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

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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 70y, 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≥60y 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<60y. Most patients with IA≥60y share many features with GCA-related aortitis.

Keywords: Aortitis, giant cell arteritis, idiopathic aortitis, isolated aortitis, aortic aneurysm

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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 syndrom, 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>2mm) is found on computed tomography (CT) or magnetic resonance imaging (MRI).

GCA is the most frequent vasculitis in patients above 60y, 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 60y 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 50-60y 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

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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.

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>4g/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 excepted the age [18], and without any diagnosis criteria for or any other causes of aortitis [19, 20].

Flow chart is summarized in figure 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

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tests when any of the expected cell counts of a 2x2 table was less than 5. Comparisons of quantitative variables were performed using Student's *t-t*est. A *p*-value < 0.05 was considered to be statistically significant. Survival curves were made to compare GCA-related aortitis and IA patients. Aortic event free-survival were compared between groups. Thus, the aortic events at the time of aortitis diagnosis were excluded from analysis, and only new subsequent events were considered. To analyze the free of aortic aneurism survival, patients with aortic aneurism at diagnosis were excluded. Progression of aortic aneurysm was not considered as an aortic event. To analyze the free of aortic surgery survival, patients with aortic surgery at diagnosis were excluded. To analyze the free of aortic dissection survival, patients with aortic dissection at diagnosis were excluded. Kaplan-Meier curves were then made and a log-rank test was performed to compare aortic event-free survivals. Stastistical analyses were all performed with Graph Pad Prism v5 software.

Results

This study analyzed 117 cases of aortitis: 73 GCA-related aortitis, 44 IA including 29 ≥60y and 15 <60y. Eighty-five patients were women, mean age at aortitis diagnosis was 67.5y [range:37-87]. In the GCA-related aortitis group, headache was present in 65.8% of the patients, weight loss in 53.4%, fever in 42.5%, *polymyalgia rheumatica* (PMR) in 26%, hyperesthesia of the scalp in 13.7%, jaw claudication in 12.3%, and ocular involvement in 9.6%. Every patient from the GCA-related aortitis group had undergone a temporal artery biopsy (TAB) with GCA consistent lesions in 51 cases. Among the 22 cases with negative TAB, 81.8% had headache, 31.8% had PMR, 27.3% had hyperesthesia of the scalp and 13.6% had jaw claudication. Among IA cases, 34.1% had weight loss, 34.1% fever and none had ocular involvement. TAB was performed in 70.5% of IA patients and was negative.

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Symptoms leading to the diagnosis of aortitis were thoracic or abdominal pain in 26% of the patients, dyspnea in 23%, cough in 17%. Thoracic or abdominal pain tended to be more frequent in patients with IA (34.1% vs 17.8%, p=0.07). Conversely, cough was more frequent among GCA-related aortitis (24.7% vs 9.1% in IA, p=0,05). In almost half of cases, aortitis was found in patients with isolated inflammatory syndrome. A majority of our GCA patients (n=52) had undergone a systematic CT-scan at the time of GCA diagnosis, and aortitis was found at that time in 50 patients. In 23 cases, aortitis was diagnosed after GCA and mean time between GCA and aortitis diagnosis was 49 months [range: 2-143] for these patients.

Main characteristics of patients with GCA-related aortitis and IA are presented in table 1. Mean age in GCA-related aortitis patients was significantly higher. Past/current smokers were significantly more found among IA patients. Location of aortic involvement was not significantly different. However, aortic aneurysm were significantly more found in patients with IA (38.6% vs. 20.5%, p=0.03) and mean maximal thickening of the aortic wall was higher in the IA group.

