Non-infectious aortitis: a report of 32 cases from a single tertiary centre in a 4-year period and literature review

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ABSTRACT

Objective. Non-infectious aortitis often presents with non-specific symptoms leading to inappropriate diagnostic delay. We intend to describe the clinical spectrum and outcome of patients with aortitis diagnosed at a single centre. Methods. We reviewed the clinical charts of patients diagnosed with non-infectious aortitis between January 2010 and December 2013 at the Rheumatology Division from a 1,000-bed tertiary teaching hospital from Northern Spain. The diagnosis of aortitis was usually based on FDG-PET-CT scan, and also occasionally on CT or MRI angiography or helical CT-scan. Results. During the period of assessment 32 patients (22 women and 10 men; mean age 68 years [range, 45–87]) were diagnosed with aortitis. The median interval from the onset of symptoms to the diagnosis was 21 months. FDG-PET CT scan was the most common tool used for the diagnosis of aortitis. The underlying conditions were the following: giant cell arteritis (n=13 cases); isolated polymyalgia rheumatica (PMR) (n=11); Sjögren’s syndrome (n=2), Takayasu arteritis (n=1); sarcoidosis (n=1), ulcerative colitis (n=1), psoriatic arthritis (n=1), and large-vessel vasculitis that also involved the aorta (n=2). The most common clinical manifestations at diagnosis were: PMR features, often with atypical clinical presentation (n=23 patients, 72%); diffuse lower limb pain (n=16 patients, 50%); constitutional symptoms (n=12 patients, 37%), inflammatory low back pain (n=9 patients, 28%) and fever (n=7 patients, 22%). Acute phase reactants were increased in most cases (median erythrocyte sedimentation rate 46 mm/hour, and a median serum C-reactive protein 1.5 mg/dL). Conclusion. Aortitis is not an uncommon condition. The diagnosis is often delayed. Atypical PMR features, unexplained low back or limb pain, constitutional symptoms along with increased acute phase reactants should be considered “red flags” to suspect the presence of aortitis.

Introduction
Aortitis is the inflammation of the aortic wall (1, 2), and can be idiopathic or associated with a cluster of infectious or non-infectious diseases (1-11). Giant cell arteritis (GCA) (12, 13) and Takayasu arteritis (TA) are the most common underlying conditions associated with aortitis (14, 15), although it can be a manifestation of other systemic diseases (16-23).
Non-infectious aortitis is often an underrecognised condition usually presenting with non-specific symptoms. Thus, high index of clinical suspicion is required (1, 2). Early diagnosis is of main importance to prevent serious complications, such as an aneurysmal rupture or an aortic dissection (24).
Aortitis is a hystopathological term (Fig. 1). However, in the clinical practice, the diagnosis is based on imaging techniques (25, 26). Thus, the introduction in the last decade of ¹⁸F(fluoro)-D-glucose (FDG) positron emission tomography (PET) computed tomography scan (FDG-PET/CT scan) and contrast-enhanced magnetic resonance imaging (MRI) (Fig. 2) has improved the diagnosis of large-vessel vasculitides (23, 26-30).
In the present study we aimed to: a) analyse the presenting features and clinical spectrum of patients with aortitis diagnosed at a single centre, and b) establish the clinical and laboratory data that may be considered as “red flags”, which may help to establish an early diagnosis of aortitis.
Patients and methods

Patient population

We reviewed the clinical charts of patients diagnosed with non-infectious aortitis between January 2010 and December 2013 at the Rheumatology Division from a 1,000-bed tertiary teaching hospital from Northern Spain. The diagnosis of aortitis was usually based on FDG-PET-CT scan, and also occasionally on CT or MRI angiography or helical CT-scan. In all the cases clinical features and/or laboratory abnormalities were also present.

GCA (31) and TA (32) were diagnosed according to the American College of Rheumatology classification criteria. Under the term “polymyalgia rheumatica” (PMR) we included patients with typical and atypical polymyalgia features. Typical PMR was defined when the patient fulfilled the PMR criteria proposed by Chuang et al. (33). On the other hand, we defined atypical PMR when patients did not fulfill the criteria for PMR shown above (33). These patients had aches and pain resembling PMR and at least one of the following features: a) inflammatory low back pain, b) diffuse pain in the lower limbs, c) persistent fever, d) constitutional symptoms and/or e) lack of improvement of PMR with low-medium dose oral corticosteroids (in all cases prednisone range: 10–15 mg/day). We also assessed whether patients with PMR features fulfilled the classification criteria proposed by EULAR/ACR 2012 (34).

Sjögren’s syndrome was diagnosed according to the European-American classification criteria (35), and psoriatic arthritis was diagnosed on the basis of CASPAR criteria (36).

Data collection and clinical definitions

Clinical and laboratory data were retrieved according to a pre-established research protocol. To minimise entry error, all the data were double-checked. Fever was defined as a temperature >38°C. Constitutional symptoms were asthenia, anorexia or weight loss greater than 5% of the normal body weight. Lower limb pain was defined as a diffuse pain involving the thighs. Inflammatory low back pain was diagnosed according to the Assessment of Spondyloarthritis International Society (ASAS) (37). ESR was considered increased when it was higher than 20 or 25 mm/1st hour for men or women, respectively. Serum CRP levels above 0.5 mg/dL were considered as high values.

