Autoimmunity Reviews xxx (2016) xxx–xxx

Contents lists available at ScienceDirect

Autoimmunity Reviews

journal homepage: www.elsevier.com/locate/autrev

Review

Comparison of idiopathic (isolated) aortitis and giant cell arteritis-related aortitis. A French retrospective multicenter study of 117 patients

Olivier Espitia [a], Maxime Samson [b], Thomas Le Gallou [c], Jérôme Connault [a], Cedric Landron [d], Christian Lavigne [e], Cristina Belizna [e], Julie Magnant [f], Claire de Moreuil [g], Pascal Roblot [d], François Maillot [f], Elisabeth Diot [f], Patrick Jégo [c], Cécile Durant [a], A. Masseau [a], Jean-Marie Brisseau [a], Pierre Pottier [a], Alexandra Espitia-Thibault [a], Anabele Dos Santos [h], François Perrin [a], Mathieu Artifoni [a], Antoine Néel [a], Julie Graveleau [i], Philippe Moreau [j], Hervé Maisonneuve [k], Georges Fau [l], Jean-Michel Serfaty [l], Mohamed Hamidou [a], Christian Agard [a,*]

[a] Department of Diagnostic Cardio-vascular Imaging, University hospital of Nantes, Nantes, France
[b] Department of Internal Medicine and Clinical Immunology, University Hospital of Dijon, Dijon, France
[c] Department of Internal Medicine, University Hospital of Rennes, Rennes, France
[d] Department of Internal Medicine, University Hospital of Poitiers, Poitiers, France
[e] Department of Internal Medicine, University Hospital of Angers, Angers, France
[f] Department of Internal Medicine, CHRU of Tours, Tours, France
[g] Department of Internal Medicine, University Hospital of Brest, Brest, France
[h] Department of Hematology, Hospital of Vannes, Vannes, France
[i] Department of Medicine, Hospital of Saint-Nazaire, Saint-Nazaire, France
[j] Department of Hematology, Hospital of Lorient, Lorient, France
[k] Department of Hematology, Hospital of La Roche sur Yon, La Roche sur Yon, France
[l] Department of Radiology, University Hospital of Nantes, Nantes, France

ARTICLE INFO

Article history:
Received 9 February 2016
Accepted 14 February 2016
Available online xxxx

Keywords:
Aortitis
Giant cell arteritis
Idiopathic aortitis
Isolated aortitis
Aortic aneurysm

ABSTRACT

Objectives: The aim of the study was to compare clinical/imaging findings and outcome in patients with idiopathic (isolated aortitis, IA) and with giant cell arteritis (GCA)-related aortitis.
Methods: Patients from 11 French internal medicine departments were retrospectively included. Aortitis was defined by aortic wall thickening >2 mm and/or an aortic aneurysm on CT-scan, associated to inflammatory syndrome. Patients with GCA had at least 3 ACR criteria. Aortic events (aneurysm, dissection, aortic surgeries) were reported, and free of aortic events-survival were compared.
Results: Among 191 patients with non-infectious aortitis, 73 with GCA and 44 with IA were included. Patients with IA were younger (65 vs 70 years, $p = 0.003$) and comprised more past/current smokers (43 vs 15%, $p = 0.0007$). Aortic aneurisms were more frequent (38% vs 20%, $p = 0.03$), and aortic wall thickening was more pronounced in IA. During follow-up (median = 34 months), subsequent development of aortic aneurysm was significantly lower in GCA when compared to IA ($p = 0.009$). GCA patients required significantly less aortic surgery during follow-up than IA patients ($p = 0.02$). Mean age, sex ratio, inflammatory parameters, and free of aortic aneurism survival were equivalent in patients with IA ≥ 60 years when compared to patients with GCA-related aortitis.
Conclusions: IA is more severe than aortitis related to GCA, with higher proportions of aortic aneurism at diagnosis and during follow-up. IA is a heterogeneous disease and its prognosis is worse in younger patients <60 years. Most patients with IA ≥ 60 years share many features with GCA-related aortitis.

Contents

* Corresponding author at: Department of Internal Medicine, Hôtel-Dieu, Centre Hospitalier Universitaire de Nantes, 1 place Alexis Ricordeau, 44093 Nantes, France. Tel.: +33 240 084 933; fax: +33 240 083 379.
E-mail address: christian.agard@chu-nantes.fr (C. Agard).

http://dx.doi.org/10.1016/j.autrev.2016.02.016
1568-9972/

1. Introduction

Aortitis is a general term for a spectrum of disorders characterized by inflammation of the aortic wall [1]. The main underlying diseases causing non-infectious aortitis are giant cell arteritis (GCA) and Takayasu arteritis [2,3] and more rarely, sarcoidosis, Behçet's disease, Cogan syndrome, granulomatosis with polyangiitis, spondylarthropathy, IgG4-related disease, or relapsing polychondritis. In addition, idiopathic (isolated) aortitis (IA) has also been described [4]. Aortitis is related to significant morbidity and mortality through the development of aortic aneurysm, aortic wall rupture, aortic acute dissection, and/or thrombotic luminal obstruction [5]. Surgical series have found that granulomatous/giant cell aortitis is the most common histological pattern of aortitis [2]. However, many patients do not require surgery and aortitis is usually diagnosed when a significant aortic thickening (generally >2 mm) is found on computed tomography (CT) or magnetic resonance imaging (MRI).

