Aortic root dilation secondary to giant cell aortitis in a human immunodeficiency virus-positive patient

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Abstract

HIV-associated vasculitis rarely involves the aorta. There is no well-established association of HIV and giant cell arteritis. We present the case of a 31-year-old HIV positive Indian woman who was referred to us with complaints of dyspnea and chest pain. Physical examination revealed a diastolic murmur in the aortic area and echocardiography showed a dilated aortic root causing severe aortic regurgitation. She was being adequately treated with anti-HIV therapy. She underwent aortic valve and root replacement and the histopathological findings of the aortic specimen showed giant cell arteritis.

Keywords: Aortic root dilation; HIV; Giant cell aortitis

1. Introduction

Vasculitis is an important, though less common, manifestation of HIV infection. Many different types of vasculitis have been associated with HIV, with predominantly medium to small visceral vessel involvement [1]. Involvement of skin, brain and neuromuscular tissue has been reported most frequently, while large vessel involvement is extremely rare [2]. Giant cell arteritis preferentially affects small and medium sized extracranial arteries but in approximately 10% of cases large arteries are affected. It can affect the ascending aorta from the root to the arch, but usually spares the aortic valve [3]. Giant cell arteritis is not typically seen with HIV. We present the case of a young Indian woman who was HIV positive with an ascending aortic aneurysm, which on histological examination revealed giant cell arteritis.

2. Case description

We report the case of a 31-year-old Indian woman, who presented to a district general hospital with acute onset of dyspnea and chest pain for 2 h. She was 4 months postpartum and had been found to be HIV positive during the antenatal period of that pregnancy. The patient had been having progressive shortness of breath for the last 2 years. On examination she had an early diastolic murmur in the aortic area. An urgent echocardiogram (transthoracic and transesophageal) showed a dilated aortic root with severe aortic regurgitation. The diameters were as follows: aortic annulus 31 mm, aortic root 50 mm, and mid-ascending aorta 55 mm. The left ventricle was globally impaired with an end-diastolic diameter of 70 mm. A CT scan of the thorax revealed no evidence of dissection. During the fifth month of her recent pregnancy she had been commenced on highly active anti-retroviral treatment (HAART), which consisted of Nevirapine 200 mg BD, Combowir BD, and Cotrimoxazole 960 mg thrice a week for prophylaxis. Her previous records had revealed a negative VDRL and TPPA screen, and CD4 count and viral load were 199 (20.4%) and <50, respectively. Although she was CMV positive, hepatitis B and C were negative and the HIV team at the referring hospital was satisfied that her HIV was controlled on therapy. The patient was a nonsmoker without any past medical history of cardiorespiratory disease.

The patient was then transferred to a cardiothoracic unit where she underwent an aortic valve and root replacement with a 25 mm Medtronic Freestyle stentless porcine bioprosthesis. A tissue valve was chosen by the patient who was unwilling to take warfarin. The intraoperative findings were a 55 mm diameter ascending aorta and a dilated left ventricle consistent with chronic aortic regurgitation. The aortic root and annulus were dilated exactly as predicted by the preoperative echocardiogram. The aortic valve leaflets were stretched and extremely thin and it was reasoned that a...
valve sparing procedure would not result in a satisfactory long-term result. The operation was straightforward and the patient was discharged from the hospital on sixth postoperative day.

The histopathological examination (Fig. 1) of the aortic wall revealed fibrosis and thickening of the intima and media with loss of normal elastic fibers. There was a severe chronic inflammatory cell infiltrate involving the full thickness of the wall with scattered histiocytic giant cells and occasional areas of medial collagen necrosis. These latter areas were associated with a neutrophilic infiltrate. Special stains for acid fast bacilli (ZN) and fungi (Grocott) were negative. The histological diagnosis was giant cell arteritis.

3. Discussion

Schwartz et al. [4] reported the first case of an HIV positive patient with AIDS who presented with eosinophilic vasculitis manifesting as Amaurosis Fugax in 1986. The exact etiology of HIV-related large vessel vasculitis is not known. Possible explanations include; immune complex mediated vasculitis secondary to dysregulation of the immune response, infections due to immunosuppression or direct HIV infection of the vessel wall. Chetty et al. [5] studied 16 HIV positive patients with large vessel disease consisting of aneurysms (often multiple) or occlusive disease and consistently found leukoclastic vasculitis of vasa vasorum and periadventitial vessels in their series. Only one aneurysm involved the thoracic aorta. The rarity of involvement of the aorta in AIDS patients is exemplified by Marks and Kuskov’s [6] report of atypical aneurysms in 16 young adult patients (12 HIV infected), in which there were only two thoracic aortic aneurysms. They found a lymphoproliferative response in the vessel wall. There is only a single case report of an HIV-related true aneurysm [7] of ascending aorta, in which the histopathology revealed a granulomatous giant cell mesoarthritis. Protopapas and Pugsley [8] have reported another case of HIV-related aneurysm of the aortic root in which the history and intraoperative findings were suggestive of syphilis as the cause and no definitive histopathological diagnosis was established.

The anti-protease part of treatment for HIV is known to increase arterial atherosclerosis. Mirza et al. [9] report a case of HIV associated ascending aortic aneurism in which histopathology showed evidence of accelerated atherosclerosis rather than vasculitis, the former being attributed to the lipodystrophy syndrome associated with protease inhibitors, used as part of HAART regimen. Our patient was on anti-protease treatment but the histology was not consistent with atherosclerosis. Interestingly, HIV has been associated with aortic root dilation in children. Lai et al. [10] showed that in children between 2 and 9 years of age, the mean aortic root measurements were significantly increased in HIV-infected children compared to HIV un-infected children.

In conclusion, the case that we have described is unique due to the uncommon findings of giant cell arteritis associated with HIV, causing aortic root dilatation with valve regurgitation. Although aneurysms are known to occur in HIV patients, they are usually due to etiologies other than giant cell arteritis and do not usually involve the ascending aorta.

References