

Horton's aortitis

Vanessa Marchal* and Muriel Sprynger

CHU de Liège, Sart Tilman B35, Liège 4000, Belgium

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Giant cell arteritis is the most common systemic vasculitis in people over the age of 50 years. Ischaemic manifestations are well known. 'Occult' manifestations as aortic aneurysmal disease need consideration. The incidence of aortic aneurysm and/or dissection is about 18.5 per 1000 person-years at risk (18.9 in Lugo(4) and 18.7 in Olmsted County(3)). Predictive factors are hypertension, polymyalgia rheumatica, coronaropathy, and hyperlipaemia. Another factor is the apparition of an aortic regurgitation murmur as in this case. So, these patients should be monitored by echocardiography.

Introduction

Giant cell arteritis (GCA) is a systemic vasculitis that affects large- and medium-sized arteries in patients of more than 50 years.^{1,2} Annual prevalence is 15–33 cases per 100.000 person aged 50 years and older. The aorta and any of its branches can be affected. The aortic wall is primarily concerned. In case of aneurysm, the dilatation of the sinuses of Valsalva can cause valvular separation and thus induce regurgitation.

Giant cell arteritis' aortitis is sometimes neglected and can lead to acute aortic dissection and/or aortic aneurysm in about 18% of the cases.^{3,4}

We report the case of a thoracic aneurysm that was incidentally discovered after 13 years of corticotherapy for GCA while investigating an aortic regurgitation murmur.

Case report

In December 2005, a 75 year old women took cardiologic advice for shortness of breath. Her medical history included chronic obstructive pulmonary disease, giant cell arteritis proved by a temporal artery biopsy, polymyalgia rheumatica (PMR) (diagnosed in 1992), recent hypertension (diagnosed in 2006), and microscopic hematury. Cardiovascular risk factors are hypertension and smoking (20 cigarettes per day). Her daily treatment consisted of Tibolone, N-acetylcystine, and methylprednisolone 2 mg every 2 days. Physical examination revealed a normal blood pressure (140/70 mm Hg), a regular heart rate (65 per min) and a mild parasternal diastolic murmur (grade 2/6). Some rhonchi were heard with disseminated wheezing. The abdominal examination was normal. The patient's weight was 55 kg and her height 150 cm. She had a good physical

condition and no signs suggesting relapsing of GCA. The electrocardiogram was normal. Biology showed a C-reactive protein value of 2.3 mg/L, Hb of 12.7 g/dL, leucocytes 6480 per mm³, erythrocytes sedimentation rate (ESR) 9 mm/1 h, fibrin 4 g/L, and no antineutrophils antibodies (ANCA). The transthoracic echocardiography showed a normal left ventricle systolic function, mild aortic regurgitation with pressure half-time of 400 ms, a moderate mitral regurgitation, and a tricuspid regurgitation (grade 2–3/4), with a mild pulmonary hypertension (systolic pulmonary pressure: 34 mm Hg). Abdominal aorta and renal arteries were normal. Ascending aorta was dilated just above the Valsalva's sine (maximal diameter: 55.5 mm). The pericard was virtual (*Figure 1*).

The thoracic aorta diameter was confirmed by CT-Scan (*Figure 2*). The preoperative assessment was completed by a coronarography which revealed a monotoncular atherosclerotic coronary disease and confirmed the aortic dilatation. The ascending aorta was preventively replaced by a 28 mm 'Vascutex' conduit, completed by a venous bypass on the posterior interventricular artery. The aortic valve was preserved after plicature of the sine of Valsalva and suspension of the corners. The corticotherapy was continued. The shortness of breath regressed with the optimization of chronic obstructive pulmonary disease treatment. The patient's evolution was uneventful.

Histological examination of the aortic wall samples (6.5 × 5 × 0.4 cm) assessed the diagnosis of GCA. The aortic internal wall was macroscopically congestive with mild lipid depositions and the external wall was irregular. Histological examination showed fibrosis and thickening of the intima and the media. The media contained an inflammatory infiltrate with lymphocytes, plasma cells, and multinucleated giant cells. Within the fibrosed adventitia, these inflammatory cells formed regular lymphoid nodes (*Figure 3*).

* Corresponding author. Tel: +32 498753676; fax: +32 43714416.
E-mail address: vanessa@produweb.be

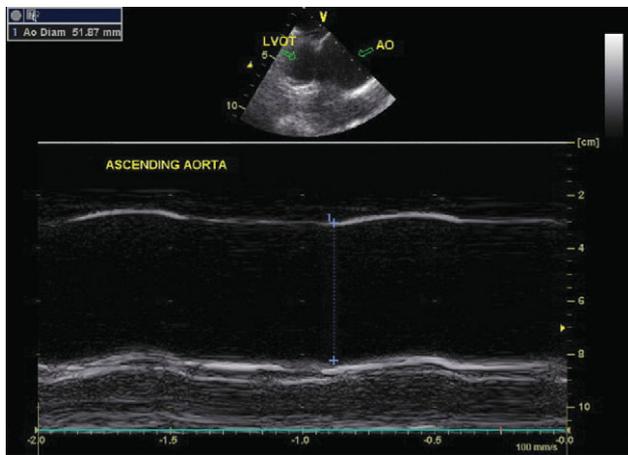


Figure 1 Echocardiography: TM mode: ascending aorta aneurysm.