Corticosteroids was given to all the 73 patients with GCA-related aortitis and to 40 patients with IA (91%). Indeed, four patients with IA did not receive any specific treatment because of well tolerated inflammation. Among IA patients, treatment was deliberately delayed in 7 cases because of non symptomatic inflammation. Prednisone was the sole oral steroid therapy used, started at the mean daily dose of 0.8±0.2 mg/kg in GCA-related aortitis and 0.82±0.2 mg/kg in IA. At aortitis diagnosis, 52 GCA-related aortitis patients (71.2%) and 27 IA patients (61.4%) were treated with oral platelet aggregation inhibitors, and respectively 17 (23.3%) and 15 (34.1%) had statins, 31 (42.5%) and 19 IA (43.2%) had high blood pressure medication, 15 (20.5%) and 13 (29.5%) had beta-blockers, which was equally distributed in both groups.

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Median follow-up was 34 months in GCA-related aortitis and 34.5 months in IA (p=0,90). The majority of our aortitis patients (n=90/117, 77%) were only treated with steroids while 17 in the GCA-related aortitis group (23%) and 10 in the IA group (22.7%) required alternate therapy: methotrexate (13 GCA, 7 IA), azathioprine (7 GCA, 3 IA), IV cyclophosphamide (5 GCA, 4 IA), tocilizumab (1 GCA, 2 IA), and anakinra 1 IA).

	GCA-Ao n=73	**IA n=44**	*p*-value
Age (median, years) [min-max]	70 [52-83]	65.0 [37-87]	**0.0003**
Female, n (%)	57 (78.1)	28 (63.6)	0.09
Biological parameters ±SD			
Mean CRP (mg/L)	111.3±78.3	109.9 ±79.6	0.93
ESR, mm/h	70.5 ±38.1	53.2 ±49.2	0.18
Fibrinogen (g/L)	7.5 ±1.8	6.9 ±2.4	0.30
Albumin (g/L)	31.1 ±6.2	33.2 ±6.0	0.42
Hémoglobin (g/dL)	10.9 ±1.5	11.2 ±2.0	0.38
Plaquettes (G/L)	429.4 ± 132.8	380.6 ±171.4	0.10
Cardiovascular risk factors, n (%)			
Obesity (BMI ≥30kg/m^2)	8 (11.0)	5 (11.4)	1.00
Arterial hypertension	30 (41.1)	20 (45.5)	0.64
Diabetes mellitus	5 (6.8)	2 (4.5)	0.71
Hypercholesterolemia	18 (24.7)	7 (15.9)	0.26
Ever smoker	11 (15.1)	19 (43.2)	**0.0007**
Location of aortic involvement, n (%)			
Ascending aorta	48 (65.8)	29 (65.9)	1.00
Arch	55 (75.3)	32 (72.7)	0.75
Descending aorta	6 (84.9)	32 (72.7)	0.15
Suprarenal abdominal aorta	46 (63.0)	20 (45.5)	0.06
Subrenal abdominal aorta l	45 (61.6)	20 (45.5)	0.09
Entire aorta	29 (39.7)	11 (25.0)	0.10
Type of aortic involvement as, n (%)			
Aneurysm, Ascending thoracic aneurysm Abdominal aneurysm	15 (20.5) 14 (19.2) 1 (1.4)	17 (38.6) 13 (39.5) 3 (6.8)	0.03 0.20 0.14
Ectasia	5 (6.8)	1 (2.3)	0.41
Wall thickening	70 (95.9)	41 (93.2)	0.67
Dissection	5 (6.8)	3 (6.8)	1.00
Aortic maximal thickening, mm, mean± SD	3.9 ±1.5	4.57 ±1.5	0.08
Aortic stenosis,	0 (0.0)	0 (0.0)	1.00
Aortic thrombosis	2 (2.7)	1 (2.3)	1.00

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Table 1**:** Comparison of demographics, biologic parameters, cardiovascular risk factors and aortic findings on CT-scan between patients with GCA-related aortitis (GCA-Ao) and patients with IA, at the time of aortitis diagnosis.

