A Case of Widespread Livedoid Vasculopathy with Pain but no Systemic Symptoms

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Sir,
Livedoid vasculopathy (LV) is characterized by persistent livedo reticularis and recurrent painful ulcers, and usually involves the lower legs. LV is considered to be a non-inflammatory thrombotic disease due to the occlusion of dermal small vessels seen in patients with certain coagulation abnormalities (1). We report here a case of LV in a young woman who showed extended livedo reticularis without an abnormal autoimmune profile or coagulation parameters.

CASE REPORT
A 26-year-old woman was referred to our department in July 2005, due to painful ulcers on her lower legs. She had noted a reticular skin eruption on her legs for 2 years, which gradually extended over her upper extremities and trunk. She did not smoke. Physical examination revealed brownish to dark-reddish reticular erythema, so called livedo reticularis, over her lower extremities. Similar eruptions were also seen on her lumbar area and arms, including the dorsum of her hands. On the distal part of her lower legs, multiple dark purplish erythema and purpura were present, accompanied by well-demarcated ulcers with a haemorrhagic crust (Fig. 1). She complained of hyperaesthesia on the dorsal side of the foot.

A biopsy specimen taken from the purpura lesion of the ankle showed perivascular lymphocytic infiltrate in the dermis and a fibrinous occlusion of dermal blood vessels without leukocytoclasis (Fig. 2). The result of direct immunofluorescence was negative. No abnormal laboratory tests, including complete blood cell count and basic blood chemistry, were noted. Although slightly elevated erythrocyte sedimentation rate (22 mm/h; normal value 3~15) and C-reactive protein (1.2 mg/dl, normal value < 0.1) were detected, other results including cryoglobulin, rheumatoid factor, antinuclear antibodies, anti-double-strand DNA, anti-SSA/Ro antibodies, and anti-neutrophil cytoplasmic antibodies were negative. Anticardiolipin β2-GPI antibodies, lupus anticoagulant, prothrombin time, activated partial thrombin time, and protein C were also negative or within normal limits. A neurological examination revealed no remarkable abnormalities. The results of a magnetic resonance imaging of the brain were normal.

Based on the clinical and histopathological findings, a diagnosis of LV was established. Administration of intravenous alprostadil, oral aspirin and ticlopidine hydrochloride rapidly led to improvement of the purpura and the ulcers on her legs, but the livedo reticularis remained. The hyperaesthesia and the pain also resolved soon after the start of treatment. Since she became pregnant 5 months after the initiation of treatment, the medications were discontinued. There has been no recurrence during a 2-year follow-up period.

DISCUSSION
LV is an uncommon dermatosis also known as “livedo vasculitis”, “livedoid vasculitis” or “livedo reticularis with summer ulcerations”. Although this condition has previously been classified as a localized vasculitic process, it appears instead to be a cutaneous vasculopathy (1). The disorder most commonly affects young to middle-aged women and the lesions are mostly located on the lower legs. The clinical appearance is commonly identified as atrophic, stellate, ivory-to-white, scar-like plaques stippled with telangiectasia and surrounded by hyperpigmentation, known as atrophie blanche (2). Histological features are typical, revealing segmental hyalinization, endothelial proliferation and thrombosis of the upper and mid-dermal blood vessels. Neutrophilic infiltrate of the blood vessel walls and fibrinoid necrosis, the hallmark of true vasculitis, are absent. It can be associated with autoimmune disorders, such as systemic lupus erythema-
mune-mediated mechanisms are considered to be responsible for vascular damage, leading to a true small vessel vasculitic process. However, in most cases, an underlying disorder is not detectable, and is known as “idiopathic” LV.

Recently, LV is considered to be an occlusive thrombotic process due to a hypercoagulable state. There have been several reports of LV associated with inherited thrombophilias (5), including factor V Leiden mutation (6, 7), heterozygous protein C deficiency (8, 9), homozygous hyperhomocysteaemia (10–12), prothrombin G20210A gene mutation (13), and plasminogen activator inhibitor-1 promoter homozygosity (14). Isolated deficiencies of coagulation factor inhibitors or increased levels of coagulation factors detected at screening do not fully explain dermal vessel thrombosis and the details of the thrombogenic mechanism. As technology advances in the coagulation laboratory, the thrombogenic mechanism may be apparently elucidated.

LV involving a wide area of the body is occasionally described (3, 15). Cardoso et al. (3) reported a patient with Sjögren’s syndrome who showed extended LV in the upper limbs accompanied by hypercoagulability. The other case, showing LV on most of the body, revealed no abnormal laboratory findings (15). The skin manifestation in our patient extended over the lumbar area and upper limbs beyond the lower extremities; however, no abnormalities were detected in the autoimmune profile and coagulation parameters. Therefore, the cause or underlying condition in our patient is unclear.

The therapy of idiopathic LV is usually difficult and disappointing (16), as shown by the wide lists of treatments with variable clinical outcome: aspirin, pentoxifylline, dipyridamole, folic acid (in patients with methylenetetrahydrofolate reductase mutation), corticosteroid, immunosuppressant, warfarin sodium, heparin, hyperbaric oxygen, tissue plasminogen activator infusion and intravenous immunoglobulin (17). These agents are often used in combination with several therapies in patients who fail to respond to a single agent. Our patient was treated successfully with oral aspirin and ticlopidine, and has remained asymptomatic without further treatments. Finally, since LV can be a manifestation of other systemic diseases, we should try to determine the underlying conditions. Idiopathic LV is the final diagnosis.

REFERENCES