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LIVEDOID VASCULOPATHY: CASE REPORT AND BRIEF REVIEW OF ITS THERAPY

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ABSTRACT

A 58-year-old man, with purpuric pericapillary lesions in the lower limbs, for 20 years, associated with local hyperesthesia. The condition evolved with local burning extending to the back of the feet. Pentoxifylline, acetylsalicylic, diosmine, syvastatin and cilostazol were used. In the current consultation, he had erythema-vinous macules in the distal part of the thighs and on both legs ranging from a few mm to 2 cm in diameter associated with continuity solutions and white atrophies. With a suspicion of Livedoid Vasculopathy, laboratory tests were performed: liver and kidney function tests, glucose, alkaline phosphatase, blood count, serum complement, ANF (antinuclear factor), urine summary, anti-RNP, native anti-DNA, anti-SLC-70, anti-U1 RNP, anti-ACA, dehydrogenases, fibrinogen, serology for syphilis, hepatitis B and C and HIV, all exams were normal, The CH50 was at 80 U/ml (normal up to 60 U/ml). The histopathological study of cutaneous fragment confirming the initial suspicion. Therapy for livedoid vasculopathy includes intravenous pulse immunoglobulin which has been shown to be effective and safe, providing responses, in some cases, from the first cycle (Kim et al 2015), and danazol, with high fibrinolytic power.

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INTRODUCTION

A 58-year-old man, with purpuric pericapillary lesions in the lower limbs, for 20 years, associated with local hyperesthesia. Initially, the suggested treatment was the alternation of hot and cold compresses at the side of the lesions. The condition evolved with local burning extending to the back of the feet. Pentoxifylline, acetylsalicylic, diosmine, syvastatin and cilostazol were used. He quit smoking and alcoholism seven years ago. He has been hypertensive for five years. His daughter had a history of lymphoma five years ago. In the current consultation, he had erythema-vinous macules in the distal part of the thighs and on both legs ranging from a few mm to 2 cm in diameter (Fig. 1A and 1B) associated with continuity solutions and white atrophies. Non-palpable fibular nerves and decreased sensitivity on the side of the right foot. With a suspicion of Livedoid Vasculopathy (LV) laboratory tests were performed: liver and kidney function tests, glucose, alkaline phosphatase, blood count, serum complement, ANF (antinuclear factor), urine summary, anti-RNP, native anti-DNA, anti-SLC-70, anti-U1 RNP, anti-ACA, dehydrogenases, fibrinogen, all exams were normal. Serology for syphilis, hepatitis B and C and HIV, non-reagents. The CH50 was at 80 U/ml (normal up to 60 U/ml). The histopathological study of cutaneous fragment demonstrated hyalinization of the vascular wall with fibrin thrombi, red blood cells extravasation and focal necrosis of the epidermis (Fig. 1C)

confirming the initial suspicion. LV is a disease that occurs by thrombo-occlusives, processes of cutaneous microcirculation. It's clinical presentation consists of the triad: racemose livedo, painful ulcers in the distal segments of the lower limbs and whitish scars called white atrophies (Marques *et al.*, 2018). The patient in the report, with a personal history of hyperthrombotic states favorable to the occurrence of LV, although undergoing anti-thrombogenic therapy with associations of multiples drugs, including vasodilators and platelet antiaggregants, showed no benefit in the course of the disease (Micieli & Alavi, 2018). A therapeutic possibility is the investigation of lipoprotein levels the [Lp(a)], since the studies suggests a correlation with the LV. Pathophysiology studies show that LP(a) has structural component similar to plasminogen, competitively binds to it's receptors, resulting in reduced fibrinolysis and, therefore, higher thrombogenic risk. One possible treatment for LV is the anabolic steroid with fibrinolytic action, the danazol. A study conducted with others patients with age range and disease evolution time similar to the case in question, demonstrated a positive effect in the reduction of Lp(a)'s levels and LV-related symptoms (Criado *et al.*, 2015). The therapies for LV are usually focused on fibrinolytic, antithrombotic and vasodilator drugs. Aspirin, pentoxifylline, prostacyclin, activated tissue plasminogen, low molecular weight heparin, dipyridamole, nicotinic acid and phenformine has been tested (Sami *et al.*, 2003).

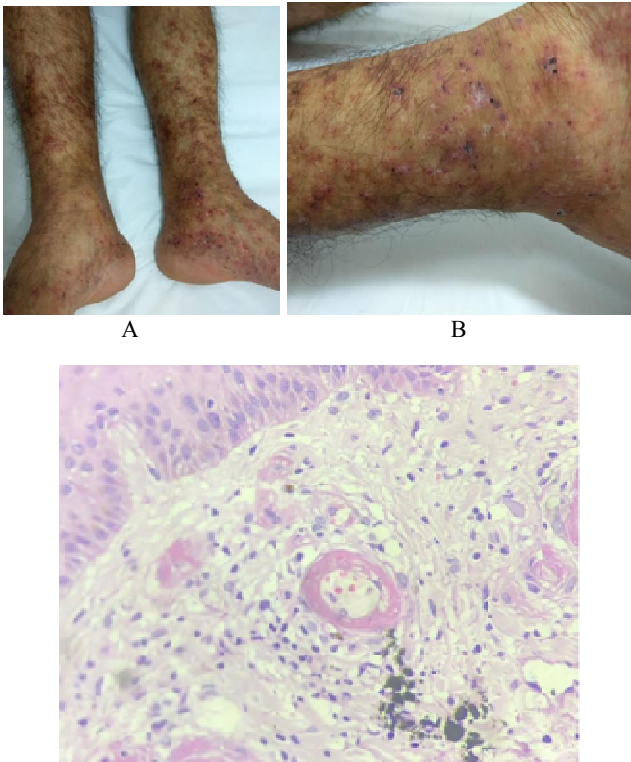


Figure 1. (A), Erythematous-vinous macules on both legs. (B), Erythematous-vinous macules ranging from a few mm to 2 cm in diameter. (C), fibrin thrombi in vascular wall and focal necrosis of the epidermis

Because the patient is already under treatment for LV, there is the possibility of instituting intravenous immunoglobulin (IVIg) therapy in pulses that proves to be effective and safe, providing answers, in some cases, from the first cycle (Kim *et al.*, 2015). The patient described did not respond to previously prescribed treatments, as IVIg therapy is a less economically accessible alternative, the danazol becomes a viable option due to its high fibrinolytic power.

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