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Case Report .

Livedoid Vasculopathy: A Case Report and Review of Literature

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ABSTRACT

Livedoid Vasculopathy (LV) is a chronic vascular disorder due to vascular coagulopathies that is characterized by recurrent painful ulcers that usually involves the lower legs especially the ankles. Here in we report a case of LV in a 50-year- old obese female presented with 6-month-history of persistent painful skin ulcers. She is a known to have hypertension on treatment. She also has history of deep vein thrombosis (DVT) 5 years ago (not on any treatment now). Skin examination showed multiple small ulcers above the medial malleolus of the left ankle. Skin biopsy showed pseudoepitheliomatous hyperplasia of the epidermis. The dermis showed mild perivascular lymphohistiocytic cellular infiltrate, hyalinization of blood vessel walls, cannonball tufting of the blood vessels and extraverted RBCs. Thrombi were also seen within some blood vessels.

Blood investigations revealed CBC, LFT, RFT, anticardiolipin antibodies, lupus anticoagulant, prothrombin time, activated partial thrombin time, and protein C serum level to be negative or within normal limits. Based on the clinical, histopathological, and laboratory findings, a diagnosis of LV secondary to venous insufficiency was made. In addition to local wound care, the patient was started on pentoxifylline 300 mg tablets 4 times a day. The ulcers completely resolved after 3 months of the treatment. The lesions recurred again after stopping the treatment, so pentoxifylline was reintroduced again.

Keywords: Livedoid Vasculopathy, Coagulopathies, Skin Ulcers.

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INTRODUCTION

Livedoid vasculopathy (LV) as the name implies is a vasculopathy and not vasculitis. LV is a rare chronic non-inflammatory thrombotic vascular disorder characterized by recurrent painful ulcers that usually involves lower legs especially ankles. It is common in females between the ages of 15 to 50 years old. It is divided into primary LV (idiopathic) and the secondary LV, which is associated with chronic venous hypertension or hypercoagulable states in 50% of patients. The causes of hypercoagulable states include antiphospholipid syndrome, protein C and S defects, factor V Leiden mutation, prothrombin mutations, hyperhomocysteinemia, antithrombin III deficiency, and sticky platelet syndrome.[1-6] The pathogenesis is not known but it is thought to be due to local defect (primary LV) or systemic defect (secondary LV) in coagulation pathway with formation of fibrin thrombi within the dermal blood vessels. Clinically LV is characterized as painful recurrent purpuric eruptions and symmetric punched-out ulcerations, primarily located on the lower extremities, and overall on the malleoli. The ulcers heal after 3 to 4 months leaving an atrophic, stellate, ivory-to white, scar stippled with telangiectasia and surrounded by hyperpigmentation, known as atrophie blanche, which is characteristic for LV.[1,3,5]

CASE REPORT

A fifty years old obese female presented with 6-month-history of persistent painful skin ulcers. She is a known to have high blood pressure controlled by treatment. She also has history of deep vein thrombosis (DVT) 5 years ago (not on any treatment now). Review of system unremarkable and there was no similar case in the family. Skin examination showed multiple small ulcers above the medial malleolus of the left ankle (figure 1). Pedal pulses are palpable. No inguinal lymphadenopathy. Skin biopsy showed pseudoepitheliomatous hyperplasia of the epidermis. The dermis showed mild perivascular lymphohistiocytic cellular infiltrate, hyalinization of blood vessel walls, cannonball tufting of the blood vessels and extraverted RBCs. Thrombi were also seen within some blood vessels (figure 2). Blood investigations revealed CBC, LFT, RFT, anticardiolipin antibodies, lupus anticoagulant, prothrombin time, activated partial thrombin time, and protein C serum level to be negative or within normal limits. Based on the clinical, histopathological, and laboratory findings, the diagnosis of LV secondary to venous insufficiency was made. In addition to local wound care, the patient was started on pentoxifylline 300 mg tablets 4 times a day. The ulcers completely resolved after 3

months of the treatment. The lesions recurred again after stopping the treatment, so pentoxifylline was reintroduced again.

