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Case Report

Atypical necrobiosis lipoidica presenting outside the pretibial region

Eman Almukhadeb[1], Nawaf Alqahtani[2]*, Ayah Sayed[3], Khlood Alattas[4], Amany Fathaddin[5]

[1]Department of Dermatology, College of Medicine, King Saud University, Riyadh, Saudi Arabia.
[2]College of Medicine, King Saud bin Abdulaziz University for Health Sciences, Riyadh, Saudi Arabia
[3]College of Medicine, King Saud University, Riyadh, Saudi Arabia
[4]College of Medicine, Jazan University, Jazan, Saudi Arabia
[5]Department of Pathology, College of medicine, King Saud University, Riyadh, KSA

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***Correspondence:**
Dr. Nawaf Alqahtani,
E-mail: alqahtanin148@gmail.com

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ABSTRACT

Necrobiosis lipoidica (NL) is a rare chronic granulomatous dermatosis classically associated with diabetes mellitus and typically affecting the pretibial surfaces. Extrapretibial involvement, particularly on the forearm, is uncommon and may create diagnostic difficulty because lesions outside the lower legs can lack the classic clinical distribution of NL. We report a 24-year-old woman with poorly controlled type 1 diabetes mellitus who presented with a slowly enlarging, occasionally pruritic plaque on the left dorsal forearm for approximately 10 years. Skin examination revealed a solitary well-defined atrophic shiny telangiectatic plaque with a central yellow hue and peripheral erythema over the extensor surface of the left forearm. Histopathological examination demonstrated necrobiotic granuloma with foci of dermal necrobiosis surrounded by lymphohistiocytic infiltrate and scattered plasma cells, confirming the diagnosis of necrobiosis lipoidica. The patient was treated with intralesional corticosteroid injections, with further therapy planned according to clinical response.Necrobiosis lipoidica can rarely present at extratibial sites, including the dorsal forearm. Recognition of this atypical presentation is important, as clinical diagnosis may be delayed when lesions occur outside the classic pretibial location. Skin biopsy remains essential for confirming the diagnosis and guiding appropriate management.

Keywords: Necrobiosis lipoidica, Extratibial involvement, Diabetes mellitus type 1

INTRODUCTION

Necrobiosis lipoidica (NL) is a rare, chronic, idiopathic granulomatous dermatosis strongly associated with diabetes mellitus, affecting women predominantly with a female to male ratio of approximately 3:1, and occurring in non-diabetic individuals in a significant minority of cases.[1] Clinically, it presents as waxy, atrophic plaques with an elevated violaceous rim arising from small reddish-brown papules that gradually coalesce into indurated lesions, typically on the pretibial surface, and may ulcerate in up to 15% of cases.[2,3] Histopathologically, NL is defined by a tiered pattern of dermal necrobiosis with palisading granulomas, lymphoplasmacytic perivascular infiltrate, and myointimal thickening of dermal vessels, extending through the full dermis into the subcutaneous fat.[1,3] The pathogenesis remains incompletely understood, with proposed mechanisms including diabetic microangiopathy, immune-complex deposition, and abnormal collagen synthesis.[3,4] While the legs account for

85% of cases, extratibial involvement is distinctly uncommon.[2,5] We present an unusual case of NL arising on the left dorsal forearm, further expanding the known anatomical spectrum of this condition.

CASE REPORT

A 24-year-old female known to have type 1 diabetes mellitus diagnosed at the age of 7 years, currently on insulin aspart and insulin glargine (Lantus), presented with skin lesion over left forearm for approximately 10 years and was associated with occasional pruritus. It had been slowly enlarged over time. She denied similar lesions elsewhere on the body, as well as musculoskeletal complaints, hair loss, or visual changes. The patient also had a history of poorly controlled type 1 diabetes mellitus with multiple prior admissions for diabetic ketoacidosis (DKA), including an ICU admission four years ago.

Skin examination revealed a single well-defined, 3x3 cm, atrophic shiny telangiectatic plaque with central yellow hue and peripheral erythema over the extensor surface of left forearm (Figure 1).