FDG-PET CT scan acquisition and image analysis

Patients had to be in fasting state for at least 6 hours before the examination. Serum glucose level was lower than 160 mg/dL in all the patients. Whole-body FDG-PET uptake was assessed 180 minutes after injection of 7 MBq/kg of $^{18}F$-FDG, using a Biograph LSO Pico 3D from Siemens Healthcare Molecular Imaging (Hoffman Estates, IL, USA). A low dose CT scan for attenuation correction and anatomic localisation was first obtained, followed by a FDG-PET scan (acquiring 250 s/bed position). Images were reconstructed using the ordered subsets-expectation maximisation (OSEM) algorithm (2 iterations, 8 subsets). Images were visually evaluated by two experienced nuclear medicine specialists according to the intensity of the $^{18}$F-FDG uptake by the vessel wall at the supraaortic trunks, thoracic aorta, abdominal aorta, iliac arteries and femoral/tibioperoneal arteries.

Statistical analysis

Results were expressed as mean ± standard deviation (SD) or as median, range and/or interquartile range (IQR) as appropriate. Analysis was performed with the STATISTICA software package (Statsoft Inc. Tulsa, OK, USA).

Results

Overall results

We studied 32 patients (22 women/10 men) with non-infectious aortitis. The mean age±SD at the time of diagnosis was 68±11 years (range 45–87 years).
Table I. Main features of 32 patients finally diagnosed as having aortitis.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age/sex</th>
<th>Associated disease</th>
<th>Delay to the diagnosis of aortitis (months)</th>
<th>Fever</th>
<th>Constitutional symptoms</th>
<th>Inflammatory low back pain</th>
<th>Lower limb pain</th>
<th>PMR</th>
<th>ESR/CRP at the time of aortitis diagnosis</th>
<th>Temporal artery biopsy</th>
<th>Imaging technique and result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>56/F</td>
<td>GCA, PMR</td>
<td>6/5</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Typical</td>
<td>NP</td>
<td>PET-CT: supraaortic vessels, left common iliac artery, femoral and peroneal arteries</td>
</tr>
<tr>
<td>2</td>
<td>59/M</td>
<td>GCA, PMR</td>
<td>2/0</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>PET-CT: aorta, supraaortic vessels and both common carotid arteries</td>
</tr>
<tr>
<td>3</td>
<td>68/M</td>
<td>GCA</td>
<td>4/0</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>PET-CT: supraaortic vessels and subclavian arteries</td>
</tr>
<tr>
<td>4</td>
<td>69/F</td>
<td>GCA, PMR</td>
<td>0/3</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>Atypical</td>
<td>12/28.2</td>
<td>Positive Sci: along the venous-arterial route of both popliteal fossae</td>
</tr>
<tr>
<td>5</td>
<td>70/F</td>
<td>GCA, PMR</td>
<td>36/0</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>PET-CT: thoracic aorta, supraaortic vessels, medium and large vessels of lower limbs</td>
</tr>
<tr>
<td>6</td>
<td>63/F</td>
<td>GCA, PMR</td>
<td>77/72</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>PET-CT: descending thoracic aorta and abdominal aorta</td>
</tr>
<tr>
<td>7</td>
<td>65/F</td>
<td>GCA, PMR</td>
<td>18/15</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
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<td>PET-CT: aorta, supraaortic vessels, subclavian arteries and both common carotid arteries</td>
</tr>
<tr>
<td>8</td>
<td>72/F</td>
<td>GCA, PMR</td>
<td>31/30</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>PET-CT: ascending and descending aorta, aortic arch, supraaortic vessels, iliac and femoral arteries</td>
</tr>
<tr>
<td>9</td>
<td>73/F</td>
<td>GCA</td>
<td>100/99</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>PET-CT: thoracic aorta, thoracic and abdominal aorta and iliac arteries</td>
</tr>
<tr>
<td>10</td>
<td>79/F</td>
<td>GCA, PMR</td>
<td>47/23</td>
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<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Typical</td>
<td>No</td>
<td>PET-CT: aortic arch, thoracic and abdominal aorta</td>
</tr>
<tr>
<td>11</td>
<td>81/F</td>
<td>GCA, PMR</td>
<td>27/17</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Atypical</td>
<td>No</td>
<td>PET-CT: ascending aorta, aortic arch, thoraco-abdominal aorta, supraaortic vessels, femoral, popliteal and peroneal arteries</td>
</tr>
<tr>
<td>12</td>
<td>82/M</td>
<td>GCA, PMR</td>
<td>10/8</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>PET-CT: large vessels, including supraaortic, thoracic and abdominal aorta and bifurcation of iliac and femoral arteries</td>
</tr>
<tr>
<td>13</td>
<td>85/F</td>
<td>GCA</td>
<td>21/19</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>PET-CT: ascending and descending aorta and aortic arch</td>
</tr>
<tr>
<td>14</td>
<td>52/F</td>
<td>Isolated atypical PMR</td>
<td>8/2</td>
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<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Atypical</td>
<td>Yes</td>
<td>PET-CT: thoracic aorta and supraaortic vessels</td>
</tr>
<tr>
<td>15</td>
<td>56/F</td>
<td>Isolated atypical PMR</td>
<td>14/2</td>
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<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Atypical</td>
<td>No</td>
<td>PET-CT: thoracic aorta and supraaortic vessels</td>
</tr>
<tr>
<td>16</td>
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<td>9/6</td>
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<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Typical</td>
<td>No</td>
<td>PET-CT: thoracic aorta and supraaortic vessels</td>
</tr>
<tr>
<td>17</td>
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<td>Isolated atypical PMR</td>
<td>120/119</td>
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<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>PET-CT: ascending aorta, aortic arch, femoral and popliteal arteries</td>
</tr>
<tr>
<td>18</td>
<td>76/M</td>
<td>Isolated atypical PMR</td>
<td>3/0</td>
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<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>PET-CT: thoracic aorta, supraaortic vessels and pulmonary artery trunk</td>
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<tr>
<td>19</td>
<td>53/F</td>
<td>Typical PMR</td>
<td>27/26</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>PET-CT: aortic arch</td>
</tr>
<tr>
<td>20</td>
<td>62/M</td>
<td>Isolated PMR</td>
<td>2/0</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
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</tr>
<tr>
<td>21</td>
<td>65/M</td>
<td>Isolated PMR</td>
<td>ND/ND</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
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<tr>
<td>22</td>
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<td>Isolated PMR</td>
<td>11/8</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
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<td>PET-CT: supraaortic vessels and thoracic aorta</td>
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<tr>
<td>23</td>
<td>79/F</td>
<td>Isolated typical PMR</td>
<td>13/1</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
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<td>No</td>
<td>No</td>
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<tr>
<td>24</td>
<td>81/F</td>
<td>Isolated PMR</td>
<td>142/136</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>PET-CT: ascending aorta and aortic arch</td>
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<tr>
<td>25</td>
<td>71/F</td>
<td>Sjögren’s syndrome</td>
<td>180/170</td>
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<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>PET-CT: without inflammatory activity</td>
</tr>
<tr>
<td>26</td>
<td>79/F</td>
<td>Sjögren’s syndrome</td>
<td>69/9</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>PET-CT: thoracic aorta and supraaortic vessels</td>
</tr>
<tr>
<td>27</td>
<td>48/F</td>
<td>TA</td>
<td>150/0</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>PET-CT: thoracic aorta</td>
</tr>
<tr>
<td>28</td>
<td>56/M</td>
<td>Sarcoidosis, PMR</td>
<td>44/39</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>PET-CT: ascending and descending aorta, aortic arch, femoral and peroneal arteries</td>
</tr>
<tr>
<td>29</td>
<td>69/F</td>
<td>Sarcoidosis, PMR</td>
<td>26/24</td>
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<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>12/0.1</td>
<td>No</td>
<td>PET-CT: thoracic aorta and femoral and peroneal arteries</td>
</tr>
<tr>
<td>30</td>
<td>45/M</td>
<td>Psoriatic arthritis</td>
<td>156/71</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>4/0.1</td>
<td>No</td>
<td>PET-CT: thoracic aorta and supraaortic vessels</td>
</tr>
<tr>
<td>31</td>
<td>64/M</td>
<td>Idiopathic</td>
<td>2/0</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>104/12.6</td>
<td>No</td>
<td>PET-CT: thoracic and abdominal aorta, supraaortic vessels and iliac arteries</td>
</tr>
<tr>
<td>32</td>
<td>87/M</td>
<td>Idiopathic</td>
<td>4/0</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>110/12.4</td>
<td>No</td>
<td>PET-CT: thoracic aorta, supraaortic vessels and large vessels of lower limbs</td>
</tr>
</tbody>
</table>