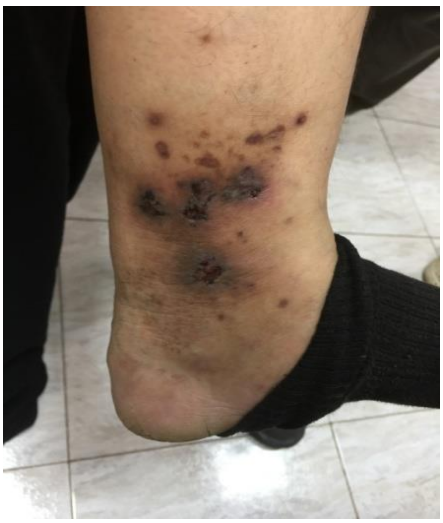

Figure 1: Multiple small ulcers and atrophic scars above the medial malleolus of the left ankle.

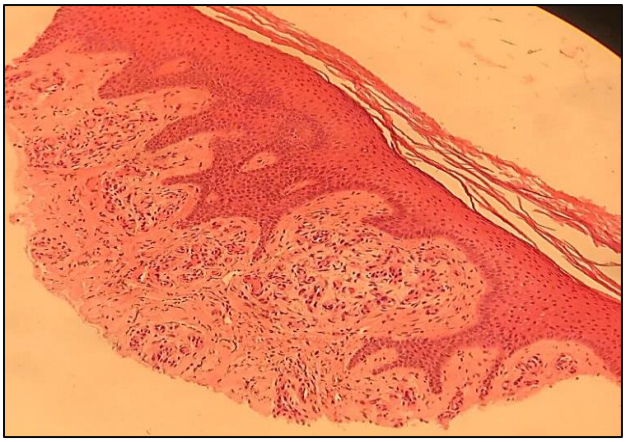

Fig 2: Skin biopsy showing pseudoepitheliomatous hyperplasia of the epidermis. The dermis showed mild perivascular lymphohistiocytic cellular infiltrate, hyalinization of blood vessel walls, cannonball tufting of the blood vessels and extraverted RBCs. Thrombi were also seen within some blood vessels.

DISCUSSION

LV is a chronic non-inflammatory vascular disorder due to thrombotic occlusion of dermal small blood vessels.[1]

The differential diagnosis of leg ulcers includes chronic venous ulcers, leukocytoclastic vasculitis, pyoderma gangrenosum, cutaneous polyarteritis nodosa, factitious dermatitis which were easily ruled out in our case.[5] Sickle cell disease and hydroxyurea-related leg ulcers can mimic LV. Porcelain white scars in the setting of venous disease but without punctate ulcerations should be considered an unrelated syndrome.[3,5] Venous ulcers (VU) is the main differential diagnosis in our case. Although VU are located above the medial malleolus similar to LV, however, VU tends to become larger, shallow with a border that is typically irregular, the bed is covered by a yellow fibrinous exudate, and lack the other characteristic features of LV.[6-8] Our case has LV secondary to venous insufficiency. The risk factors for chronic venous insufficiency in our patient include the age, female gender, obesity and hypertension. Treatment of LV is challenging, however the current approach is based on drugs that stimulate endogenous fibrinolytic activity, inhibit thrombus formation or cause vasodilatation.[3]

Multiple treatments have been proposed, including low-dose aspirin, dipyridamole, hyperbaric oxygen, and intraveneous immunoglobulin.[4,7] Many studies have also investigated the use of anticoagulant drugs such as warfarin, unfraction¬ated heparin, or low molecular weight heparin, all of which have been found to help with pain relief and hasten healing. Another treatment modality is pentoxifylline, which used in our patient and she responded successfully but with recurrence of the lesion upon discontinuation. It has been proven to lower the blood viscosity and im¬prove flow.[6] The use of topical, systemic and intralesional corticosteroids has shown little or no efficacy5. Anabolic agents such as danazol and stanozolol have been helpful in some instances, as has PUVA therapy. For recalcitrant disease, prostanoids (e.g. alprostadil [PGE-1]), rivaroxaban, and IVIg have been used.[2] The pain is best managed using tricyclic antidepressants. If these are unsuccessful or not tolerated, gabapentin, pregabalin, or carbamazepine can be added.[7]

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