Aortic CT-scan was performed during follow-up in 63% of GCA-related aortitis patients and in 68.2% of IA patients. The entire aortic complications identified in both groups are presented in table 2. Twenty-four IA patients (54.5%) underwent at least one aortic event as compared to 20 GCA-related aortitis patients (27.4%, p=0.003). Respectively, 4 patients with GCA-related aortitis and 6 patients with IA developed new aortic aneurysm during follow-up. Thus, a total of 41 patients (35%) had aortic aneurismal disease, which was significantly more frequent among IA patients (50% vs 26%, p=0.008). In 2 cases, aortic aneurysm occurred in IA patients who had not been specifically treated for aortitis.

	GCA-Ao n=73	**IA n=44**	***p*-value**
Aortic aneurysm, n (%)	19 (26.0)	22 (50.0)	**0.008**
Aortic ectasia, n (%)	3 (4.1)	2 (4.5)	1.00
Aortic dissection, n (%)	6 (8.2)	6 (13.1)	0.36
Aortic stenosis, n (%)	0 (0)	1 (2.3)	0.37
Aortic surgery, n (%)	10 (13.7)	16 (36.4)	**0.004**

Table 2: Aortic events identified at diagnosis and during follow-up (median follow-up=34 months) among 117 patients with aortitis, including 73 patients with GCA-related aortitis and 44 with idiopathic aortitis (IA).

Aortic surgery during follow-up was required in 4 patients with GCA (5.4%) and in 8 patients with IA (18%). Overall, aortic surgery (at diagnosis and during follow-up) was more frequently performed in the IA group than in the GCA-related aortitis group (36.4% vs 13.7%,

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p=0.004). Bentall procedure was performed in 17 cases (7 GCA, 11 IA), aortic endoprosthesis in 5 cases (3 GCA, 2 IA) and aortic repair in 3 IA patients. In 23 cases, (8 GCA, 15 IA), aortic pathological examination was available and giant cells in the aortic wall were noted in 17 patients (5 GCA, 12 IA).

At the end of the follow-up, 10 patients had died, 6 in the GCA group (8.2%), and 4 in the IA group (9.1%, NS). Median time between aortitis diagnosis and death was 26 months for GCA-related aortitis and 30 months for IA. Five patients (2 with GCA, 3 with IA) had relapsing inflammatory disease and uncontrolled aortitis. One IA patient died because of aortic aneurysm rupture.

As IA may represent a heterogeneous entity, we compared IA patients according to the age < or ≥60y. The proportion of women was higher among IA patients ≥60y (69% vs. 53.3%). Inflammatory parameters and cardiovascular risk factors except smoking were not different. Past/current smokers were predominantly found in patients with IA under 60y (66.7% vs. 31%, p=0.02). Aneurism, ectasia, wall thickening, and aortic dissection were similar in both groups. IA patients ≥60y had more descending aorta involvement (82.4%) than IA patients <60y (53.3%, p=0.04).

As IA≥60y may overlap with GCA-related aortitis, we then compared these both groups (table 3). Median age, sex ratio and biological inflammatory parameters were remarkably close. Cardiovascular risk factors were equally distributed. Proportions of patients with involvement of ascending aorta, descending aorta, and the entire aorta were also very close in both groups.

