Idiopathic Livedoid Vasculopathy: About a case report

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Abstract

Livedoid Vasculopathy (LV) is a chronic condition that presents as recurrent, painful, ulcers of the lower leg, ankle, or dorsal foot. It is an uncommon disease entity. We present a case of livedoid vasculopathy in a young woman.

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Short Title: Livedoid Vasculopathy: About a case report.

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Dear Editor;

Livedoid Vasculopathy (LV) is a chronic condition that presents as recurrent, painful, ulcers of the lower leg, ankle, or dorsal foot. It is an uncommon disease. We present a case of LV in a young woman.

A 30-year-old woman with no past medical history, presented in our dermatology department for a 4-years history of painful ulcerated lesions with serosanguineous crusts in the dorsal surface of her feet, that appeared in summer season then healed spontaneously on winter. She did not present Raynaud's phenomenon or other systemic symptoms. She had no history of spontaneous recurrent abortions or deep vein thrombosis. Physical examination revealed several, painful, well-defined, purpuric ulcerations, erythematous red-violaceous plaques and stellate, porcelain white scars surrounded by punctate telangiectasia and brownish pigmentation around ankle and dorsum of her feet **(Figure 1a, 1b, 1c)** . All peripheral pulses were palpable. Examination with dermoscopy revealed central crusted violaceus ulcers surrounded by peripheral ivory white areas

(Figure 2a) . Skin biopsy showed a superficial dermal vessel with thrombi, and hyalinized vessel walls **(Figure 2b)** . Antinuclear antibodies and cytoplasmic antineutrophil cytoplasmic antibodies were negative. A coagulation investigation including prothrombin activity, cephalin and thrombin times, functional fibrinogen, protein C and protein S, antithrombin III, Factor V Leiden mutation, anticardiolipin and anti-2-glycoprotein I antibodies, lupus anticoagulant, cryoglobulins and cold agglutinins was negative. Serum homocysteine levels were normal. The diagnosis of LV was made based on clinics and histologic findings. The patient was treated with anticoagulation with good improvement within two weeks **(Figure 3a, 3b)** .

Livedoid Vasculopathy (LV) is a noninflammatory condition, characterized by recurrent painful purpuric ulcerations around the ankle and dorsum of the feet that heal with stellate porcelain white scars called "atrophie blanche". It is an uncommon disease entity affecting 1% to 5% of the population **(1)** classically presents in a middle-aged woman. it does occur in males, and there are a few case reports of disease in children **(2)** . It was originally described as a clinical manifestation of vasculitis. However, at present, the main physiopathogenic mechanism considered is a vasoocclusive phenomenon due to intraluminal thrombosis of dermal venules. In general, there are three main factors that predispose to thrombosis: endothelial damage, changes in the blood flow and blood disorders leading to hypercoagulability **(3)** . The presence of lupus anticoagulant and/or anticardiolipin antibodies may also be demonstrated. Although there is no consensus, and many cases such our patient, remain idiopathic. Histopathology reveals segmental vessel wall hyalinization of dermal blood vessels with fibrinoid deposition in lumen and minimal perivascular lymphocytic infiltration **(1)** . Polyarteritis nodosa (PAN) is a close differential diagnosis. Skin biopsies that include the deep subcutaneous tissue are required for diagnosis. Its shows a necrotizing vasculitis affecting medium-sized vessels **(4)** . However, the coexistence of clinical and histological features of cutaneous PAN and LV has been described **(5).** Several therapeutic approaches have been employed with varying degrees of success: hyperbaric oxygen therapy, Anticoagulant and Vasodilator drugs are the most efficient. Livedoid vasculopathy remains an enigmatic condition in terms of its etiopathogenesis. More investigations are needed to explain it.

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Figure legends:

Figure 1a, b, c: Purpuric ulcerative lesions, with porcelain white scars around ankle and dorsum of feet.

Figure 2a: central, crusted, violaceus ulcers surrounded by peripheral, ivory, white areas in dermoscopic examination.**Figure 2b:** HE*200: Superficial dermal vessel with thrombi

Figure 3a, b, c: cutaneous lesions after two weeks of treatment.

Authors contribution: Marwa Thabouti wrote the manuscript and contributed to the management of the patient. Marouane Ben Kahla and Nadia Gahriani Fetoui revised the manuscript. Badreddine Sriha contributed to the anatomopathological examination. Mohamed Denguezli reviewed the manuscript and gave final approval.

