**Case report**

**Occlusive vasculopathy in human immunodeficiency virus (HIV)-associated vasculitis: unusual clinical and imaging course**

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**ABSTRACT**

Human immunodeficiency virus (HIV)-associated vasculitis is a rare secondary systemic vasculitis involving small and medium arteries. We report a 42-year-old man with uncontrolled HIV infection presenting with long-lasting abdominal pain. An abdominal CT angiography revealed multiple microaneurysms and stenoses in intrarenal arteries, with involvement of mesenteric and hepatic arteries. HIV-associated vasculitis was diagnosed and glucocorticoids and raltegravir-based antiretroviral therapy were administered with good initial clinical and virological response. Several episodes of acute intestinal ischaemia were later developed requiring bowel resections of which histological examination showed vascular occlusive fibrotic changes without active vasculitic lesions. Vasculitis persisted in remission and intrarenal microaneurysms disappeared.

**Introduction**

Polyarteritis nodosa (PAN) is a rare primary systemic vasculitis involving small and medium arteries (1, 2). Hepatitis B virus (HBV) and human immunodeficiency virus-1 (HIV-1) may cause necrotising vasculitis almost identical to PAN. Such secondary vasculitides have been recently classified and renamed as HBV- and HIV-associated vasculitis, respectively (1). Clinical manifestations of PAN and HBV- and HIV-associated vasculitis range from non-specific symptoms to severe organ damage due to tissue ischaemia and/or haemorrhage. The main abdominal visceral arteries (celiac trunk, superior and inferior mesenteric and renal arteries) and their branches are typically affected (2). Gastrointestinal involvement confers a poor prognosis to all necrotising vasculitides (2, 3). When conventional biopsies (muscle, peripheral nerve, skin) are not performed or result negative or inconclusive to confirm diagnosis, angiographic changes showing microaneurysms, dilatations, stenoses and/or occlusions of the visceral arteries may also support PAN diagnosis (2, 4).

We report the unusual clinical and imaging course of an HIV-infected man with severe immunosuppression, who was diagnosed with HIV-associated vasculitis by an abdominal CT angiography after presenting with long-lasting abdominal pain.

**Case report**

A 42-year-old man, heavy smoker, with a past medical history of HIV infection diagnosed in 2000, Pneumocystis jirovecii pneumonia in 2009, and poor adherence to antiretroviral therapy, presented with recurrent episodes of diffuse abdominal pain associated to weakness and mild diffuse myalgia during the previous year. He did not present with cutaneous, joint, ocular or neuropathic symptoms, or fever. Fibrogastroscopy, endoscopic capsule, colonoscopy (plus intestinal biopsies), MR enterography and several contrast-enhanced abdominal CT had revealed multiple ulcers and inflammatory changes in jejunum (by imaging and biopsies, respectively). All serology tests and intestinal biopsies resulted negative for bacterial, mycobacterial, protozal and viral infections. Empirical treatment for a possible cytomegalovirus infection was given with no improvement. Intestinal ischaemia was finally suspected. An abdominal CT angiography showed multiple microaneurysms, stenoses and occlusions of intrarenal arteries, irregular lesions of...
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mesenteric arteries, fusiform dilatations of hepatic branches, and splenic and kidney infarctions (Fig. 1). Abnormal laboratory results revealed increased acute phase reactants, normocytic anaemia, low lymphocyte (800/mm³) and CD4 (29 cells/mm³) counts and viral load of 274,000 copies/mL. Hepatic profile, renal function, urinalysis, hepatitis B and C virus serology, antineutrophil cytoplasmic antibodies, antiphospholipid antibodies, cryoglobulins, rheumatoid factor and complement were normal or negative. An electromyogram did not show myopathic or neuopathic abnormalities. A blinded (deltoid) muscle biopsy revealed normal vessels with tissue microinfarcts, suggesting ischaemia due to occlusion of proximal small arteries beyond the examined sample.

The patient was finally diagnosed with HIV-associated vasculitis (November 2014) and antiretroviral therapy (raltegravir, emtricitabine and tenofovir) was started, with intravenous high-doses methylprednisolone (1gr/day per 3 days), followed by prednisone (1mg/kg/day). Three months later, while vasculitis was considered to be in remission, the patient presented with sudden abdominal pain caused by intestinal ischaemia, and required surgical resection of the affected intestine. At month 11 of starting treatment, intestinal abdominal pain occurred, requiring new intestinal resection. At month 11 of starting treatment, intense abdominal pain occurred and an abdominal CT angiography identified a progressive stenotic lesion with distal occlusion of a superior mesenteric artery branch and marked artery wall thickening (Fig. 3a-b). Since a distal thrombosis could not be excluded, oral anticoagulation therapy was started and no surgery was needed. After 16 months, a new ileal perforation due to segmental ischaemia required a new resection of the affected ileum. Histopathological examination of the three resected samples of small intestine revealed focal transmural necrosis with small and medium arteries showing preocclusive and occlusive lesions with remarkable intimal hyperplasia, neovascularisation and thrombotic changes, without signs of active vasculitis (Fig. 2a-e). A proximal medium-sized artery from the initially resected ileum disclosed a notable intimal hyperplasia degree (Fig. 3c), similar to that observed in the superior mesenteric artery (Fig. 3a-b).