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	GCA n=73	**IA ≥ 60 y n=29**	*p*-value
Median age (years) (min-max)	70 (52-83)	69.8 (60-87)	0.90
Female, n (%)	57 (78.1)	20 (69.0)	0.19
Biological parameters ±SD			
Mean CRP mg/L	111.3 ±78.3	117.5 ±76.3	0.74
ESR, mm/h	70.5 ±38.1	65.6 ±30.9	0.71
Fibrinogen g/L	7.5 ±1.8	6.9 ±2.4	0.38
Albumin g/L	31.1 ±6.2	34.2 ±6.2	0.18
Hémoglobin g/dL	10.9 ±1.5	11.2 ±1.9	0.41
Plaquettes G/L	429.4 ±132.8	380.2 ±192.1	0.21
Cardiovascular risk factors, n (%)			
Obesity (BMI ≥30kg/m^2)	8 (11.0)	2 (6.9)	0.53
Arterial hypertension	30 (41.1)	14 (48.3)	0.78
Diabetes mellitus	5 (6.8)	1 (3.4)	0.67
Hypercholesterolemia	18 (24.7)	6 (20.7)	0.67
Ever smoker	11 (15.1)	9 (31.0)	0.07
Location of aortic involvement, n (%)			
Ascending aorta	48 (65.8)	19 (65.5)	0.33
Arch	55 (75.3)	21 (72.4)	0.76
Descending aorta	62 (84.9)	24 (82.4)	0.77
Abdominal aorta, suprarenal	46 (63.0)	14 (48.3)	0.17
Abdominal aorta, subrenal	45 (61.6)	11 (37.9)	**0.03**
Entire aorta	29 (39.7)	7 (24.1)	0.14
Type of aortic involvement, n (%)			
Aneurysm	15 (20.5)	14 (48.3)	**0.005**
Ectasia	5 (6.8)	1 (3.4)	0.51
Wall thickening	70 (95.9)	26 (89.7)	0.34
Dissection	5 (6.8)	2 (6.9)	1.00
Mean of maximum thickening, mm (± SD)	3.9 (±1.5)	4.7 (±1.8)	**0.03**
Aortic stenosis	0 (0.0)	0 (0.0)	1.00
Aortic thrombosis	2 (2.7)	0 (0.0)	1.00

Table 3: Comparison of demographics, biologic parameters, cardiovascular risk factors and aortic findings on CT-scan between patients with aortitis related to GCA (GCA-Ao) and patients with IA≥60y, at the time of aortitis diagnosis.

Further, we analyzed the free of aortic events long-term survival (figure 2). Among 85 patients (58 GCA, 27 IA) without aortic aneurism at aortitis diagnosis, subsequent development of aortic aneurysm was significantly lower in GCA when compared to IA (p=0.009) (figure 2A). Analysis of 101 patients free of aortic surgery at aortitis diagnosis (65

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GCA, 36 IA), showed that GCA patients required significantly less aortic surgery during follow-up than IA patients (p=0.02) (figure 2B). Among 109 patients (68 GCA, 41 IA) without aortic dissection at aortitis diagnosis, subsequent occurrence of aortic dissection was lower in GCA (figure 2C). Finally, the free of aortic aneurism survival was equivalent among GCA-related aortitis patients and IA≥60y patients (figure 1D). Moreover, this survival was significantly worse among IA patients<60y (p=0.003).

Discussion

Both IA and GCA-related aortitis are rare diseases for which epidemiological or therapeutic data are lacking. In the revised nomenclature of vasculitis, IA is considered separately [21], but the hypothesis that some IA patients over 50 or 60y do have GCA is often discussed in practice. Our study is the first to investigate clinical and imaging findings, outcome and prognosis of GCA-related aortitis and IA in a large French population. Our aortitis cases distinguish from previous surgical series describing aortic histological findings [3, 15, 22, 23].

Our study describes how GCA-related aortitis and IA were diagnosed. Weight loss, fever, inflammatory state were found in both conditions. Cough was more frequent among patients with GCA, which is not surprising, as it is a frequent manifestation of GCA [24]. The main finding was that clinical signs of aortic aneurismal disease like dyspnea, thoracic or abdominal pain were more frequent in IA. This is consistent with the fact that aortic aneurism were more frequent. Surprisingly, we found more smokers among IA patients and smoking could contribute to generate more severe aortic damages. In fact, we hypothesize that aortitis among GCA patients was found at an earlier stage, with less aneurism and lower aortic wall thickening. This can be explained by the systematic screening of aortic involvement in

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numerous patients with GCA [8, 12]. Such systematic screening is controversial but recently, the French research group on GCA recommended an aortic CT-scan for each patient at the time of diagnosis of GCA [25].

