Giant cell aortitis in treatment-naïve human immunodeficiency virus and hepatitis C coinfection

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ABSTRACT

Giant cell arteritis is a common form of vasculitis, although involvement of the aorta is unusual. There is no established association between giant cell aortitis and human immunodeficiency virus (HIV) and hepatitis C virus (HCV) coinfection. We present the case of a 46-year-old female coinfecting with HIV and HCV who had never received antiretroviral therapy and developed symptoms of deteriorating shortness of breath on exertion. Investigations demonstrated aortic root and ascending aorta dilatation, along with severe aortic valve regurgitation, for which the patient underwent valve-sparing aortic root replacement (a David procedure). Histopathology confirmed giant cell aortitis.

KEYWORDS

Aortitis – HIV infection – HCV infection – Aortic aneurysm – David procedure

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Giant cell arteritis (GCA) is the leading cause of inflammatory vasculitis affecting medium and large sized vessels, and presents predominantly in women over 50 years of age. GCA typically involves the branches of external carotid artery, while the aorta and its branches are uncommon sites.¹ Human immunodeficiency virus (HIV) and hepatitis C (HCV) infection are recognised as causes of systemic vasculitis.

We present the case of an HIV and HCV co-infected, treatment-naïve individual who developed an ascending aortic aneurysm, with histological findings revealing giant cell aortitis.

Case presentation

A 46-year-old female presented with a 2-month history of fatigue, pleuritic chest pain and shortness of breath. Symptoms included memory impairment and poor coordination and vision. She had been diagnosed with HIV infection 22 years previously, but had persistently refused antiretroviral treatment (ART). Her CD4 count was 268 cells/mm³ and her viral load was 815,142 copies/ml.

In healthy adults, CD4 counts range from 500–1,200 cells/mm³. A CD4 count below 200 cells/mm³ is a qualification for diagnosis of stage 3 infection (acquired immune deficiency syndrome) and it is recommended that ART is started urgently. The viral load provides information on the patients' health status and is declared 'undetectable' if it is below 40 copies/ml.

The patient was also infected with Hepatitis C-3a, at a viral load of 2.9 million copies/ml. An HCV viral load of more than 2 million copies/ml is considered high. Syphilis serology was negative.

Transthoracic echocardiography demonstrated a trileaflet aortic valve (AV) with malcoaptation of the leaflets and moderate-to-severe central aortic regurgitation (AR). The aortic root was 5.3cm at the level of the Sinuses of Valsava. The left ventricle (LV) was dilated. Computed tomography demonstrated dilatation of the ascending aorta up to 6cm in diameter. The aortic arch and descending aorta were not aneurysmatic.

Brain magnetic resonance imaging (MRI) showed left frontal and cerebellar lesions and the patient's serum was positive for toxoplasma antibody. She was therefore started on sulphadiazine and pyrimethamine for cerebral toxoplasmosis, and ART with elvitegravir/cobicistat/emtricitabine/tenofovir. Cardiac MRI confirmed moderate-to-severe AR (at a regurgitant fraction of 35%) and a severely dilated LV (left ventricular end-diastolic volume 282mL, left ventricular end-systolic volume 102mL) with preserved systolic function, at an ejection fraction of 64% (Fig 1).

The patient underwent valve-sparing aortic root replacement involving re-implantation of the aortic valve (a David procedure). A 30-mm straight Hemashield graft (Maquet, Rastatt, Germany) was used. The procedure and postoperative course were uncomplicated. Echocardiography before discharge and at 2-month follow up showed good AV leaflet coaptation and a mild central regurgitant jet.

Figure 1 Cardiac magnetic resonance imaging showing moderate-to-severe aortic regurgitation, a severely dilated left ventricle and aortic root dilatation. 94×134mm (96×96 DPI)

The histopathologic findings of the aortic specimen revealed fulminant giant cell aortitis and intimitis, with extensive media necrosis. The aortic wall showed precocious atheroma with foamy macrophages, lymphocytic inflammation and giant cells. There was fulminant media necrosis bordered by lymphocytes, plasma cells and giant cells. Elastic-Van-Gieson stain showed abundant destruction of elastic fibers. The adventitia showed fibrosis and inflammation (Fig 2).

Discussion

An aneurysm of the ascending aorta and AR caused by GCA was first described in 1965, but there are few further reports in the available literature. GCA can affect the ascending aorta from the root to the arch without the involvement of the AV and is rarely related to HIV infection.

A recent UK study demonstrated a two-fold increase in the relative risk of developing ascending aortic aneurysm in patients with GCA versus matched controls from the general population. Smoking and male gender are independent predictors for aortic aneurysm in both GCA and non-GCA cohorts.²

A previous case report from our institution presented a young woman, under treatment for HIV, who was found to have symptomatic dilatation of aortic root and severe AR. She underwent aortic valve and root replacement, and the histopathology examination showed GCA.³ Patients with GCA have an increased risk of developing aortic aneurysms,

Figure 2 Section of the aorta with destruction of media. Hematoxylin and eosin stain. 186×162mm (96×96 DPI)

and patients with HIV and/or HCV coinfection are more inclined to develop GCA.

HIV-infected individuals are prone to cardiovascular dysfunction and large artery vasculitis. Moreover, patients on protease inhibitors used as part of a highly active ART regimen have accelerated atherosclerosis of their coronary arteries.⁴

HCV infection may be an independent factor for the development of cardiovascular disease and is also associated with vasculitic changes. In individuals co-infected with HCV and HIV, types II and III cryoglobulinemias occur with higher prevalence than in mono-infected HIV patients. Polyarthritides Nodosa is also associated with HCV infection, usually presenting after prolonged HCV infection, predominantly in older females.⁵

We present an uncommon case of an aortic root aneurysm caused by GCA and associated with treatment-naïve HIV infection and positive hepatitis C serology. The patient underwent valve-sparing aortic root replacement (a David procedure), which is considered the best option for a young patient in terms of prognosis and life expectancy. If the operation is performed by experienced surgeons, a valve-sparing approach could be a life-long operation, provided the patient is under well-controlled ART so that further degeneration of the tissues is not expedited. Regular clinical and imaging follow-up is necessary to monitor inflammatory changes and further dilatation affecting the rest of the aorta and other medium- and large-size vessels.

Conclusions

It is notable that patients with HIV and HCV coinfection who are immune compromised could be susceptible to GCA. When an aortic root and ascending aorta aneurysm is

diagnosed, an operation should be considered promptly in an experienced centre. The follow-up after surgery is of paramount importance, as patients with GCA could present with inflammatory vasculitis and aneurysms of other vessels.

Conflicts of interest

None declared

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