International Journal of Research in Dermatology
Hidayat A et al. Int J Res Dermatol. 2023 Jan;9(1):36-39
http://www.ijord.com

DOI: https://dx.doi.org/10.18203/issn.2455-4529.IntJResDermatol20223343

Case Report

Successful treatment of refractory livedoid vasculopathy in a patient with systemic lupus erythematosus

Arie Hidayat[1], Nanda Earlia*[1], Mahda R. Liana[2]

[1]Department of Dermatology and Venereology, Faculty of Medicine, Universitas Syiah Kuala, Dr. Zainal Abidin General Academic Hospital, Banda Aceh, Indonesia
[2]Department of Dermatology, Faculty of Medicine, Universitas Syiah Kuala, Dr. Zainal Abidin General Academic Hospital, Banda Aceh, Indonesia

Received: 23 September 2022
Revised: 14 October 2022
Accepted: 17 October 2022

***Correspondence:**
Dr. Nanda Earlia,
E-mail: nandaearlia01@gmail.com

ABSTRACT

Livedoid vasculopathy (LV) also known as livedoid vasculitis is a rare and chronic thrombotic vasculopathy that typically affects the lower extremities. Diagnosis of LV are not well defined and the treatment are widely varied. We report a case of 30 years old female patient diagnosed with Livedoid vasculopathy and systemic lupus erythematous as underlying diseases. Patient presented with chief complain painful ulcer on her bilateral lower extremities. Hypercoagulable state was normal. Skin biopsy found that dermal vessels filled with erythrocytes extravasation and inflammation cell PMN. ANA profile was positive for RNP/Sm suggest that a systemic lupus erythematosus. Patient underwent successful treatment planning for methylprednisolone 16 mg twice daily, hydroxychloroquine 200 mg once daily and methotrexate 10 mg once a week. The ulcer completely resolved after 2 months of the treatment and had no recurrent ulcer. LV is a rare, chronic, and occlusive disease of the veins supplying the upper parts of the skin. Histopathological finding is thickening or hyaline changes in the walls of superficial dermal vessels and luminal fibrin deposition. Red cell extravasation and perivascular lymphocytic infiltrates are expected findings. SLE was underlying condition that cause endothelial injury and leads to thrombosis of the dermal vessels. no treatment guidelines are available for LV. Methylprednisolone, hydroxychloroquine and methotrexate have successful therapeutic response to a patient with ulcerative LV and SLE as underlying diseases.

Keywords: Livedoid vasculopathy, SLE, Treatment

INTRODUCTION

Livedoid vasculopathy (LV) also known as livedoid vasculitis is a rare and chronic thrombotic vasculopathy that typically affects the lower extremities. The term livedoid vasculopathy has been used because primary pathology is hypercoagulability.[1,2] LV is different from inflammatory vasculitis and classified as a coagulating disorder, a vasculopathy, which occurs when a thrombus forms in the arterial lumen and comprises blood flow.[2-4] livedoid vasculopathy is a rare diagnosis with an approximate incidence of 1 in 100,000 per year. It is 3 times more common in females than in males, especially in patients aged 15 to 50 years. A triad of livedoid vasculopathy is livedo reticularis, atrophie blanche and painful small punched out ulcer.[2,3]

Diagnosis of LV are not well defined and the treatment are widely varied.[4] We present a case of LV with SLE in a woman that successfully treated with methylprednisolone, hydroxychloroquine and methotrexate. As a result, the purpose of this study is to report on the use of combination between

methylprednisolone, hydroxychloroquine and methotrexate in planning for the treatment of LV.

CASE REPORT

A 30 years old woman referred to Zainal Abidin general hospital presented with chief complain painful ulcer on her bilateral lower extremities for 3 months. Before the ulcer, she had red rash on bilateral feet which progressed to purpuric patches and blister then become ulcer and finally to atrophy scar. She had this recurrent ulcer condition since 5 years ago. She got methylprednisolone 16 mg once daily from dermatologist in district hospital but the ulcer still recurrent. She also complaint painful joint, recurrent oral ulcer and malaise. there were unknown history of Autoimmune disorder and coagulation disorder. There was no family member with this complain

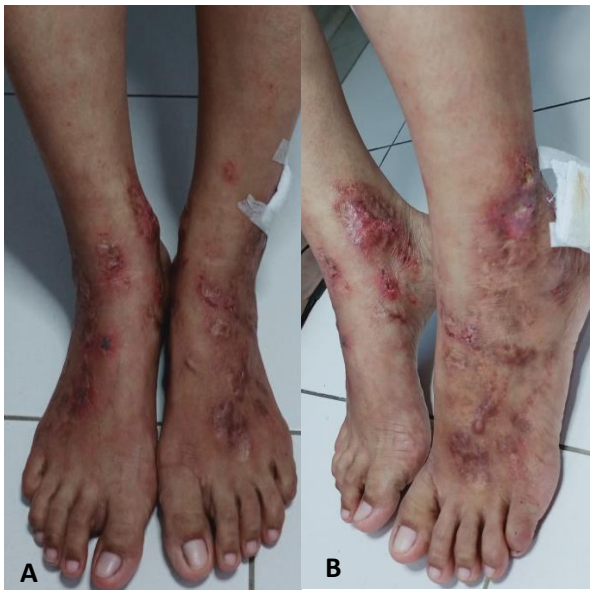

Figure 1: Clinical appearance before therapy. A: purpuric lesion are surrounded by an erythematous ring and atrophic scars. B: Multiple small ulcer on the left and right feet at the time before treatment.

