Livedoid vasculopathy (LV) is a rare cutaneous chronic disease characterized by recurrent, painful ulcerations of the legs and feet, following focal infiltrated purpura. Widespread LV may also occur (1). LV is commonly associated with livedo reticularis (2). Healing is slow, and leaves atrophie blanche, an irreversible scar (3). LV is characterized histologically by fibrin occlusion of dermal vessels without vasculitis (4). Although the pathophysiology is not fully understood, LV is classified either as idiopathic, or as a secondary form associated with hypercoagulable and/or autoimmune disorders, which justify anticoagulants or intravenous immunoglobulins (5–7). We describe here 2 patients who developed LV associated with human immunodeficiency virus (HIV)-1 infection and severe nephropathy; an association that has not been reported previously.

CASE REPORTS

Case 1. A 56-year-old Caucasian man had a 15-year history of HIV-1 infection, complicated by membranoproliferative glomerulonephritis that led to chronic renal failure and haemodialysis for 2 years. He had lipodystrophy, metabolic syndrome and hypertriglyceridemia secondary to highly active antiretroviral therapy (HAART), associated with human immunodeficiency virus (HIV)-1 infection and severe nephropathy; an association that has not been reported previously.

Fig. 1. (a) Ulcerated lesions and atrophie blanche of the ankle in patient 1. (b) Perimalleolar purpuric papules associated with livedo reticularis and small superficial ulcers in patient 2.

DISCUSSION

These 2 original cases lead us to discuss the role of the association of HIV-1 and nephropathy in the development of LV, although, a mere coincidence
LV is a rare chronic dermatosis with an estimated prevalence of approximately 1:100,000 (8). This condition is frequently associated with comorbid conditions that lead to thrombosis of dermal vessels; for example, connective tissue diseases, malignancies, or venous insufficiency (4, 9). LV pathophysiology is linked to abnormalities in coagulation and/or fibrinolysis (4, 8). Associations of several factors are expected to lead to the disease, as in our 2 cases. Indeed, HIV has been shown to be an independent risk factor for thrombosis and hypercoagulable state: elevated markers of inflammation and coagulation are reported not only in patients who have AIDS, but also in HIV-infected patients on effective HAART (10, 11). In addition, HAART contributes to the development of a metabolic syndrome, characterized by lipodystrophy, dyslipidaemia and insulin resistance (12). This metabolic process contributes to the development of a proinflammatory state and subsequent chronic, subclinical vascular inflammation, which modulates and results in atherosclerotic developments (13). In addition, protease-inhibitors have been associated with higher PAI-1 levels in HIV-infected patients, which might result in a hypofibrinolytic state (14). Altogether, these elements argue for procoagulable and hypofibrinolytic states in HIV-1-infected patients; conditions that are found in LV. Moreover, these patients had nephropathy, which itself might induce coagulation abnormalities. Before transplantation, patients are frequently prone to acquire antiphospholipid antibodies and hyperhomocysteinaemia, particularly in haemodialysed patients, as observed in case 2. As the first patient had a renal transplantation before the development of LV and the second after the development of LV, renal transplantation appeared to not be directly involved in LV pathophysiology. However, renal transplantation has been associated with acquired hypercoagulable state and hypofibrinolytic state, with a multifactorial origin. Although the data are conflicting, some of these main factors are immunosuppressive drugs, especially corticosteroids and calcineurin inhibitors (15). Renal transplant recipients often develop hyperhomocysteinaemia, which contributes to an elevated risk of vascular thrombosis.

In conclusion, this report presents two patients with an original association of HIV-1 infection and LV, highlighting the possible increased risk of microcirculation thrombosis associated with HIV-1 and nephropathy. However, we shall not leave out the hypothesis of a coincidence between LV and HIV-1, more than a causal relationship.

REFERENCES


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