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# Clinical Insights and Diagnostic Dilemmas: Two Cases of Livedoid Vasculitis

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### Abstract

Livedoid Vasculitis (LV) is a thrombo-occlusive vasculopathy involving dermal vessels, especially in the lower extremities. Clinical symptoms of LV are chronic, recurrent, scarring, and painful purpuric ulcers. Diagnosing and providing therapy for LV is a challenge because there are no standard guidelines. We present clinical insights and diagnostic approaches on two cases of LV to improve early diagnosis and prevent misdiagnosis, which were confirmed based on history, dermatological examination, and skin biopsy. In the first case, it was a woman, 23 years old, who came with complaints of a blackish-red rash appearing on both legs. There were scars on several parts of the patient's legs, which felt painful, and the legs looked swollen. On histopathological examination, fibrin deposition in the vessel walls, endothelial proliferation, and intraluminal hyaline thrombin were found. In the second case, it was a man, 19 years old, who came with complaints of pain when walking accompanied by wounds on both lower legs. On histopathological examination, fibrin deposition in the vessel walls, endothelial proliferation, and intraluminal hyaline thrombin were found. The conclusion from the histopathology results was LV. After receiving therapy, both cases showed improvement: swelling in the legs was reduced, red and black rashes began to disappear, ulcers improved, pain decreased, and scars became blurred. These two cases provide examples of success in diagnosing LV. Being able to diagnose LV early and correctly is very important so that adequate therapy can be given and good outcomes can be achieved.



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## 1. Introduction

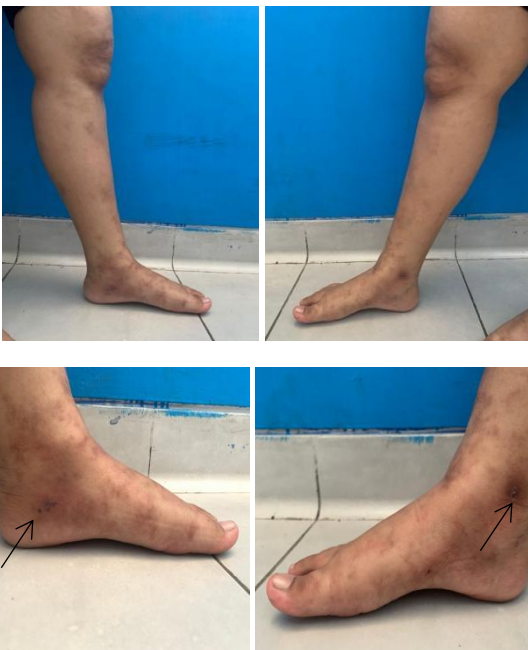
Livedoid Vasculitis (LV) is a thrombo-occlusive vasculopathy that involves the dermal vessels, especially in the lower extremities. Typical clinical symptoms of LV are chronic, recurrent, scarring and painful purpuric ulcers [1, 2]. This disease occurs 3 times more often in women than men, especially in patients aged 15 to 50

years. LV is a rare case with an estimated incidence of 1:100,000 [3].

The pathogenesis of LV is currently still unclear. It is thought that this occurs due to an increase in local and systemic thrombotic activity and a decrease in fibrinolytic changes, resulting in coagulation disorders and fibrin thrombus formation in superficial dermal blood



**Figure 1.** On dermatological status before treatment: Macules and erythematous patches accompanied by ulcers and covered with crusts (arrow sign).



**Figure 2.** Follow up at 1 month: Atrophie blanche appears at the location where there was previously a wound (arrow sign). The lesions have dried up and pain has decreased.

vessels. The formation of fibrin thrombi causes tissue hypoxia in the dermis, causing wounds that are difficult to heal and scarring [2, 4].

There are no standard guidelines for diagnosing LV [5]. Diagnosis is made based on dermatological examination, clinical and histopathological findings [1]. The characteristic features of LV are livedo racemosa, very painful ulcers located in the lower extremities, and white porcelain-like scars (*atrophie blanche*) [1].

Histopathologically, it shows intraluminal fibrin deposition and thrombosis, segmental hyalinization, and endothelial proliferation [2]. Accuracy in diagnosis and therapy can reduce pain, recurrence, scar tissue, disability, and other complications [5].

Diagnosing and providing therapy for LV is a challenge for healthcare practitioners. The clinical appearance of other violaceous skin diseases, overlap in nomenclature, and other ulcerative disorders is a source of confusion. Here we present clinical insights and diagnostic approaches on two cases of LV to improve early diagnosis, prevent misdiagnosis and achieve good outcomes.