M: male; F: female; GCA: giant cell arteritis; PMR: polymyalgia rheumatica; TA: Takayasu arteritis; CT: computed tomography; MRA: magnetic resonance angiography; PET: 18FDG positron emission tomography. NP: Not performed; ESR: Erythrocyte sedimentation rate (mm/1st hour); CRP: C-reactive protein (mg/dL).
Table II. Main features of 13 patients with giant cell arteritis (GCA) and aortitis.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (yrs) / sex (at aortitis diagnosis)</th>
<th>Headache* / Jaw claudication</th>
<th>Visual symptoms</th>
<th>PMR</th>
<th>Abnormal temporal artery exploration</th>
<th>ESR / CRP</th>
<th>Temporal artery biopsy</th>
<th>Initial prednisone dose / response at first month</th>
<th>Clinical features</th>
<th>ESR / CRP</th>
<th>Time from GCA to aortitis diagnosis (months)</th>
<th>Initial prednisone dose and other therapy / response at 1 month</th>
<th>Other therapy in follow-up</th>
<th>Follow-up (months)</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>56/F</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>75/ND</td>
<td>NP</td>
<td>40/PclinR, ClabR</td>
<td>Fever, ILBP, ComS</td>
<td>-</td>
<td>Synchronous</td>
<td>MTX 10 mg/w</td>
<td>36</td>
<td>CclinR, ClabR</td>
</tr>
<tr>
<td>2</td>
<td>59/M</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>67/13.4</td>
<td>Negative</td>
<td>40/CclinR, ClabR</td>
<td>Fever, sweating, ComS</td>
<td>-</td>
<td>Synchronous</td>
<td>-</td>
<td>36</td>
<td>CclinR, ClabR</td>
</tr>
<tr>
<td>3</td>
<td>68/M</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>101/11.75</td>
<td>Positive</td>
<td>50/PclinR, PlabR</td>
<td>Malaise, fever, sweating, ComS</td>
<td>-</td>
<td>Synchronous</td>
<td>MTX 15 mg/w</td>
<td>66</td>
<td>CclinR, PlabR</td>
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<tr>
<td>4</td>
<td>69/F</td>
<td>No</td>
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<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>126/28.2</td>
<td>Positive</td>
<td>45/PclinR, PlabR</td>
<td>Fever, pain in thighs, diffuse pain in lower limbs</td>
<td>-</td>
<td>Synchronous</td>
<td>MTX 15 mg/w</td>
<td>24</td>
<td>CclinR, ClabR</td>
</tr>
<tr>
<td>5</td>
<td>70/F</td>
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<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>78/3.3</td>
<td>Negative</td>
<td>30/CclinR, ClabR</td>
<td>ComS</td>
<td>-</td>
<td>Synchronous</td>
<td>-</td>
<td>3</td>
<td>CclinR, ClabR</td>
</tr>
<tr>
<td>6</td>
<td>63/F</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>131/24</td>
<td>Positive</td>
<td>50/CclinR, PlabR</td>
<td>Chest pain</td>
<td>4.0/1.6</td>
<td>72</td>
<td>45/CclinR, ClabR</td>
<td>MTX 15 mg/w</td>
<td>6</td>
</tr>
<tr>
<td>7</td>
<td>65/F</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>108/13.2</td>
<td>Positive</td>
<td>40/PclinR, ClabR</td>
<td>Headache and loss of weight when prednisone is tapered below 25 mg/d</td>
<td>12.0/9</td>
<td>6</td>
<td>25+TCZ (8 mg/kg/m) / PclinR, ClabR</td>
<td>-</td>
<td>36</td>
</tr>
<tr>
<td>8</td>
<td>72/F</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>46/1.9</td>
<td>Positive</td>
<td>40/CclinR, ClabR</td>
<td>Headache, pain in scapular guirdle when prednisone is tapered below 10 mg/d</td>
<td>100/15</td>
<td>30</td>
<td>10+ MTX 7.5 / PclinR, ClabR</td>
<td>-</td>
<td>7</td>
</tr>
<tr>
<td>9</td>
<td>73/F</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>85/4.6</td>
<td>Positive</td>
<td>30/CclinR, ClabR</td>
<td>Asthenia</td>
<td>46/ND</td>
<td>99</td>
<td>TCZ (8mg/km) / CclinR, ClabR</td>
<td>-</td>
<td>20</td>
</tr>
<tr>
<td>10</td>
<td>79/F</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>84/ND</td>
<td>Positive</td>
<td>20/PclinR, PlabR</td>
<td>ComS, PMR, headache, ashenia</td>
<td>32/1.2</td>
<td>35</td>
<td>7.5+MTX (12.5mg/w) / CclinR, ClabR</td>
<td>-</td>
<td>5</td>
</tr>
<tr>
<td>11</td>
<td>81/F</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>76/2.92</td>
<td>Positive</td>
<td>45/CclinR, ClabR</td>
<td>Asymptomatic</td>
<td>43/0.2</td>
<td>17</td>
<td>20+ MTX10 / CclinR, PlabR</td>
<td>-</td>
<td>7</td>
</tr>
<tr>
<td>12</td>
<td>82/M</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>49/5.5</td>
<td>Negative</td>
<td>10/CclinR, ClabR</td>
<td>Asymptomatic but 1 of acute phase reactants</td>
<td>119/1.6</td>
<td>8</td>
<td>40/CclinR, ClabR</td>
<td>-</td>
<td>15</td>
</tr>
<tr>
<td>13</td>
<td>85/F</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>113/1.4</td>
<td>Positive</td>
<td>45/CclinR, ClabR</td>
<td>ILBP and lower limbs pain</td>
<td>27/1.2</td>
<td>19</td>
<td>7.5+MTX (25 mg/w) / PclinR, PlabR</td>
<td>-</td>
<td>34</td>
</tr>
</tbody>
</table>