Because all episodes of intestinal ischaemia were attributed to the progression of the vascular repair with fibrosis and occlusion of the previously inflamed vessels, no increase in prednisone doses taken by the patient was indicated in any of them. After the last small bowel resection, the patient has quitted smoking and has remained stable for the past six months on the same antiretroviral therapy and prednisone 2.5 mg/day. Viral load always remained undetectable and CD4 counts ranged between 30–90 cells/mm³. However, he suffered from mild partial bowel obstruction symptoms, which were alleviated with low-residue diet. A control abdominal CT angiography showed good patency of the main visceral arteries, persistence of fusiform dilatations of intrahepatic artery branches and resolution of renal microaneurysms (Fig. 1F).

Discussion

Initial descriptions of necrotising vasculitis in HIV-1 patients were reported in the late 1990s (5). In HIV-associated vasculitis, as it occurs with HBV-associated vasculitis (2, 6-8) and, more recently with HCV-associated cryoglobulinaemic vasculitis (9), disease activity is directly related to viral load and treatment should be focused on targeting the potential causal agent. Therefore, antiviral agents in combination with glucocorticoids are the mainstay of therapy for these secondary vasculitides (2, 6-9).

Although patients with HIV-associated vasculitis have been reported to have milder disease and better long-term prognosis than (idiopathic) PAN (7, 10), HBV-related vasculitis patients present more often with abdominal involvement and more severe outcome than PAN patients (11). Nevertheless, abdominal involvement ensues “per se” a poor prognostic factor, with higher morbidity and mortality, in patients with all vasculitis types (6, 11), even in those patients with single-organ vasculitis localised in abdominal structures (12).

In PAN patients multiple dilatations or aneurysms of the abdominal vasculature are the most commonly reported arterial lesions, occurring in up to 60% of patients (4, 13). Patients with abdominal angiographic abnormalities commonly have involvement of other vascular beds when extended studies

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**Fig. 1.** CT and CT angiography of the kidneys. Arterial phase contrast-enhanced coronal CT (A and D), CT angiography using Maximum Intensity Projection (B and E) and three-dimensional CT angiography using volume rendering reconstructions (C and F). Images at the time of diagnosis show multiple artery microaneurysms in both kidneys (arrows) (A-C) and stenotic lesions (short arrows) and a proximal occlusion (asterisk) (B). Sixteen months after starting treatment, microaneurysms are not visualised (D-F).
are performed (4). The presence of microinfarcts in a muscle biopsy might also indicate multisystem involvement in our case.

Acute vascular lesions in necrotising vasculitis are characterised by segmental weakening, usually leading to microaneurysms formation or disruption of the artery wall, and/or luminal narrowing due to intimal hyperplasia (2). However, in advanced or healing lesions, vascular remodelling with fibrotic changes of the vessel wall and progression of myointimal proliferation with neovessel formation may lead to stenotic and occlusive lesions (2, 14, 15). The occurrence of thrombosis is frequently reported and may also contribute to the final occlusive event (2). In our patient, on one hand, chronic inflammatory state induced by HIV infection, cigarette smoking and also the proved effects of antiretroviral agents on the progression of carotid artery intima-media thickness (16) may have influenced in the development of occlusive vasculopathy present in histological and imaging studies. On the other hand, the prolonged low-grade grumbling disease experienced prior the treatment was started seems to have favoured the development of chronic vascular remodelling and fibrotic damage leading to artery occlusions and intestinal ischaemia as a delayed complication of vasculitis. Of note, and because we realise this is an exceptional situation, in similar cases, if biopsy or surgical resection of the ischaemic territory cannot be obtained to assess the final aetiology, active disease should be still considered to play a role and dose increase of immunosuppressive agents has to be warranted.

HIV-associated vasculitis has been previously reported to occur as long-lasting abdominal pain in a HIV patient with mesenteric microaneurysms (17). However, structural regression of microaneurysms has been reported only in patients with PAN and HBV-related vasculitis, a fact that indicates good response to therapy (18). To the best of our knowledge, this is the first case of HIV-associated vasculitis, in whom regression of renal microaneurysms was observed after appropriate therapy was begun.

In summary, HIV-associated vasculitis has to be considered a potential diagnosis in HIV patients presenting with abdominal pain, mainly in those with poor HIV control. CT angiography of

![Fig. 2. Histology of the vessels in the resected small intestine. Samples from the first (A-C), second (D) and third (E) small intestinal resections. (A-C) A medium- (A) and small-sized arteries (B and C) in the ileal serosa show a remarkable intimal hyperplasia with severe stenosis and occlusion of the lumen due to a fibrotic and irregular tissue within the intima (A) and also destruction of all artery layers (B and C), with multiple neovessel formation (several of them marked with tiny arrows). D-E) Significant lumen reduction due to vessel wall thickening and intimal hyperplasia is present in small arteries in the intestinal serosa (arrows); some of them are occluded (arrowheads). Original magnification x10 (A, D and E) and x20 (B and C).](image)

![Fig. 3. Distal occlusion of a superior mesenteric artery branch. Contrast-enhanced (portal phase) coronal (A) and consecutive axial sections (B) of a branch of the superior mesenteric artery showing progressive and distal occlusion (tiny black arrows in B) with a non-enhancing segment of small bowel, suggesting ischaemia (white arrow in A). Proximal segment of the affected branch depicts a thickened artery wall (arrows in B). A medium-sized artery from the previous ileal resection also shows a wall thickening due to a marked intimal hyperplasia (C). Original magnification x10.](image)
the splanchnic circulation maybe useful for detecting vasculitic lesions. When abdominal involvement is detected, a vigorous treatment with glucocorticoids has to be promptly initiated, together with the reassurance of an effective antiretroviral therapy. A good response to treatment may be accompanied by the regression of renal microaneurysms.

References