Case Report

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Livedoid Vasculitis: Case Report

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ABSTRACT

The natural history of livedoid vasculitis is one of relapse and remission, with an estimated prevalence of 1:100,000 inhabitants per year, mainly women with a mean age of 30 years. The objective of the present study is to report the case of a patient with livedoid vasculitis and the difficulties in diagnosis and treatment. A 51-year-old white female patient, a 30-year smoker of one pack of cigarettes per day, presented with a chronic ulcer for 9 years on the pretibial surface of the right lower limb after local trauma. During this period, the lesion progressed with an increase in extension, affecting the lateral surface of the leg and becoming pale and painful to manipulation. The biopsy confirmed the diagnosis of livedoid vasculitis, which made it possible to establish a more appropriate therapy, although there is no consensus on one. The patient evolved with good healing of the lesion.

Keywords

Vasculitis, Livedoid, Treatment, Diagnosis.

Introduction

Livedoid vasculitis is a rare condition characterized by thrombosis of dermal vessels without significant inflammatory infiltrate, leading to the appearance of recurrent and painful ulcers in the lower limbs. Although the exact etiology of livedoid vasculitis is not understood, it is believed that immunological and vascular factors play important roles in the development of the disease. Triggering factors, such as local trauma, infections and hematologic disorders, can trigger or exacerbate the symptoms of the condition [1,2]. The blood vessels undergo thickening, local fibrosis and degeneration of the vascular layers, being a veno-occlusive phenomenon due to intraluminal thrombosis of the dermal venules [3,4].

The disease is chronic and can be primary when it is not related to other diseases, while secondary disease can be related to systemic autoimmune diseases and hypercoagulable states, such as systemic lupus erythematosus (SLE), systemic sclerosis, rheumatoid arthritis, Sjogren's syndrome and antiphospholipid syndrome (APS). However, in 50% of cases there is an underlying thrombophilia with coagulation or fibrinolysis abnormalities [5].

The natural history of livedoid vasculitis is one of relapse and remission, with an estimated prevalence of 1:100,000 inhabitants per year, especially women with an average age of 30 years. Furthermore, it tends to affect mainly the lower limbs and patients report intense pain due to its ischemic nature, limiting their daily activities [1,4,6].

It can evolve in several stages, with the initial formation of livedo racemosa, followed by the formation of erythematous-wine-colored and purpuric papules, which are painful and pruritic. They can evolve into vesicles with hemorrhagic content and later, ulcerations occur, sometimes multiple and may converge into

large ulcers, which have “star-shaped” edges, an ivory color and an atrophic center [4].

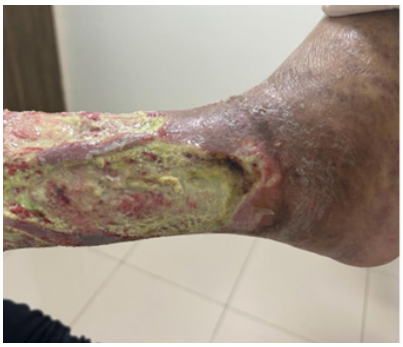
Livedoid vasculitis mainly affects young and middle-aged women, in a ratio of 3:1 and the average age of onset is around 30 years, and it can manifest up to 53 years of age [1,2]. Diagnosis is a medical challenge since the clinical manifestations of livedoid vasculitis resemble other skin diseases, delaying histological diagnosis by 3-4 years.

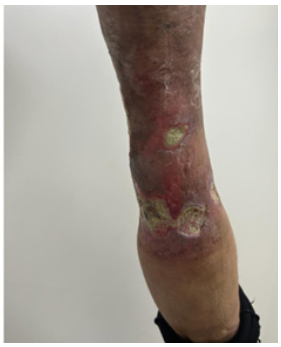
There are several treatments for this condition, but no standardized and evidence-based therapeutic strategy has been published. The aim of this study is to report the case of a patient with livedoid vasculitis and the diagnostic and treatment difficulties.

Case Report

A 51-year-old white female patient, a 30-year smoker who smoked one pack of cigarettes a day, had a chronic ulcer for 9 years on the pretibial surface of the right lower limb after local trauma. During this period, the lesion progressed with an increase in extension, affecting the lateral surface of the leg and becoming pale and painful to manipulation. As shown in figures one and two. Due to the difficulty in healing, even with distal pulses present, it was decided to perform an arteriography of the limb, which showed spastic arteries, with no conditions for angioplasty. A sympathectomy was performed on the right due to refractory pain, with partial and momentary improvement of the symptoms.