*Headache: recent onset or different from usual.
F: female; M: male; CclinR: complete clinical response; PclinR: partial clinical response; ClabR: complete laboratory response; PlabR: partial laboratory response; MTX: methotrexate; ILBP: inflammatory low back pain; ComS: constitutional symptoms; ESR: Erythrocyte sedimentation rate (mm/1st hour); CRP: C-reactive protein (mg/dL); NP: Not performed.
The median interval from the onset of the symptoms to the diagnosis of aortitis was 21 (IQR: 6–69) (range 0.3–180) months. Table I summarises the main features of the patients included in our series.

**Clinical subtypes of aortitis**

- **Giant cell arteritis and aortitis**
  We diagnosed 13 patients (10 women/3 men) with aortitis associated with GCA. The mean age±SD was 71±9 years (range 56–85 years). GCA was histologically confirmed in 9 of 13 cases. The main clinical and laboratory characteristics of these patients are summarised in Table II. Of note, in 8 cases initially diagnosed of GCA, the diagnosis of aortitis was made later (median [IQR]: 24.5 (8–72) months; range 6–99 months).

- **Typical and atypical polymyalgia rheumatica associated with aortitis**
  Twenty-three patients had PMR (typical in 15 cases). It was isolated in 11 (typical in 6 cases and atypical in 5), and associated with other conditions in the remaining (10 cases with GCA, 1 with sarcoidosis and 1 with ulcerative colitis).