In GCA, the management of aortic involvement (initial dose regimen, duration, monitoring) remains to be determined [26-28]. The benefit of steroid therapy to prevent subsequent development of aortic aneurismal disease remains elusive in patients with aortic inflammatory thickening. To treat large large-vessel vasculitis, the EULAR (european league against rheumatism) recommends corticosteroid at 1 mg/kg/day of prednisone during 1 month, followed by a gradual decline [25, 29], associated with antiplatelet treatment and management of cardiovascular risk factors [7]. Moreover, blood pressure should be controlled, and the use of beta-blocker is recommended to reduce the growth of aneurysms. Antiplatelet therapy was given to 79 of the entire cohort (67.5%). The benefit of this therapy in patients with aortitis is not really demonstrated and a recent cumulative meta-analysis showed that antiplatelet/anticoagulant therapy prior to the diagnosis of GCA was not associated with reduction in severe ischemic complications [30]. However, such a therapy may prevent from severe ischemic events after the diagnosis of GCA [30].

Despite the absence of any subsequent systematic scheduled aortic monitoring, our study have described the occurrence of new aortic aneurism, in 4/73 cases of GCA and in 6/44 cases of IA. The incidence of new aortic aneurism is probably higher. As 2 of 4 untreated IA patients developed new aneurism, therapeutic abstention should be avoided. However, treatment with prednisone 0.7 or 1 mg/kg/day might be not sufficient to prevent aortic aneurysm occurrence.

Aortic involvement in GCA is potentially serious and the incidence of aortic aneurysm/dissection has been found to be increased 5 years after GCA diagnosis [31, 32]. Moreover, the survival of GCA patients with aortic aneurysm or dissection has been found to

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be decreased regarding standardized mortality ratio at 2.63 [31]. The prognosis of GCA patients with aortitis is unknown, but inflammatory thickening of the aortic wall may completely regress after 3-6 months steroids therapy [7, 13] and in some patients, aortitis might be totally cured. We previously found that these patients might have more GCA relapses, and higher cardiovascular mortality [14] but this remains to be prospectively demonstrated. Currently, predictive factors for aortic involvement in GCA and for severe aortic injury (dissection, aortic aneurysm) are unknown [33]. Through our study, it seems that outcome is worse in IA than it is in GCA-related aortitis. In this way, proportions of new aortic aneurism and of patients requiring aortic surgery were higher during IA. However, this distinction might be due to differences in inflammatory stage at the time of diagnosis.

Our study provides new informations regarding IA, which is an uncommon disorder characterized by giant cells or lymphoplasmacytic inflammation of the aorta; female gender, smoking, and older age are possible risk factors for the disease [15, 23]. IA appears as a multifocal and progressive aortic disease [23]. In patients with IA starting at the ascending aorta, subsequent distal aortic aneurysms and/or dissections occur in almost 50% of cases [34]. Studies specifically focusing on the long-term follow-up of ascending IA are very limited.

Herein, we particularly show that GCA-related aortitis and IA over 60y share numerous remarkable similarities in terms of age, inflammatory parameters, sex-ratio, location of aortic involvement and risk of development of new aortic aneurism. By the way, 17/44 IA patients were considered as having GCA by the clinicians. IA in patients over 60y should lead to perform TAB, and if negative, should be considered as authentic GCA with negative TAB. The term “IA” has sometimes been used to design aortitis occurring in patients with prior systemic disease. Among 52 cases of IA reported by Rojo-Leyva *et al.*, 4 had prior

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diagnosis of GCA (positive TAB in 2 cases), up to 8 years before aortitis [15]. The term GCA-related aortitis" would be more suitable in this situation.

On the opposite, IA under 60y affects males as much as females, and involvement of descending aorta is less frequent. Above all, the prognosis of IA under 60y seems to be worse regarding the development of aortic aneurism. The higher proportion of smokers in this sub-group of patients could explain this point.

