Unusual Presentation of HIV Vasculopathy in a Child

Deenadayalan Munirathnam · Revathi Raj

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Abstract The manifestations of human immunodeficiency virus (HIV) infection are protean and vasculopathy is an uncommon but important complication. A large-vessel (aorta and femoral and carotid arteries) vasculopathy is rarely described in children. We describe a HIV positive thalassaemic child who developed multiple aneurysms in abdominal aorta. HIV vasculopathy resulted in severe aortoarteritis and was fatal in this child.

Case Report

A 10 year old female child, a known case of thalassaemia major since the age of 6 months, had been on regular blood transfusions since then. She underwent splenectomy at the age of four. She presented to us with history of abdominal pain and pain in both lower extremities which was progressively increasing in nature. On examination, she had pallor and hepatomegaly of 3 cm below the costal margin with no other specific signs. Complete haemogram showed haemoglobin 11.1 g/dl, total leukocyte count 23780 cells/cum, platelet count $7.41 \times 10^9/\mu\text{l}$ with ESR of 32 mm at the end of 1 h. Mantoux was negative. Work up for tuberculosis, cytomegalovirus syphilis, hepatitis B and C was negative. HIV I/HIV II by Elisa were positive. Absolute CD 4 count was 2552 cells/ μl . CT abdomen was done in view of persistent abdominal pain and showed an aneurysm with mural thrombosis of infra renal segment of abdominal aorta

and bilateral common iliac vessels. Abdominal angiogram revealed irregular circumferential wall thickening with formation of multiple aneurysms in infra renal aorta, bilateral common iliac and left internal iliac arteries and complete occlusion of right external and internal iliac, right common femoral and left external iliac arteries (Pictures 1 and 2). Screening of thoracic aorta shows no significant abnormality.

Antiretroviral therapy was not started in view of patient presenting with HIV CDC stage A1. She was started on Inj.Enoxaparin 20 mg subcutaneously and Aspirin 75 mg once daily orally. Vascular and cardiothoracic consults were obtained. The aneurysm was deemed inoperable as it was extensive and medical management was continued. Abdominal pain continued to increase despite medical management and the child succumbed within 7 weeks of the onset of illness due to rupture of the aneurysm.

Discussion

Vasculopathy is an uncommon feature of HIV disease but has significant morbidity and mortality. HIV vasculopathy commonly affects small and medium sized vessels but large-vessels involvement can rarely occur in children. The prevalence of cerebrovascular disease has been reported as 2.6% in children with HIV [1]. Medium- and large-vessel involvement can result in either multiple aneurysm formation or occlusive disease, as seen in our patient. Unusual sites such as the descending aorta, subclavian, internal carotid arteries, and renal arteries can be affected.

A wide range of vasculitides has been described ranging from vasculitis resulting from specific infective agents to a non-specific vasculitis. Vasculopathic process in

D. Munirathnam · R. Raj (✉)
Department of Paediatric Haematology, Apollo Speciality
Hospital, 320, Padma Complex, Anna Salai, Chennai 600035,
India
e-mail: revaraj@yahoo.com

Picture 1 Abdominal aortic aneurysm

Picture 2 CT Angiogram pictures of the aortic aneurysm

HIV can be classified as infective, necrotizing, systemic hypersensitive, angiocentric, immunoproliferative, primary angitis of the central nervous system, large-vessel

vasculopathy, and miscellaneous [2]. Cytomegalovirus, herpes zoster, toxoplasmosis, pneumocystis, salmonella, and *Mycobacterium tuberculosis* have been associated with vasculitis in patients with HIV [3]. Infectious agents can cause vasculitis of arteries and veins of all sizes and in all organs. Aneurysm formation or occlusive disease of large elastic arteries is mostly of infective aetiology [4, 5]. However, large-vessel involvement has been noted in the absence of overt infection [6]. Affected patients tend to be young and present with multiple aneurysms or occlusions of the carotid, femoral, and popliteal arteries. Damage to large-vessel wall culminating either in aneurysm formation or occlusive disease was most probably due to leucocytoclastic vasculitis of the vasa vasora and periadventitial vessels. The exact aetiopathogenesis by which the vasculitic processes occur in HIV is still not clear and is most probably multifactorial. An immune complex-mediated mechanism or a direct infection of vascular or perivascular tissue has been postulated as the cause of HIV vasculopathy [7]. Direct infection of aortic fibroblasts by HIV virus is more likely to be a contributing factor than the process of molecular mimicry [8]. In HIV there is an oligoclonal expansion of T cells especially CD8 positive cells resulting in increased secretion of vascular endothelial cells growth factor A (VEGF-A) that may induce vascular leakage and stimulate proliferation of vascular endothelial cells [9]. Aneurysmal dilatation of ascending aorta due to pseudo-xanthoma elasticum like syndrome in a patient with beta thalassemia intermedia has been reported [10]. Spontaneous regression of an abdominal aortic aneurysm in an immunocompromised patient has been documented [11].

Severe immunosuppression may accentuate the vasculitis in HIV patients [12]. Immune-reconstitution inflammatory syndrome (IRIS) could be implicated in the pathogenesis of their vascular complications [13]. Definitive diagnosis can be achieved by ultrasonography, Doppler ultrasound, or computed tomography [14]. The optimal management of these patients has not yet been well established. A low CD4 is associated with poor surgical outcome, but preoperative HAART has improved surgical outcomes. Medical management including HAART has been used in children with good results [15, 16]. Further studies are required to determine if early treatment with HAART will prevent the occurrence of HIV vasculopathy.

To conclude, HIV vasculopathy involving large vessels may occur due to part of a leucocytoclastic vasculitis of the vasa vasora or periadventitial vessels. An infective aetiology should always be sought so that appropriate treatment can be instituted. The precise mechanisms by which vasculopathy occurs and whether early treatment with HAART will prevent the occurrence of vasculopathy need further study.

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