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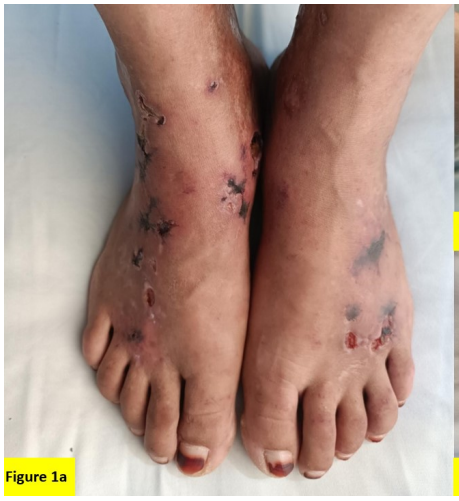

Figure 1a

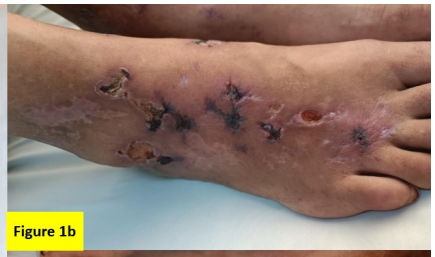

Figure 1b

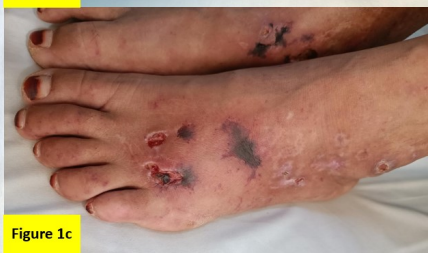

Figure 1c

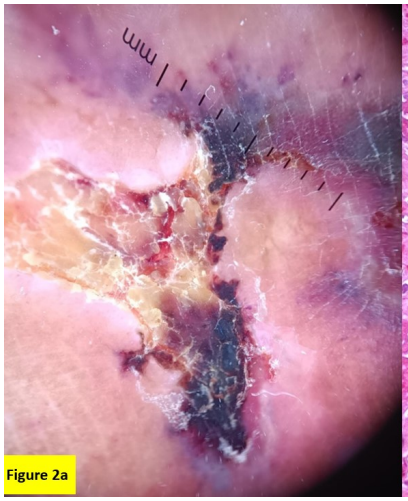

Figure 2a

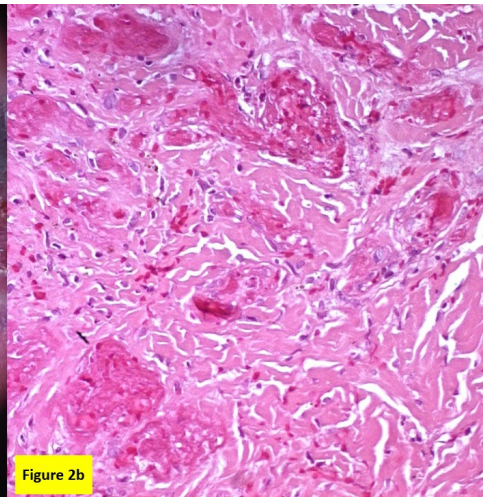

Figure 2b

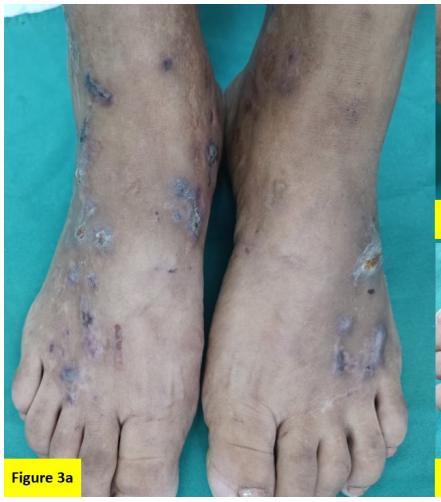

Figure 3a

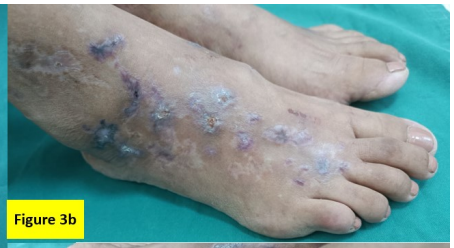

Figure 3b

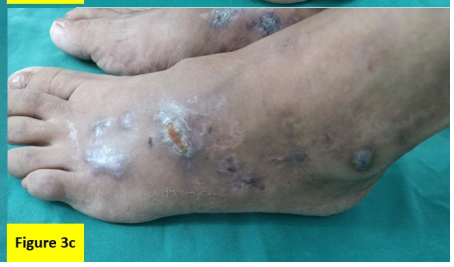

Figure 3c