## 2. Cases

### 2.1. Case 1

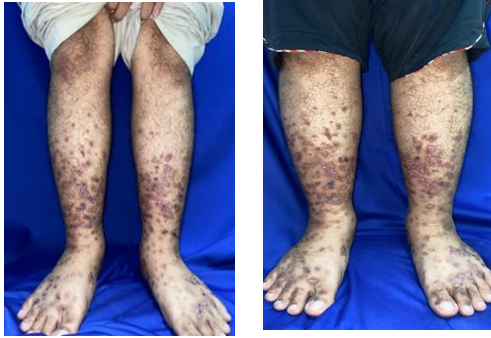
A 23-year-old female was referred to dermatology and venereology polyclinic Dr. Zainoel Abidin General Hospital with complaints of a blackish-red rash appearing on both legs. There were scars on several parts of the patient's legs, which felt painful and looked swollen. The rash appeared in 2020. The rash initially appeared in the ankle area and then spread to the calf area (Figure 1). No family member had a similar complaint.

The general examination was normal, with a visual analog scale of 6. On dermatological status, In the region of the tibia dextra et sinistra, malleolus dextra et sinistra there are macules and erythematous patches with ill-defined borders and irregular edges accompanied by ulcers and covered with crusts. Multiple numbers, lenticular to guttate size and nummular shape with some confluent lesions, discrete arrangement, bilateral distribution. Atrophie blanche appears at the location where there was previously a wound. On histopathological examination, fibrin deposition in the vessel walls, endothelial proliferation, and intraluminal hyaline thrombin were found. The conclusion from the anatomical histopathology results was LV. From the history, clinical features, and skin biopsy, we diagnosed the patient with LV.

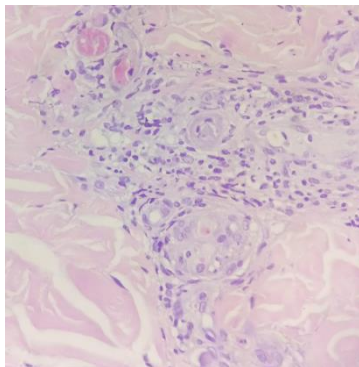
The patient was given oral clindamycin 300mg three times a day for a week, oral methylprednisolone 4mg twice a day, and apply mupirocin cream twice a day. After receiving therapy for one months, the swelling in both legs has begun to decrease, accompanied by lesions that have dried up and pain has decreased (Figure 2).

### 2.2. Case 2

Male, 19-year-old, referred to dermatology and venereology polyclinic dr. Zainoel Abidin General Hospital with complaints of pain when walking accompanied by wounds on both lower legs. The first complaint appeared



**Figure 3.** On dermatological status before treatment: Erythematous macules with sharp borders, erosion, crusts and multiple ulcers. Typical livedo racemosa lesions also found on the lower legs with erythematous to purple network pattern.



**Figure 4.** Histopathological examination showed fibrin deposition in the vessel walls, endothelial proliferation and intraluminal hyaline thrombin.

five months ago, and has gotten worse over the past two weeks. At first, the rash was still small, red, and then turned black. One month later, the patient complained again of the rash which had been at the base of the foot, slowly rising to the heel and ankle. The rash is accompanied by blisters and ulcers, the patient also complains of swelling in both legs (Figure 3). There was no known previous autoimmune disorder and the family did not experience similar complaints like this.

The general examination was normal with a visual analogue scale of 5. On dermatological status, in the tibia-ankle region right and left, erythematous macules appeared, with sharp borders, erosion, crusts and multiple



**Figure 5.** Follow up at three months: Atrophie blanche appears at the location where there was previously a wound (arrow sign). The ulcers showed improvement and pain has decreased.



multiple ulcers, the size of the guttate-nummular lesions, discretely distributed in a symmetrical distribution. On histopathological examination, there were found to be hyperkeratosis, and the subcutis appeared to be thickened blood vessels with fibrin deposition (thrombosis), a few of which were lymphocytes. The conclusion from the histopathology results was LV (Figure 4). We diagnose the patient with LV.

The patient was given oral clindamycin 300mg three times a day for a week and oral methylprednisolone 8mg twice a day. To treat wounds, normal saline compresses are given twice a day for 10 minutes, and mupirocin cream is given twice a day. After receiving therapy for three months, the swelling in both legs has begun to decrease, accompanied by lesions that have dried up and pain has decreased (Figure 5).

### 3. Discussions

An accurate diagnosis of LV requires patient history data, physical examination, histopathology, and laboratory parameters [3]. In the two cases above, there were complaints of pain when walking accompanied by injuries to both lower legs. A dermatological examination of both dorsal pedis tibialis anterior regions revealed macules accompanied by erythematous patches with blackish ulcers and crusts, and in some areas, there was atrophie blanche, which was in accordance with the clinical features of LV.