Few studies comparing IA and GCA have been conducted so far. As in our study, Talarico *et al.* found that patients with IA were younger with a different male/female sex-ratio than the one usually noted in GCA [16]. However, aortitis was asserted by aortic abnormal [^{18}F] fluorodeoxyglucose PET uptake, and only 18% of the patients with GCA underwent aortic PET-scan to look for aortitis. Moreover, patients with "isolated aortitis" under 50y were excluded as they only reviewed medical notes of patients with suspicion of GCA.

Our study has several limits, mainly due to its retrospective design. Monitoring of aortitis was not performed in each patient. Our cohort is homogenous in that aortic CT-scan was the unique imaging technique to diagnose aortitis, but precise modalities of CT-scan may differ between radiologists and centers. Treatment of aortitis was not standardized and data are lacking about the duration, the decrease protocol, and the cumulative dose of steroids. Moreover, data regarding levels of inflammatory parameters during follow up are not provided. We hypothesize that uncontrolled aortitis may lead to aortic complications such aneurism but our data do not allow us to be affirmative. Management of cardiovascular risk factors, which is not precisely described in the follow-up of our patients, also represents an important issue to prevent such complications.

Conclusion

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This work provides new data regarding initial characteristics and outcome of patients with aortitis seen in French internal medicine units. GCA-related aortitis and IA are the main causes of aortitis. IA appears to be more severe at the time of diagnosis, with more frequent aortic aneurisms and higher aortic wall thickenings. The screening for aortitis at the time of diagnosis of GCA enables early diagnosis. From this perspective, we speculate that early diagnosis of aortitis would be a potential mean to better prevent aortic aneurism development, but this remains to be established. Our main finding is that IA in patients ≥60y share many similarities with GCA-related aortitis: mean age around 70y, predominance of women, identical inflammatory parameters, same risk for development of aortic complications. This raises the question of the potential place of aortitis as a criteria for the diagnosis of GCA.