GCA is the most frequent vasculitis in patients above 60 years, typically affecting temporal arteries, but involving also large arteries like proximal limb arteries [6] or the aorta. The estimated prevalence of aortitis in GCA ranges from 33 to 65% [7–9], and aortic aneurysm or ectasia have been found in about 10–15% of patients at the time of diagnosis of GCA [7,8,10–14]. Regarding IA, most of studies have included surgical cases, especially of the ascending aorta, and IA accounts for 4–8% of surgical aortitis cases [15]. In practice, many patients with IA above 50 or 60 years are considered as having GCA-related aortitis, even if they do not fulfill the "American College of Rheumatology" (ACR) diagnosis criteria. However, it is not clear if IA above 50–60 years represent variants of GCA [16]. In fact, differences between IA and GCA-related aortitis have never been clearly studied in a non-surgical population. Data are also lacking about epidemiology, optimal treatment, monitoring, and prognosis of patients with IA or with GCA-related aortitis. Thus, the aim of the present study was to describe the initial clinical and CT-scan aortic characteristics, the treatment used, and to analyze the outcome of patients with IA or GCA-related aortitis, in order to underline potential common features or differences between these 2 entities.

2. Methods

This retrospective multicenter study was conducted in French internal medicine departments of 7 university hospitals and 4 non-university hospitals. All patients with a diagnosis of non-infectious aortitis assessed between January 2000 and December 2014 were identified. Aortitis was defined by an aortic wall thickening >2 mm on CT-scan [17] and/or an aortic aneurysm, associated to inflammatory syndrome (CRP > 5 mg/L and/or fibrinogen > 4 g/L) unexplained by any other cause. Finally, only patients with GCA-related aortitis or IA were included. At least 3 ACR criteria, including age over 50, were required for GCA diagnosis [18]. IA was defined by aortitis, associated to inflammatory syndrome, without any other ACR criteria for GCA except the age [18], and without any diagnosis criteria for any other causes of aortitis [19,20].

The flowchart is summarized in Fig. 1. Two physicians (OE, CA) analyzed each medical file, aiming to classify patients with either GCA-related aortitis or IA. Initial clinical data, biological parameters, aortic CT-scan findings, and treatment modalities were analyzed. Follow-up data, therapeutics, evolution of aortic imaging, and aortic events were also studied at the time of the last visit. An aortic event was defined by the subsequent occurrence of an aortic aneurysm, ectasia, dissection, or stenosis, on CT-scan or on Doppler ultrasonography, only in patients free of any of these features at the time of diagnosis. This study was approved by the local ethics review committee.

For statistical analyses, results were expressed by mean ± standard deviation (SD) or as median, range. Categorical variable were compared using chi-square tests or Fisher's exact tests when any of the expected cell counts of a 2 × 2 table was less than 5. Comparisons of quantitative

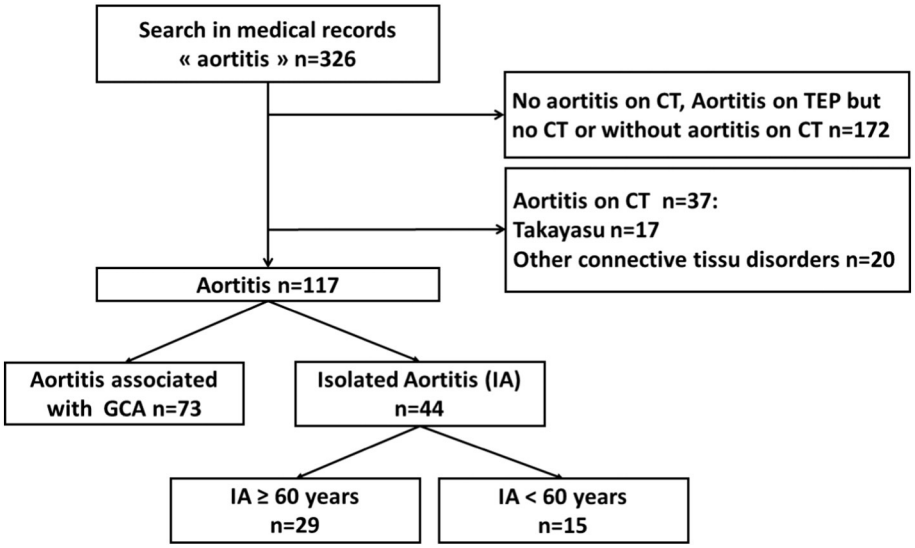

Fig. 1. Flowchart of patient's selection.

Please cite this article as: Espitia O, et al, Comparison of idiopathic (isolated) aortitis and giant cell arteritis-related aortitis. A French retrospective multicenter study..., Autoimmun Rev (2016), http://dx.doi.org/10.1016/j.autrev.2016.02.016