LV is characterized by three clinical symptoms, namely livedo racemosa, atrophie blanche, and ulcers which have a tendency to heal very poorly and feel painful [1]. Livedo racemosa is a lesion caused by reduced blood flow followed by hypoxia and ischemia in the local tissue so that a reddish rash appears as a discontinuous circle. Ulcerations are often very small and very painful. Changes in the form of linear or angular macules, papules, or nodules that are deep and not very visible or erythematous. Other clinical findings are usually ulcers with severe pain. These ulcers take months to heal, eventually forming smooth, ivory-white atrophic plaques known as atrophie blanche and are often surrounded by hyperpigmentation and telangiectasias [6].

Apart from a complete physical examination, a laboratory examination is needed in the form of a complete blood count, coagulation tests, autoimmune markers such as anti-nuclear factor, anti-DNA antibodies, and it is very important to do a Doppler ultrasonography on the lower limbs with the aim of detecting chronic venous-biligic insufficiency [5].

In both cases, the biopsy results showed hyperkeratosis and the subcutis showed thickened blood vessels and few lymphocytes. Proper histopathological examination can also differentiate LV from vasculitis. The main histopathological characteristic of biopsies in LV is focal thrombosis within the vessel lumen in the distal dermis layer. Capillaries are generally affected predominantly, followed by venules and arterioles. Capillaries may also have intramural fibrin deposition while the lumen is still open and well. Thrombosis is usually limited to the upper to middle dermis and rarely occurs in the subcutaneous fatty tissue. The epidermis is not involved except in areas of ulceration, or may become thinner in areas of atrophie blanche, whereas vasculitis is characterized by infiltration of inflammatory cells that cause damage to the blood vessel walls [1, 2].

The differential diagnoses in LV are polyarteritis nodosa (PAN) and sneddon syndrome (SS). Signs and symptoms of PAN usually involve the skin and peripheral nerves. On the skin, purpura, livedoid reticularis, subcutaneous nodules, and necrotic ulcerations can be found. Livedo reticularis (LR) is a bluish discoloration (livid) of the skin that occurs in a net-like pattern. The location of clinical manifestations on the skin is usually limited to the lower extremities. Sneddon syndrome (SS) is a rare non-inflammatory thrombotic vasculopathy characterized by livedo racemose and no ulcers [5]. The main diagnostic criteria for excluding other differential diagnoses are typical histopathological findings on skin biopsy. Histopathological examination on PAN showed necrotizing vasculitis with nodules (mid-size and small

arteries) and secondary thrombosis, whereas on SS there was focal proliferation of subendothelial muscle cells [1].

There are no standardized and evidence-based therapeutic strategies in LV [1]. In both cases above, the patient received drugs such as methylprednisolone, clindamycin, and mupirocin. Most therapies aim to improve clinical manifestations and reduce pain. Treatment that can be done is bed rest, analgesics, low-dose heparin, and platelet aggregation inhibitors. Pain can be relieved and healing accelerated with systemic glucocorticoids. LV therapy obtained from the literature consists of antiplatelets and anticoagulants, fibrinolytic agents, vasodilators, phototherapy with PUVA, immunosuppressants (corticosteroids, cyclosporine) and intravenous immunoglobulin. Pain relief can be achieved with the use of non-steroidal anti-inflammatory drugs (NSAIDs) and tricyclic antidepressants. Wound care uses general principles of wound healing and management [4, 7].

These two cases provide examples of success in diagnosing LV by taking a clinical approach based on the patient's history, physical examination, and supporting examinations such as skin biopsies and other laboratory examinations so that appropriate therapy can be given and provide good outcomes. Clinically, both patients showed improvement after treatment; swelling in the legs is reduced, red and black rashes begin to disappear, ulcers improve, pain decreases, and scars become blurred.

#### 4. Conclusions

Diagnosing and providing therapy for LV is a challenge for provider because there are no standard guidelines for diagnosing. The characteristic features of LV are livedo racemosa, very painful ulcers located in the lower extremities, and white porcelain-like scars (*atrophie blanche*). Although the clinical features of LV are very typical, the diagnosis should only be made based on clinical manifestations and histological findings to exclude differential diagnoses. Histopathologically, it shows intraluminal fibrin deposition and thrombosis, segmental hyalinization, and endothelial proliferation.

A multidisciplinary team approach is important, involving professionals from the departments of pathology, radiology, cardiac surgery, infectious diseases, and oncology for accurate diagnosis and optimal treatment to prevent disability and other complications. Because LV is a rare case, there are few large-scale studies and high evidence levels. It is necessary to conduct randomized controlled trials with a high evidence level to determine the best therapeutic approach in the treatment of LV.

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**Conflicts of Interest:** All the authors declare that there are no conflicts of interest.

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