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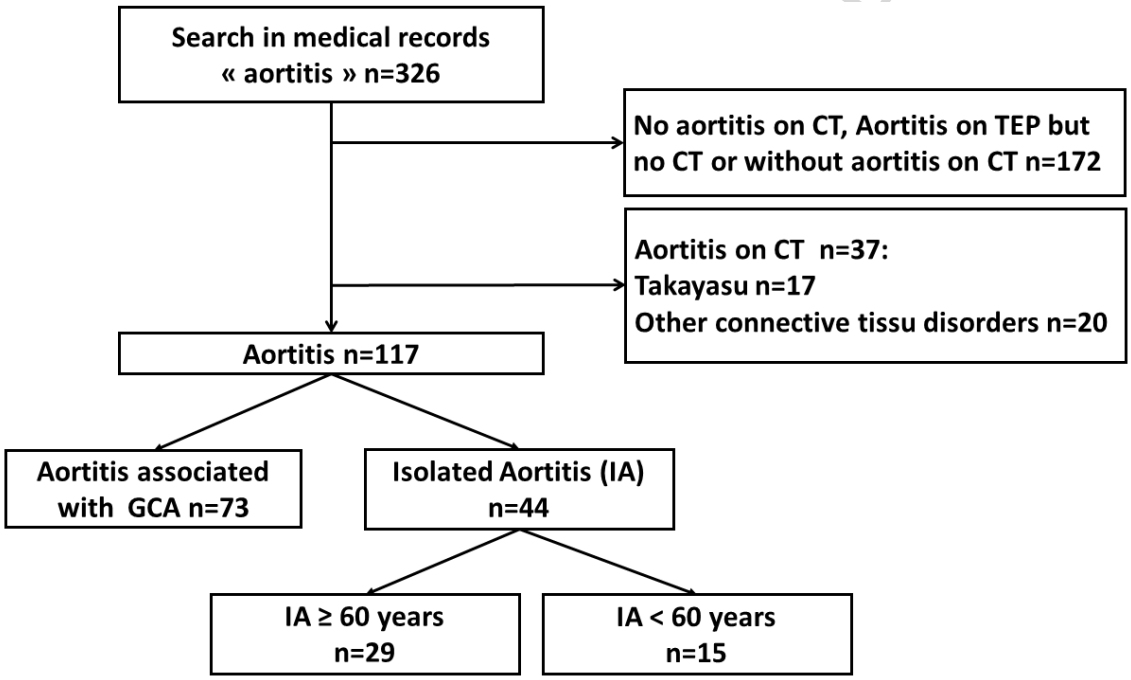
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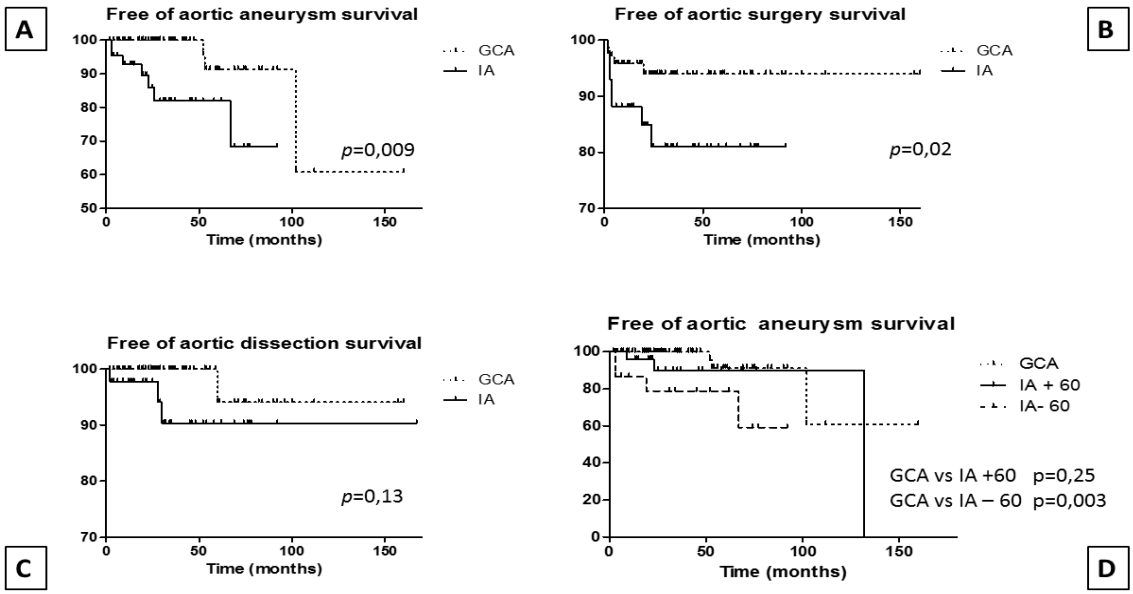
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Figure 1: flow chart of patient's selection.



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Figure 2: Long-term survival comparison of patients with GCA-related aortitis (GCA) and idiopathic aortitis (IA). In each type of aortic event (aortic aneurism, aortic surgery, aortic dissection), only subsequent aortic events are taken into account in patients free of the event at diagnosis. Figure 2A shows free of aortic aneurism survival, figure 2B shows free of aortic surgery survival, figure 2C shows free of aortic dissection survival. Free of aortic aneurysm survival in patients with GCA-related aortitis, IA≥60y and IA<60y is shown on figure 2D.



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Highlights

- At the time of diagnosis of aortitis, aortic aneurisms are more frequent and aortic wall is more thickened in patients with IA than in patients with GCA-related aortitis.
- IA represents a heterogenous group of patients, with numerous smokers, especially in patients under 60y.
- IA in patients≥60y share many similarities with GCA-related aortitis: mean age around 70y, predominance of women, identical inflammatory parameters, same risk for development of aortic complications.

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