  Patients (8 women and 3 men) with isolated PMR had a mean age of 67±10 years (range 52–81 years). Table III shows the main clinical features and laboratory parameters of patients with isolated PMR. Noteworthy, patients with atypical PMR experienced, more commonly, symptoms such as thigh pain, low back pain or lower limb pain, and they had a poor response to low-medium doses of prednisone (10–15 mg/day). Besides, none of the 5 patients with isolated atypical PMR fulfilled the ACR/EULAR criteria (34).

- **Sjögren’s syndrome and aortitis**
  Two patients were diagnosed with Sjögren’s syndrome and aortitis. The first one was a 71-year-old woman diagnosed with Sjögren’s syndrome in 1997. Aortitis was suspected because persistently high ESR levels despite treatment with low-dose corticosteroids. A FDG-PET CT scan showed increased vascular uptake with a typical pattern suggestive of aortitis. Rituximab (375 mg/m² weekly for 4 weeks) was added with an excellent clinical response at 3 months.

  The second patient was a 79-year-old woman diagnosed with Sjögren’s syndrome in 1997 and with persistently elevated ESR and anaemia. The patient underwent a FDG-PET CT scan that disclosed an increased FDG uptake in the thoracic aortic wall consistent with aortitis.

- **Aortitis related to other diseases**
  We found a patient with Leriche syndrome and exertional angina pectoris in which TA was diagnosed on the basis of raised acute phase reactants and an arteriography with high-grade stenosis in the infrarenal aorta, dilation of the arc of Riolan and 50% left subclavian artery stenosis. She was started on prednisone (30 mg/day) and MTX (15 mg/week). Normalisation of acute phase reactants and resolution of asthenia was achieved 2 months later. Then, prednisone was tapered until complete discontinuation one year later. Sarcoidosis and ulcerative colitis associated with aortitis was diagnosed in two patients previously reported (16, 17).

  Finally, a 45-year-old man with psoriatic arthritis and aortitis was diagnosed in our centre. He suffered a stroke and then began with diffuse pain in lower limbs. Because of the lower limb pain and the previous history of stroke, a large-vessel vasculitis was suspected and confirmed by increased FDG uptake in the thoracic aortic wall consistent with aortitis.

- **Large-vessel vasculitis that also involved the aorta**
  Two cases of large-vessel vasculitis that also involved the aorta were included in this series. The first one was...
Table III. Main features of 11 patients with isolated typical and atypical polymyalgia rheumatica (PMR) and aortitis.

<table>
<thead>
<tr>
<th>Case*</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Cervical pain</th>
<th>Scapular pain</th>
<th>Pelvic pain</th>
<th>Other features</th>
<th>ESR / CRP</th>
<th>Typical or atypical PMR</th>
<th>Initial prednisone dose / response at 1 month</th>
<th>Features at PMR diagnosis</th>
<th>Features at the time of aortitis diagnosis</th>
<th>Aortitis: therapy and outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>ESR / CRP</td>
<td>Typical or atypical PMR</td>
<td>Initial prednisone dose / response at 1 month</td>
<td>Clinical features</td>
<td>ESR / CRP</td>
<td>Time from PMR diagnosis to aortitis diagnosis (months)</td>
<td>Prednisone dose and other therapy at 1 month</td>
</tr>
<tr>
<td>14</td>
<td>52/F</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>87/1.1</td>
<td>Atypical</td>
<td>10/ No clinical improvement and PlabR</td>
<td>Pain in thighs and shoulder girdle</td>
<td>530.9</td>
<td>2</td>
<td>10/CclinR, PlabR</td>
</tr>
<tr>
<td>15</td>
<td>56/F</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>33/ND</td>
<td>Atypical</td>
<td>No corticosteroids/ No improvement</td>
<td>Pain in thighs, lower limbs and arms</td>
<td>43/ND</td>
<td>2</td>
<td>20/PclinR, PlabR. because the patient discontinued the treatment for fear of side effects</td>
</tr>
<tr>
<td>16</td>
<td>63/F</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>980/1.1</td>
<td>Atypical</td>
<td>10/CclinR, ClabR</td>
<td>Pain in thighs, ILBP and abdominal pain</td>
<td>250.1</td>
<td>6</td>
<td>5/PclinR, PlabR</td>
</tr>
<tr>
<td>17</td>
<td>68/M</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>100.1</td>
<td>Atypical</td>
<td>10/CclinR, ClabR</td>
<td>Pain in thighs</td>
<td>20.1</td>
<td>119</td>
<td>40/CclinR, ClabR</td>
</tr>
<tr>
<td>18</td>
<td>76/M</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>62/2.11</td>
<td>Atypical</td>
<td>30/CclinR, PlabR</td>
<td>ConsS</td>
<td>622/1.1</td>
<td>Synchronous</td>
<td>30/CclinR, PlabR</td>
</tr>
<tr>
<td>19</td>
<td>53/F</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>59/2.9</td>
<td>Typical</td>
<td>10/PclinR, PlabR</td>
<td>Pain in thighs</td>
<td>460.4</td>
<td>26</td>
<td>40/CclinR, ClabR</td>
</tr>
<tr>
<td>20</td>
<td>62/M</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>70/7.7</td>
<td>Typical</td>
<td>20/PclinR, PlabR</td>
<td>Shivering, shore throat</td>
<td>707/7</td>
<td>Synchronous</td>
<td>20/PclinR, PlabR</td>
</tr>
<tr>
<td>21</td>
<td>65/F</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>ILBP</td>
<td>65/5.5</td>
<td>Typical</td>
<td>5/PclinR, PlabR</td>
<td>ILBP</td>
<td>65/5.5</td>
<td>Synchronous</td>
<td>5/PclinR, PlabR</td>
</tr>
<tr>
<td>22</td>
<td>79/F</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>Headache</td>
<td>30/1.9</td>
<td>Typical</td>
<td>10/CclinR, ClabR</td>
<td>Reactivation of the symptoms when prednisone is tapered</td>
<td>25/1.5</td>
<td>8</td>
<td>6.25/PclinR, PlabR</td>
</tr>
<tr>
<td>23</td>
<td>79/F</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>Muscle pain in thighs, headache</td>
<td>330.6</td>
<td>Typical</td>
<td>20/PclinR, ClabR</td>
<td>Muscle pain in thighs</td>
<td>330.6</td>
<td>Synchronous</td>
<td>20/PclinR, ClabR</td>
</tr>
<tr>
<td>24</td>
<td>81/F</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>ConsS</td>
<td>28/2.2</td>
<td>Typical</td>
<td>10/CclinR, PlabR</td>
<td>Muscle pain in cervical, shoulder and pelvic girdles</td>
<td>78/2.1</td>
<td>136</td>
<td>30/PclinR, PlabR</td>
</tr>
</tbody>
</table>