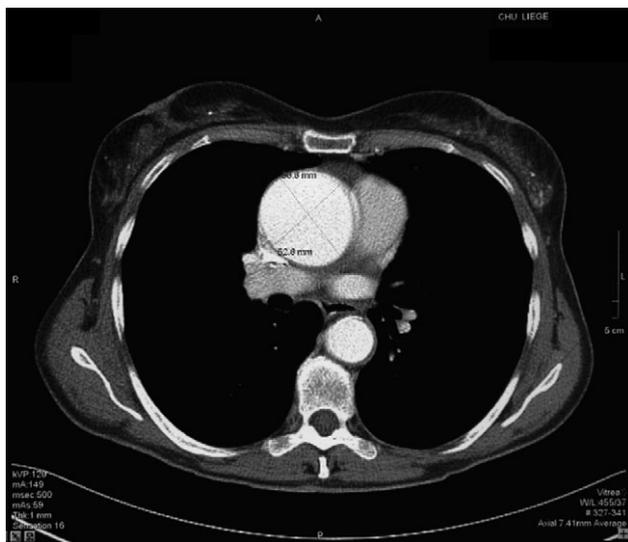


Figure 2 CT scan: ascending aorta aneurysm.

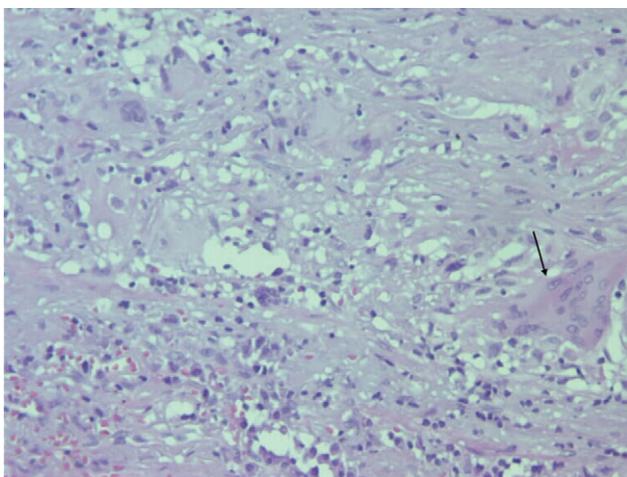


Figure 3 Multinucleated giant cells and fibrosed media.

Discussion and conclusion

This clinical case enhances the need of looking for aortic aneurysm in the follow-up of GCA. This adverse evolution

is not as rare as one could think. It was confirmed by two studies carried out by Nuenninghoff *et al.* and Gonzales-Gay *et al.* Annual prevalence is 15–33 cases per 100.000 person aged 50 years and older. The histological examination showed fibrosis and thickening of the intima, the media,⁵ and the adventitia⁶ with loss of normal elastic fibres and a chronic inflammatory cell infiltrate involving the full thickness of the wall⁵ with many multinucleated giant cells, lymphocytic, and plasma cellular infiltrates. In our patient, giant cell aortitis was histologically confirmed. There are many clinical manifestations of giant cell's aortitis: aortic aneurysm, aortic dissection, aortic rupture, aortic arch syndrome, aortic wall haematoma, aortic regurgitation, aortopulmonary, or aortodigestive fistula.⁷ However, some clinical signs are well correlated with the higher incidence of aneurysms^{3,4}: systemic hypertension, PMR, coronaropathy, and hyperlipaemia. The most predictive association is, nevertheless, essential hypertension and PMR with an acute severe inflammatory response at the time of diagnosis. Another predictive factor is the apparition of an aortic regurgitation murmur as in this case. The aortic regurgitation is due to the dilatation of the sinuses of Valsalva which causes the aortic valve commissure separation.^{7,8} Aortitis is often diagnosed in patients with well-known GCA, but most of the time when the corticoids are progressively lowered or even when they are stopped.^{7–13} Therefore, the thoracic aorta aneurysm is often lately diagnosed. The mean delay between GCA's diagnosis and discovery of the thoracic aneurysm varies from 5 to 7 years.^{1,7} The normalization of the sedimentation rate does not exclude the evaluative capacities of the aortitis.^{8–10,13,14} The ongoing inflammatory process could explain the late expression of the disease.⁷

The originality of this case is the extremely late expression of the aortic disease (diagnosis of aortitis 13 years after the diagnosis of GCA, despite of a non-regressive corticoid treatment). Many aetiologies could be evoked: infection aneurysm (syphilis, salmonella, etc.), inflammatory non-septic aneurysm, Marfan's syndrome, Takayasu's disease, Cogan's syndrome, Behcet's syndrome, and spondylarthropathies.¹⁵ In our case, the confirmation was histological. Giant cell aortitis can lead to aneurysmal evolution, to dissection, aortic rupture, and sudden death. Those issues are due to elastophagia and elastolysis of the arterial wall,^{16,17} acute aortitis of the whole wall, and vasa vasorum arteritis. Patients with giant cell arteritis require a long-term follow-up because of the risk of aortic dissection and sudden death. Lots of authors recommend a yearly lateral chest radiography.^{8,10,14,18–20,22}

However, the use of transthoracic echocardiography (eventually completed by CT-scan, MRI, and PET scan) is much effective. Potential advantages are no irradiation, good sensitivity and specificity, low cost, and easy availability.^{1,15,21} Particular attention must be paid to patients with arterial high blood pressure, hyperlipaemia, and/or new aortic regurgitation murmurs. Prospective imaging studies are needed¹ in order to elaborate the guidelines of the follow-up of GCA.

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