General examinations were normal with visual analogue scale of 6. On dermatologic examination there were patch erythematous-violaceus with multiple ulcers and atrophie blanche in bilateral pedis (Figure 1). for further assessment, punch biopsy of the left foot was performed and found that dermal vessels filled with erythrocytes extravasation and inflammation cell PMN suggest that a livedoid vasculitis (Figure 2). The patient underwent workup for haemostasis and autoimmune disorder. We found that bleeding time, clothing time, international normalized ratio, prothrombin time, activated partial thromboplastin time, D-dimer, and fibrinogen were normal. ANA profile was positive for RNP/Sm suggest that a systemic lupus erythematosus. We diagnose the patient LV with SLE. We performed pedis doppler ultrasonography and found that dorsalis pedis artery and venous dorsalis were normal. This result to exclude the differential diagnosis of venous ulcer. She had treatment planning for methylprednisolone 16 mg twice daily, hydroxychloroquine 200 mg once daily and methotrexate 10 mg once a week. In addition to local wound care, patient was started on topical mupirocin calcium 2% once daily. The ulcer completely resolved 2 months of the treatment. At 4 months follow up, the patient was continued therapy and had no recurrence of her ulcer (Figure 3).

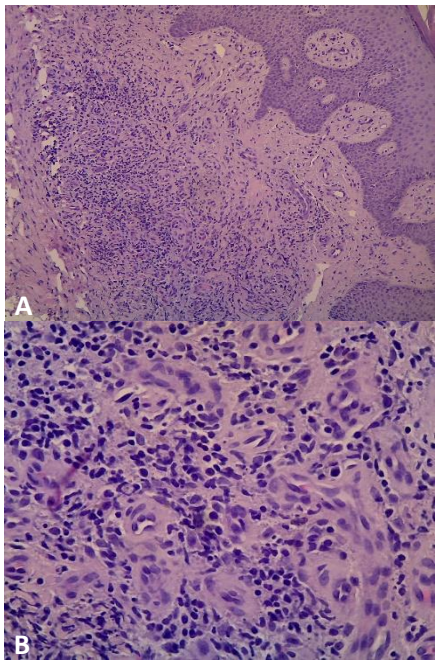

Figure 2: Microscopic appearance of superficial dermal vessels filled with erythrocytes and infiltration of inflammatory cell PMN. (Hematoxylin-eosin staining, (A) original magnification ×200; (B) original magnification ×400).

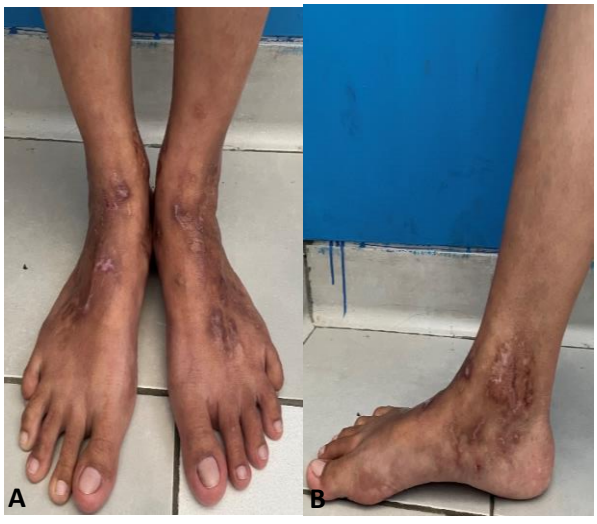

Figure 3: 4 months follow up after received therapy. (A) porcelain-white atrophic scarring tissue; (B) Complete reepithelization of ulcer.

DISCUSSION

LV is a rare, chronic, and occlusive disease of the veins supplying the upper parts of the skin. LV occurs as recurrent, painful ulcers of the lower extremities in association with persistent livedo racemosa that often is deep purple in colour.[5,6] Healing results in sclerotic pale areas that are surrounded by telangiectasias termed atrophie blanche.[7] LV has a 3:1 female predominance with a young-middle age.[7,8] LV can be categorized as primary (idiopathic) or secondary LV, in which a known underlying condition causes the disease.[5] Histopathological finding is thickening or hyaline changes in the walls of superficial dermal vessels and luminal fibrin deposition. Red cell extravasation and perivascular lymphocytic infiltrates are expected findings.[8]