* Related to case number from Table I.
F: female; M: male; CclinR: complete clinical response; PclinR: partial clinical response; ClabR: complete laboratory response; PlabR: partial laboratory response; MTX: methotrexate; ILBP: inflammatory low back pain; ConsS: constitutional symptoms. HQ: hidroxichloroquine; ESR: Erythrocyte sedimentation rate (mm/1st hour); CRP: C-reactive protein (mg/dL); Prednisone dose: mg/day; NP: Not performed.
a 64-year-old man presented with high-grade fever and dyspnea. He had mild leukocytosis, ESR 104 mm/1st hour and serum CRP 12.6 mg/dL. Complete microbiological studies were all negative. Temporal artery biopsy was normal. A body CT-scan disclosed slightly enlarged left paratracheal lymph nodes, and a biopsy showed reactive lymphadenitis. A 18F-FDG PET/CT scan revealed an increased uptake in the thoracic and abdominal aorta, supraaortic vessels and iliac arteries. The patient started on prednisone (40 mg/day) and MTX (10 mg/week) reaching complete clinical remission.

The second patient was an 87-year-old man who was sent to the hospital because of chest wall pain and inflammatory low back pain as well as asthenia, anorexia and weight loss of 3 months’ duration. The ESR was 110 mm/1st hour and serum CRP 12.6 mg/dL. Complete microbiological studies were all negative. Temporal artery biopsy was normal. A body CT-scan disclosed slightly enlarged left paratracheal lymph nodes, and a biopsy showed reactive lymphadenitis. A 18F-FDG PET/CT scan revealed an increased uptake in the thoracic and abdominal aorta, supraaortic vessels and iliac arteries. The patient started on prednisone (30 mg/day) and MTX (10 mg/week) reaching complete clinical remission.

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Non-infectious aortitis presents with a wide range of symptoms. Apart from fever, headache, back pain, and PMR manifestations (38), severe aortic insufficiency, aortic aneurysms and rarely, aortic dissection and rupture have been described at the time of disease diagnosis (1, 13, 39, 40). In our series, the most common clinical manifestations of aortitis were PMR, diffuse lower limb pain, constitutional symptoms, inflammatory low back pain, and fever. Most of these manifestations are non-specific. Therefore, a high index of suspicion for aortitis is needed. Thus, in patients with chronic or relapsing inflammatory diseases, these clinical features along with elevated acute phase reactants should be considered as “red flags” to suspect the presence of aortitis (Fig. 4).

Regarding imaging techniques, angiography has been replaced by angioCT-scan and MRI in the diagnosis and assessment of distribution and activity of aortitis (23, 41-43). MRI is recommended in patients in whom close follow-up is needed as this technique is very useful to assess wall thickening and wall oedema, and to determine the presence of specific lesions such as the presence of aneurysms or thrombus, (1, 41, 42, 46). In recent years, however, FDG-PET CT scan has gained considerable acceptability as a non-invasive tool potentially useful for the diagnosis and management of patients with large-vessel vasculitis by providing a metabolic functional image of the vessel wall inflammation before structural changes are seen (47-52). This technique is especially helpful in atypical presentations of vasculitis (17, 53). Previous studies have shown good results evaluating the aortic involvement in patients with large-vessel vasculitis (28).

GCA is more common in Caucasian individuals older than 50 years (54-57), and may affect the aorta and its major branches (12, 13, 29, 58-63). Table IV summarises the main series on patients with GCA and aortic involvement. The use of new imaging techniques has disclosed that extracranial large-vessel involvement in GCA is more common than initially thought (26). Aortitis in the setting of GCA is a potentially serious and usually underdiagnosed complication (64). A prospective study using CT-scan within the month after GCA diagnosis showed that aortic thickening occurs frequently at the time of diagnosis of GCA, and that this condition predominantly affects the ascending aorta (65). In keeping with these observations, using CT angiography Prieto-González et al. found aortitis in 67% of 40 newly diagnosed GCA patients (66). Histopathology studies disclosed ascending aorta involvement in 8–13% of GCA patients (13, 61, 62). Aortitis in patients with GCA may lead to aortic aneurysms. The risk of aneurysmal rupture and aortic dissection is increased in patients with GCA. They occur mostly in the ascending thoracic aorta. Comparing with people of the same age and sex, Evans et al. reported that patients with GCA had a 17-fold increased risk of thoracic aortic aneurysm and a 2.4-fold increased risk of abdominal aortic aneurysm (39). Simi-
Table V. Review of published cases of polymyalgia rheumatica and involvement of the aorta and/or its major branches.