She was hospitalized several times due to the pain, which improved with the use of a heparin pump and returned after its discontinuation. The lesion was debrided and a biopsy was collected, which showed intravascular hyaline thrombi in the papillary dermis, suggestive of livedoid vasculitis. After diagnosis, acetylsalicylic acid, pentoxifylline and rivaroxaban were prescribed, in addition to medications for pain control. The patient showed significant improvement in symptoms, walking without difficulty and the lesion with a granulation base, figures 1 to 4.





Figures 1 and 2: Lesions before diagnosis and vasculitis, the first referring to the anterolateral aspect of the limb and the second, the posterior aspect.

Regarding diagnostic care, deep punch or excision biopsies of 4 to 6 mm are suggested at the ulcer margin, including surrounding normal tissue. In terms of histology, the patient presents superficial dermal edema, extravasation of red blood cells and intraluminal capillary thrombi, with minimal associated inflammatory infiltrate or absence thereof [1].

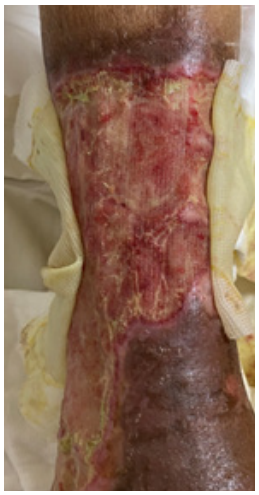


Figure 3: Injury in the 1st postoperative day, after debridement and biopsy collection, associated with clinical treatment for livedoid vasculitis.

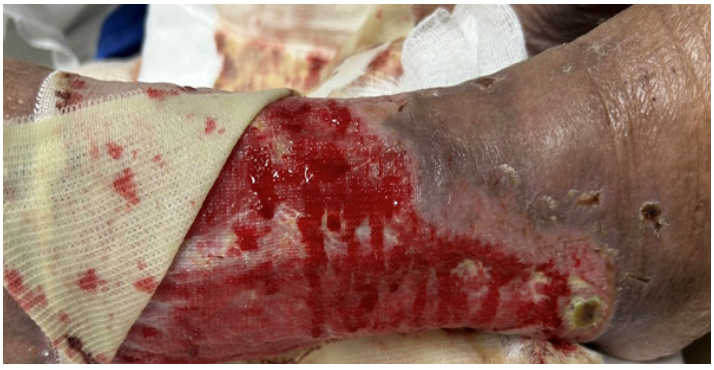


Figure 4: Injury on the 10th postoperative day with continued clinical treatment for livedoid vasculitis.

Discussion

This report presents the diagnostic and therapeutic difficulties of livedoid vasculitis, where biopsy was essential in this definition. The warning is about the difficulties in healing and the limiting pain. The use of heparin was one of the therapeutic options adopted throughout this period during hospitalizations and it brought momentary improvement while it was being used. However, when the anticoagulation was withdrawn, the pain recurred.

The biopsy was decisive in the diagnosis of vasculitis and can better guide the approach to the patient. Regarding the literature, there is no definitive and consensual conduct, but the use of anticoagulants, vasodilators, analgesics and antiplatelet agents has been suggested [5,6]. However, the cure of vasculitis is a challenge over time.

With the emergence of oral anticoagulants, they have become a long-term therapeutic option at reduced doses [7], but the

association with antiplatelet agents may increase the possibility of bleeding and the conduct should be carefully evaluated. However, the association with congenital thrombophilias and hypercoagulation states should be evaluated. Patients with antiphospholipid antibodies (APS) are particularly at increased risk of recurrent thrombotic events during DOAC therapy. Intravenous immunoglobulins (IVIG) are recommended as first-line therapy, but due to their high cost, they should be used in cases refractory to the initial therapy of choice at a dose of 0.5 g/kg/day [5].

Venous ulcers are the main causes of lesions in the lower limbs, but they have more specific locations, mainly associated with perforations and distal reflux of the great saphenous vein, and have significant emotional consequences and are part of the differential diagnosis 8,9. Pain in venous ulcers can be aggravated when infected and bring significant pain impairment 10. In summary, differential diagnosis is essential in the management of lower limb ulcers.

Conclusion

Livedoid vasculitis is a rare cause of lower limb ulcers, but the painful aspect of this lesion and its atypical location in relation to venous ulcers raises a warning regarding the diagnosis, where biopsy with histology is essential for diagnosis.

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