We reported a women 30 years old with diagnosis Livedoid vasculopathy and systemic lupus erythematous. The diagnosis was based on anamnesis, dermatologic examination, laboratory and histopathological finding. We found multiple painful recalcitrant ulcers, white porcelain atrophy and livedo racemosa. The histologic features of red cell extravasation and polymorphonuclear infiltrate in superficial dermal vessels. we made the diagnose of LV from this clinical finding and it may be associated with autoimmune diseases such as systemic lupus erythematosus and hypercoagulable disorders that we can identified from laboratory blood test. In this case we found the positive ANA profile as a marker of systemic lupus erythematosus (SLE). Differential diagnoses include venous statis ulcers, systemic vasculitis, peripheral arterial disease, pyoderma gangrenosum, and trauma.[5-8] Venous stasis ulcer is the main differential diagnoses of our case. It located above the medial malleolus similar with LV but the ulcer has irregular border, shallow and covered by yellow fibrinous exudate and lack of the other LV clinical features.[9] Furthermore, we exclude the venous ulcer from doppler ultrasonography result.

SLE was underlying condition that cause endothelial injury and leads to thrombosis of the dermal vessels.[10] although LV pathogenesis is not yet fully known, recent consensus suggests that changes in the local or systemic control mechanism of coagulation lead to the formation of fibrin thrombi in superficial dermal vessels. the thrombotic effect results from defects in the endothelial cell, platelet dysfunction or enhanced fibrin formation.[5,10] This dermal-vessel thrombosis leads to ischemia and tissue necrosis that causing ulceration and pain. Low tissue perfusion further leads to poor wound healing.[5,6]

Treatment of LV is challenging, no treatment guidelines are available for LV. Current treatment paradigms are based on low levels of evidence, primarily case reports and case series. This case provides an example of a successful therapeutic response to methylprednisolone, hydroxychloroquine and methotrexate in a patient with ulcerative LV and SLE as underlying diseases. The patients give a good response to the therapy, complete reepithelization in 2 months on treatment and no recurrency until 4 months follow up without discontinued therapy.

Hydroxychloroquine as a LV treatment has a function to reduces inflammatory response. Methylprednisolone has Anti-inflammatory action, antifibrinolytic effect and immunosuppressive effect.[6] MTX has immunosuppressive effect and anti-inflammatory effect that associated with adenosine metabolism. MTX could increase adenosine accumulation. It stimulates endothelial cells and fibroblast to release adenosine that can inhibit neutrophil adhesion.[9]

CONCLUSION

In summary, this was a case of livedoid vasculopathy with systemic lupus erythematous as underlying condition. Clinical features and biopsy provided excellent depiction in suggesting the diagnosis of LV. Further test to find out underlying condition is needed. We conclude that combination between methylprednisolone, hydroxychloroquine, and methotrexate can be treatments option for LV with SLE.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Khenifer S, Thomas L, Balme B, Dalle S. Livedoid vasculopathy: thrombotic or inflammatory disease. Clin Exp Dermatol. 2010;35:693-8.
2. Kerk N, Goerge T. Livedoid vasculopathy - a thrombotic disease. Vasa. 2013;42(5):317-22.
3. Criado PR, Rivitti EA, Sotto MN, Valente NYS, Aoki V, de Carvalho JF, et al. Livedoid vasculopathy: an intringuing cutaneous disease. An Bras Dermatol. 2011;86(5):961-77.
4. Micieli R, Alavi A. Treatment for Livedoid Vasculopathy: A Systematic Review. JAMA Dermatol. 2018;154(2):193-202.
5. Bilgic A, Ozcobanoglu S, Bozca BC, Alpsoy E. Livedoid vasculopathy: A multidisciplinary clinical approach to diagnosis and management. Int J Women's Dermatol. 2021;7(5 Part A):588-99.
6. Vaseduvan B, Neema S, Verma R. Livedoid vasculopathy: A review of pathogenesis and principles of management. Indian J Dermatol Venereol Leprol. 2016;82:478-88
7. Soter NA, cutaneous necrotizing venulitis in Kang S, Amagai M, Bruckner AL, Alexander H, Margolis D, et al (eds). Fitzpatrick's Dermatology 9th ed. New York, McGraw-Hill; 2019(2): 2527-2538.
8. Levell NJ, Mukhtyar C, cutaneous vasculitis in Christopher E, barker J, Bleiker T, Chalmers R, et

al. Rooks textbook of Dermatology 9th edition. West Sussex, Willey Blackwell; 2016(4): 102.1
9. Bedoui Y, Guillot X, Sélambarom J, Guiraud P, Giry C, Jaffar-Bandjee MC, et al. Methotrexate an Old Drug with New Tricks. Int J Mol Sci. 2019;20(20):5023.
10. Freitas TQ, Halpern I, Criado PR. Livedoid vasculopathy: a compelling diagnosis. Autops Case Rep. 2018;8(3):34.

Cite this article as: Hidayat A, Earlia N, Liana MR. Successful treatment of refractory livedoid vasculopathy in a patient with systemic lupus erythematosus. Int J Res Dermatol 2023;9:36-9.