<table>
<thead>
<tr>
<th>Reference</th>
<th>n/sex</th>
<th>Age (years)</th>
<th>Fever (n)</th>
<th>Constitutional symptoms (n)</th>
<th>PMR (n)</th>
<th>Headache (n)</th>
<th>Abnormal arterial on physical exam (n)</th>
<th>Jaw claudication (n)</th>
<th>Visual manifestations (n)</th>
<th>ESR</th>
<th>Positive temporal artery biopsy (n)</th>
<th>Imaging technique</th>
<th>Response to corticosteroids (initial dose)</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kataoka et al. (74)</td>
<td>1/M</td>
<td>61</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>120</td>
<td>11.57</td>
<td>Yes (prednisolone, 30 mg/day)</td>
<td>CT and MRA</td>
<td>Rapid resolution</td>
<td></td>
</tr>
<tr>
<td>Kataoka et al. (74)</td>
<td>1/F</td>
<td>63</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>90</td>
<td>6.07</td>
<td>Yes (prednisolone, 15 mg/day)</td>
<td>CT and MRA</td>
<td>Relapse</td>
<td></td>
</tr>
<tr>
<td>Koga et al. (75)</td>
<td>1/F</td>
<td>64</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>105</td>
<td>12.6</td>
<td>Yes (prednisolone, 40 mg/day)</td>
<td>CT</td>
<td>6 months later the patient required surgery for a dissecting aortic aneurysm, recovering without complications</td>
<td></td>
</tr>
<tr>
<td>Narváez et al. (29)</td>
<td>1/F</td>
<td>73</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>82</td>
<td>40</td>
<td>Yes (prednisolone, 20 mg/day)</td>
<td>MRI</td>
<td>Treatment failure</td>
<td></td>
</tr>
<tr>
<td>Narváez et al. (29)</td>
<td>1/F</td>
<td>67</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>42</td>
<td>19</td>
<td>Yes (prednisolone, 20 mg/day)</td>
<td>MRI</td>
<td>Treatment failure</td>
<td></td>
</tr>
<tr>
<td>Mückert et al. (76)</td>
<td>1/F</td>
<td>66</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>Yes (methylprednisolone, 500 mg/day)</td>
<td>CT</td>
<td>Rapid resolution</td>
<td></td>
</tr>
</tbody>
</table>

M: male; F: female; CT: computed tomography; MRA: magnetic resonance angiography; MRI: magnetic resonance imaging; PET: positron emission tomography; US: ultrasonography; NA: not available. ESR: erythrocyte sedimentation rate (mm/1st hour); CRP: C-reactive protein (mg/dL).
lar data have been reported in biopsy-proven GCA patients from Northwest Spain (61). Of note, Liozon et al. indicated that the presence of classic cranial features of GCA at the time of disease diagnosis may be a negative predictor of an aortic complication (67).

Overall, patients with large-vessel involvement in the setting of GCA are younger than patients with classic cranial temporal arteritis (66 vs. 72 years) and most of them are women (83% vs. 66%). Moreover, the time between the onset of symptoms and the diagnosis of the disease is usually longer in patients with aortitis (7 months vs. 2 months) (26).

Patients with large-vessel GCA may present with typical cranial temporal arteritis or PMR, but they may also present with non-specific symptoms such as fever, sweating, malaise, anorexia, weight loss, fatigue, lower extremity claudication or arthralgias (26, 42, 68). Therefore, in many GCA patients the diagnosis of aortitis may be extremely difficult, leading to a long diagnostic delay. Thus, in a series of biopsy-proven GCA patients from Northwest Spain, the mean time between the diagnosis of GCA and the diagnosis of aortic involvement was 57 months (61). Similar results have been reported in other studies (29, 58).

Aortic complications may be the cause of death in 3–12% of patients with large-vessel GCA (13). A recent epidemiologic study on large-vessel involvement in GCA patients from Olmsted County (MN) has shown that incidence of any large-vessel event is high within the first year of GCA diagnosis. The incidence of aortic aneurysm/dissection is also increased 5 years after GCA diagnosis (69). Survival of GCA patients with aortic aneurysm or dissection was found decreased (standardised mortality ratio, 2.63; 95%CI 1.78–3.73) (69).

PMR is also a common disorder in individuals older than 50 years from Western countries (55, 56). This may appear as an isolated disease or associated with ischemic features of GCA (70, 71). In the literature a handful of case reports emphasise the presence of aortitis in patients with PMR (29, 57, 70, 72-76). Table V summarises the lit-
erature cases of PMR with involvement of aorta and/or its major branches. The diagnosis of PMR is very straightforward when typical features, such as pain in the neck, and shoulder and pelvic girdles are present (77, 78). However, the presence of atypical symptoms or a poor response to corticosteroids should be considered warning signs for the presence of a condition mimicking this disease but different from isolated and typical PMR (79). It was also the case for some of our patients diagnosed with aortitis. Compared to the vast majority of patients with PMR reported in clinical studies, complications of large-vessel vasculitis in PMR patients are extremely rare. A search for aortitis should only be undertaken in PMR under special circumstances. This may be the case if atypical findings such as low back pain or pain involving mainly the legs associated to elevation of acute phase reactants are present.

