inflammatory infiltrate consisting of plasma cells, lymphocytes, and macrophages [5], and in half of the cases also includes aspects of small-vessel vasculitis [6].

In IAAAs and perianeurysmal retroperitoneal fibrosis, also referred to as the aneurysmal forms of CP, the fibro-inflammatory tissue develops around a dilated aorta; these forms share with their non-aneurysmal counterpart (i.e. IRF) clinical, laboratory and histological characteristics, and are therefore part of the same disease spectrum.

The present review will focus on the clinical and pathogenetic aspects of CP, and discuss the diagnostic elements that may help differentiate this condition from other inflammatory disorders with aortic and periaortic involvement.

Nomenclature

The confusion about the nomenclature of CP is due to the heterogeneity of the diseases it embraces and to its rarity, which so far has not allowed the development of diagnostic or classification criteria.

CP may develop around an undilated or a dilated aorta. In the former case, the definition of *IRF* is commonly used; in the latter, clinicians used to distinguish between *perianeurysmal retroperitoneal fibrosis* and *IAAAs* on the basis of the presence or absence, respectively, of encasement of adjacent structures [7]. Since the boundaries between the latter two forms may be artificial, and patients may present with varying degrees of periaortic inflammation and fibrosis, we prefer to include them under the definition of *aneurysmal forms of CP*. When the aorta is only slightly dilated, the distinction between aneurysmal and non-aneurysmal CP may not be clear-cut; it has been proposed that the definition of aneurysmal CP requires an abdominal aortic diameter (i.e. excluding the periaortic inflammatory tissue) >3 cm [8].

The clinical entities included in the spectrum of CP are *idiopathic*. This consideration is important, as some cases (about one third) of retroperitoneal fibrosis, particularly if non-aneurysmal, are instead *secondary* to different aetiologies, such as the use of drugs (e.g. ergot alkaloids, methysergide, β-blockers, dopamine agonists), primary retroperitoneal cancer (e.g. lymphoma, sarcoma) or retroperitoneal metastatic disease (e.g. various carcinomas), carcinoid syndrome, trauma, radiotherapy, major abdominal surgery and infections (e.g. tuberculosis, actinomycosis) [2,4,9]. Being secondary to definite causes, these conditions are not part of CP but always need to be considered in the differential diagnosis.

In most of the cases CP only affects the abdominal aorta, the iliac arteries, and the perivascular soft tissues, but in some patients it also involves the thoracic aorta and the surrounding tissue (*thoracic periaortitis*), as well as the origin of the epiaortic vessels [10,11].

CP, especially its non-aneurysmal form (i.e. IRF), may be *isolated* or may develop in the setting of a *systemic* immune-mediated disease, such as systemic lupus erythematosus [12], rheumatoid arthritis [13–15] and small- and medium-sized-vessel vasculitides (e.g. Wegener granulomatosis, polyarteritis nodosa) [16,17], or found to be *associated* with organ-specific autoimmune disorders such as autoimmune thyroiditis [18,19].

CP, again particularly its non-aneurysmal form, may occasionally arise in the context of a newly recognized clinico-pathological systemic disorder called *IgG4-related sclerosing disease*, characterized by extensive T-lymphocyte- and IgG4-bearing plasma-cell infiltration of various organs in which fibrosis with obliterative phlebitis is pathologically induced [20]. Clinical manifestations are apparent in the pancreas (autoimmune pancreatitis), bile duct (sclerosing cholangitis), gallbladder (cholecystitis), salivary glands (chronic sialoadenitis), mediastinum (mediastinal fibrosis), retroperitoneum (retroperitoneal fibrosis), kidney (tubulo-interstitial nephritis), lung (interstitial pneumonia), and prostate (prostatitis); some inflammatory pseudotumours can also be associated [21–23]. Patients with multiple lesions of IgG4-related sclerosing disease tend to have high serum levels of IgG4, which seems to be a useful marker of disease activity [24]. Recent reports have shown that a group of patients with aneurysmal CP (i.e. IAAAs) also have high serum IgG4 concentrations and IgG4-plasma-cell infiltration; this has led some authors to suggest that a subset of IAAAs can belong to the IgG4-related sclerosing disease [25–28].

Multifocal fibrosclerosis is a rare fibroproliferative systemic entity characterized by multiple conditions such as IRF, sclerosing colangitis, Riedel's thyroiditis and orbital pseudotumour. As this entity basically has the same histopathological features as IgG4-related sclerosing disease, it has been