

Complete Resolution of Recurrent Livedoid Vasculopathy with Rivaroxaban

Fedaa Saleh Andijani^{1*} and Mohammad Ali Basendwh²

¹Department of Dermatology, King Abdulaziz University, Jeddah, Saudi Arabia

²Department of Dermatology, King Fahad Armed Forces Hospital, Jeddah, Saudi Arabia

*Corresponding Author: Fedaa Saleh Andijani, Department of Dermatology, King Abdulaziz University, Jeddah, Saudi Arabia.

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Abstract

This report describes a case of 28-year-old female patient presented with painful skin ulcers that were diagnosed as livedoid vasculopathy. Livedoid vasculopathy is a thrombotic cutaneous disorder affecting the skin microcirculation. The incidence is rare (1 per 100.000) but the disease has a huge burden on affected cases. Pentoxifylline was administered for three weeks; however, no improvement was detected. Thus, rivaroxaban 10mg once daily was alternatively prescribed. The patient came up after two months with entire resolution of symptoms and full healing of the ulcers.

Keywords: Livedoid Vasculopathy; Rivaroxaban; Pentoxifylline; Skin Ulcers

Abbreviations

CRP: C-Reactive Protein; CBC: Complete Blood Count; ESR: Erythrocyte Sedimentation Rate; INR: International Normalized Ratio; LMWH: Low-Molecular-Weight Heparin

Introduction

Livedoid vasculopathy is a chronic skin disorder that results from closure of the skin microcirculation leading to skin ischemia and ulcerations [1,2]. The incidence of this chronic condition has been estimated at 1 per 100.000 cases, mainly affects females at late adolescence with an average age of 45 years [3]. Initially, the disease presents as painful erythematous papules or plaques on the lower extremities that may ulcerate on a later stage, and subsequently heal, leaving an irreversible stellate white scar recognized as atrophic blanche [1,2]. Thus, the main goal of disease management is to preclude lesions' ulceration as well as repeated incidence of active lesions. To the best of our knowledge, there is no single standard effective treatment for livedoid vasculopathy [4]. Few recently published reports deduced that rivaroxaban can effectively ameliorate disease symptoms [5-7]. Herein, we present a case of young female that experienced complete resolution of livedoid vasculopathy after two months of rivaroxaban administration. More studies are recommended to build conclusive evidence.

Case Presentation

A 28-year-old female patient presented to our clinic with a 10-year history of recurrent painful ulceration over her lower extremities. The patient had no previous history of abortion, venous thrombosis, or stroke. Also, no family history or chronic systemic conditions were reported. On physical examination, there were multiple irregular ulcers with violaceous border and necrotic base over the dorsum of the right foot, surrounded by multiple porcelain white atrophic scars (Figure 1). Laboratory investigations revealed no significant abnormalities; normal ranges of erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and complete blood count (CBC). In addition, no positive profile for autoimmune diseases or thrombophilia. Consequently, the diagnosis of livedoid vasculopathy was attained and pentoxifylline was initially prescribed at 400 mg three times per day. Two weeks later, the patient presented once again

with maculopapular skin rash all over the body. Pentoxifylline dose was reduced to once daily for one week; however, there was no improvement. Therefore, pentoxifylline was discontinued and rivaroxaban was initiated at 10 mg once daily. After two months, the patient attended for a follow-up appointment with complete resolution of her symptoms as well as fully healed ulcers (Figure 2).



Figure 1: Irregular ulcers with violaceous border and necrotic base over the dorsum of the right foot, surrounded by multiple porcelain white atrophic scars.



Figure 2: Fully healed ulcers after two months of rivaroxaban administration.

Discussion

Livedoid vasculopathy is a thrombotic cutaneous disorder manifests as persistent painful skin ulcers owing to occlusion of the superficial microcapillaries of skin, resulting in residual stellate white scar known as atrophic blanche [1]. Livedoid vasculopathy may occur secondary to thrombophilia that occludes the skin microcirculation; however, a large number of cases could be of unknown etiology [1,8]. Of note, the patients of livedoid vasculopathy reported poorer quality of life in comparison to other serious conditions such as psoriasis, tumours, and diabetes [9]. Atrophic blanche are irreversible scars and are not condition-specific lesions. In addition, there is a lack of standardized criteria of diagnosis and different vascular disorders may be involved in the differential diagnosis of livedoid vasculopathy, including antiphospholipid antibody syndrome and venous stasis. Thus, a histopathological evaluation may assist in determining the diagnosis. The pathologists may detect intravascular thrombosis, small vessels tissue degeneration, and lymphocytes infiltration [10].

Livedoid vasculopathy may occur in patients with or without coagulation disorders [8]. Hence, anticoagulants may show salutary outcomes in specific cases. For instance, low-molecular-weight heparin (LMWH) and warfarin had satisfactory effects in a large number of cases [11,12]. However, livedoid vasculopathy is a chronic condition that necessitates repeated LMWH injections and frequent monitoring of international normalized ratio (INR), which interfere with the patients' compliance. Furthermore, fibrinolytic agents are of limited preference owing to a high risk of haemorrhage [13]. Accordingly, recent experimental and human studies have been investigating the promising role of rivaroxaban in the management of livedoid vasculopathy. Rivaroxaban is an orally administered drug that can directly inhibit factor Xa inhibitor [14]. Recent studies have revealed that rivaroxaban can alleviate resistant pain and cutaneous ischemia among livedoid vasculopathy cases. RILIVA was the first trial to examine rivaroxaban efficacy and safety in patients with livedoid vasculopathy [15]. This trial deduced that rivaroxaban can efficiently ameliorate disease symptoms including pain, ulceration, and erythema as well as enhancing the quality of life. Furthermore, the patients receiving rivaroxaban reported well compliance since the drug does not involve frequent bothersome injections like LMWH or regular assessment of INR like warfarin [16]. The patient in our report experienced complete resolution of symptoms and full healing of ulcers after two months of rivaroxaban administration. However, these findings must be cautiously interpreted and thus future large-scale randomized trials are warranted to assist in establishing standard management protocols for this rare disorder. In addition, it is recommended to identify the appropriate age of administration as well as drug dosage and duration through subgroup analysis.

Conclusion

The current report describes a female patient, with no identifiable coagulation disorder, diagnosed with livedoid vasculopathy, for which she received rivaroxaban. Interestingly, rivaroxaban showed effective role in disease management and all symptoms were alleviated on a two-month follow-up. Nonetheless, there is a lack of high-quality evidence regarding drug efficacy and tolerability.

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