TA is a large-vessel vasculitis uncommon in Western countries (54). However, it has been widely described in people from Asia, Africa and Latin America (80). TA affects commonly young people (81). The aorta, carotid and subclavian are the arteries most frequently involved (82, 83). The development of arterial stenosis and aneurysms leads to claudication, bruises, limb pain, and diminished or even absent pulses (84). Sometimes patients may also complain of visual loss or stroke, constitutional symptoms, fever, malaise, weight loss or anorexia (84). Table VI shows a set of series of patients with TA in which information on major branch involvement is described (82, 83, 85, 86). In our study, we only disclosed one patient with multiple arterial stenosis features and complications related to severe vascular involvement in the setting of a delayed diagnosis of TA.

Aortitis may be associated with other diseases. It may occur in the setting of sarcoidosis, a multisystemic disease that involves the lungs, eyes and skin (87). Sarcoidosis may coexist with rheumatic diseases including PMR (16, 88). Although uncommonly reported (89), sarcoidosis may be associated with vascular involvement (90-96), mostly with small-vessel vasculitis (90). Sarcoidosis has also been associated with Wegener’s granulomatosis, microscopic polyangiitis, polyanarthitis nodosa and TA (19, 90, 96). Association of sarcoidosis with aortitis and Crohn’s disease has also been described (97).

Inflammatory bowel disease is characterised by diarrhoea with blood and mucus, and may be accompanied by extraintestinal symptoms. Although aortitis has been reported in patients with Crohn’s disease (98-100), large-vessel involvement in ulcerative colitis is very uncommon (83, 101, 102).

Sjögren’s syndrome is a heterogeneous systemic autoimmune disease characterised by keratoconjunctivitis sicca, xerostomy and a wide spectrum of systemic symptoms and signs. Although vascular involvement is very uncommon, in the present study we described two cases associated with aortitis. We also reported a patient with aortitis and psoriatic arthritis. This association has previously been reported as single case report (103-106).

In our series, we described two patients with large-vessel vasculitis that also involved the aorta. The main features were unexplained fever along with high ESR in one patient and low back pain in the other.

Idiopathic aortitis is an uncommon disorder characterised by giant cells or lymphoplasmacytic inflammation of the aorta (107). Female gender, smoking, and older age are risk factors for the disease (108-111). Two related entities have been proposed: isolated idiopathic thoracic aortitis, and chronic periaortitis, that encompass idiopathic retroperitoneal fibrosis, inflammatory abdominal aortic aneurysm, periaortitis aortitis and idiopathic isolated abdominal periaortitis (1). Isolated aortitis usually manifests as an aneurysm of the ascending aorta, and is often an incidental finding during the histopathological study of the aortic wall after thoracic surgery (107). Nevertheless, sometimes it may present with symptoms related to aortic inflammation. Liang et al. published 64 patients who underwent an aortic aneurysm dissection at Mayo Clinic. Most of them (81%) were classified as isolated aortitis. Forty-five percent of the patients had aneurysm-related symptoms, 33% were asymptomatic, 12.5% had constitutional symptoms, 9.4% polymyalgia and 4.7% suffered symptoms related to the involvement of cranial arteries. Seventy-two per cent of patients had additional vascular imaging abnormalities (109). Miller et al. found isolated aortitis in 47% of patients from a series of 45 patients who underwent an ascending aortic resection (112).

Patients with retroperitoneal fibrosis, inflammatory abdominal aortic aneurysms or periaortitis are aortitis may develop constitutional symptoms, abdominal or back pain, fever, fatigue, night sweats and elevated inflammatory markers (1, 4, 5, 9, 113-117). In patients with retroperitoneal fibrosis, ureteral obstruction and renal failure have been reported (1, 5); and sometimes blood and lymphatic vessels may be also involved (116, 118).

There are other diseases that may be also included under the term “idiopathic aortitis”. This is the case of Erdheim-Chester disease and immunoglobulin G4-related disease. Erdheim-Chester disease is a rare systemic disease characterised histopathologically by histiocytosis of non-Langerhans cells. It may present with retroperitoneal infiltration and cardiovascular involvement. When the aorta is affected, the periarticular tissue is infiltrated circumferentially from ascending aorta to the iliac bifurcation (7, 42, 119, 120). Immunoglobulin G4-related disease encompasses a broad spectrum of disorders presenting with fibrosis of several organs, increase of serum IgG and IgG4 and autoantibodies. The aorta and its major branches show a lymphoplasmyctatic infiltration and irregular fibrosis in the adventitial layer (42, 121). In this entity, chronic periaortitis is more frequent than aortitis. Differences between isolated aortitis and GCA have recently been described by Talarico et al. (122).

In conclusion, aortitis is not an uncommon entity. However, the search for aortitis should only be undertaken in patients presenting with typical PMR under special circumstances. It may be the case in the evaluation of corticosteroid-resistant PMR patients (123).
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Concurrence of
Idiopathic retroperitoneal
fibrosis