A skin biopsy performed in 2021 confirmed the diagnosis of necrobiosis lipoidica. Microscopic examination of the hematoxylin, and eosin (H & E) stained tissue sections from the skin punch biopsy from the low power revealed an unremarkable epidermis. The dermis demonstrates diffuse involvement by a layered inflammatory process and alternating zones of necrobiotic collagen. (Figure 2A). On high power, the areas of necrobiosis show degenerate eosinophilic collagen without associated dermal mucin. The inflammatory infiltrate is composed of an admixture of lymphocytes, histiocytes and plasma cells. (Figure 2B)



Figure 1: A solitary well-defined atrophic shiny telangiectatic plaque measuring approximately 3-3 cm in diameter with a central yellow hue and peripheral erythema over the extensor surface of the left forearm.

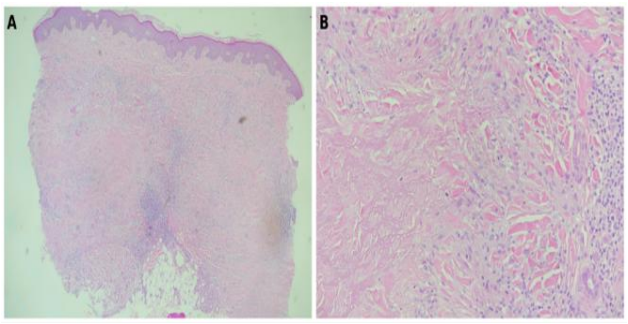


Figure 2: Skin biopsy: (A) The low power shows diffuse involvement by a layered inflammatory process and alternating zones of necrobiotic collagen (H&E, x40). (B) High power reveals degenerate eosinophilic collagen with inflammatory infiltrate (H&E, x 200).

Treatment

The patient was started on intralesional kenacort injections (ILK) which led to slow progression of the lesion in the beginning. Further intralesional corticosteroid therapy is planned depending on clinical response. At follow up, the skin lesion showed clinical improvement after multiple sessions of intralesional corticosteroid and a course of topical corticosteroids. Patient was counseled on the importance of achieving and maintaining good glycemic control, as poorly controlled blood glucose may exacerbate her skin condition. The current topical treatment will be continued as maintenance therapy, with further follow up planned to assess the sustained treatment response and monitor disease progression.

DISCUSSION

NL is a well-characterized granulomatous dermatosis with a strong predilection for the pretibial surfaces; however, a subset of cases arises at extratibial locations including the forearms, dorsal hands, trunk, back, nipple, face, scalp, and genitalia posing considerable diagnostic challenges due to the frequent absence of classical features.[1-5] Tee et al documented forearm NL in which biopsy not only confirmed palisading necrobiotic granulomas but also revealed concurrent elastophagocytosis a histological feature not previously associated with NL illustrating that morphological and histopathological atypicality may coexist when disease arises outside its classic distribution.[1] Truncal NL has similarly been reported, where the lack of characteristic yellowish discoloration and telangiectasia renders clinical recognition difficult, and dermoscopy showing linear branching vessels against a yellow-white structureless background has been proposed as a useful pre-biopsy adjunct.[2,5] Kavanagh et al described a patient with biopsy-proven NL confined entirely to the back and nipple with no lower extremity involvement, fundamentally challenging the assumption that leg sparing excludes the diagnosis.[3] NL of the face and scalp

has also been reported, where the differential includes scarring alopecia, discoid lupus erythematosus, and cutaneous sarcoidosis, making histopathological examination the sole reliable diagnostic tool.[4] Further rare localizations include penile NL and lesions arising within surgical scars via a Koebner isomorphic mechanism.[1-5] The dorsal forearm, as in the present case, represents one of the least frequently reported extratibial sites, where inherently low clinical suspicion makes early biopsy essential histopathological demonstration of tiered dermal necrobiosis with palisading granulomas and myointimal vascular thickening remains the diagnostic gold standard regardless of location.[1,3,4]

Limitations

As a single case report, this study cannot support generalizable conclusions regarding the frequency, natural history, or optimal management of dorsal forearm NL. The paucity of analogous published cases limits meaningful literature-based comparison.[1-5] Prospective registries or multicenter studies are needed to better define the epidemiology and clinical behavior of extratibial NL.

CONCLUSION

The case report of NL arising on the left dorsal forearm an anatomical site rarely implicated in this condition and one that deviates markedly from its classical pretibial distribution. This case reinforces that NL should be included in the differential diagnosis of indurated atrophic plaques at any cutaneous site, including the upper extremities. Skin biopsy with histopathological evaluation is indispensable for confirming the diagnosis at atypical locations, where clinical recognition alone is insufficient. Heightened clinician awareness of extratibial NL is essential to prevent diagnostic delay and to ensure timely, appropriate management.

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