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Case Report

# Neutrophilic Urticarial Dermatitis Revealing Lupus with Lupus Nephropathy: a Case Report.

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## Abstract

Neutrophilic urticarial dermatosis is a rarely discovered entity manifesting as a maculopapular, barely pruritic or non-pruritic, fleeting erythema with a specific histological appearance showing a perivascular infiltrate of unaltered neutrophils without associated vasculitis. This entity is associated with certain systemic diseases such as Still's disease, Gougerot-Sjögren syndrome and Schnitzler syndrome. We report the case of a patient who presented with neutrophilic urticarial dermatosis (NUD) associated with systemic lupus erythematosus (SLE) with extracutaneous renal involvement.

**Keywords:** Neutrophilic urticarial dermatosis; lupus nephropathy

## Introduction

Neutrophilic urticarial dermatosis (NUD) is an entity that belongs to the group of neutrophilic dermatoses (ND). It is characterized histologically by an intense inflammatory dermal infiltrate composed of neutrophils without vasculitis [1].

Neutrophilic urticarial dermatosis (NUD) is a recently described entity that often occurs in the context of systemic disease and may be complicated by extracutaneous involvement [2].

We report the case of a patient who presented with neutrophilic urticarial dermatosis (NUD) associated with systemic lupus erythematosus (SLE) with extracutaneous renal involvement.

## Case report

A 23-year-old woman without particular medical history, presented with multiple reddish, infiltrated plaques in the upper limbs. The lesions were non pruriginous. They were associated with photosensitivity, erythema in vespertilio and joint pain. Skin biopsy showed an interstitial dermal neutrophilic infiltrate without vasculitis. Direct immunofluorescence tests were negative and revealed a lupus band. The following criteria: skin with urticaria and neutrophilic infiltrate without vasculitis on histology, photosensitivity and erythema in vespertilio, In the presence of the following criteria: renal involvement such as proteinuria, hematuria without alteration of the renal function with histological class IV lupus nephropathy at the renal biopsy, hematological involvement such as lymphopenia, skin with urticaria and neutrophilic infiltrate without vasculitis at the histology, erythema in vespertilio and photosensitivity. Immunological criteria with positive anti-nuclear antibodies of type 1/1280 anti-native DNA and consumption of C3 and C4 complements the diagnosis of DUN associated with SLE was retained. The treatment was essentially based on high dose corticosteroids with mycophetil mofetil associated with hydroxychloroquine. The evolution was marked by the disappearance of NUD lesions upon initiation of corticosteroids.

## Discussion

The association LES and NUD is rare. Only few cases were reported in the literature [1]. On 2009, Kieffer et al [2], was the first who defined, the NUD as a sort of urticarial eruption which is pale, flat or only slightly raised, nonpruritic macules, papules, or plaques, which disappear within hours without any sequale. It's histologically different from urticaria with an important interstitial neu-

trophilic infiltration. This infiltration is concentrated along the collagen bundles and in the deep part of the reticular dermis [3]. The infiltrate is composed almost exclusively from neutrophils [4] with leukocytoclasia but without vasculitis. Kieffer et al [4] reported the cases of nine patients who presented a NUD and 3 of whom, this NUD was associated to SLE. Pavlidakey et al [5], cited the cases of 6patients with NUD revealing a SLE. Hau et al [6], NUD was the presenting mode in 3patients conducting to the diagnosis of SLE. On 2014, Larson and Granter [7], reviewed neutrophilic dermatitis encountered in the SLE and NUD associated to SLE was described within 3patients. The pathogenesis of this neutrophilic dermatitis is due to a disorder of the innate immunity which probably results in autoinflammation as it is associated to systemic diseases [8]. The therapeutic strategies of NUD target neutrophils by modulating their activation, maturation, or migration. Oral corticosteroids has been proven to be effective. Other drugs were beneficial as Sulphones (dapsone), colchicine, potassium iodide, retinoid (acitretin), clofazimine, sulfasalazine, and thalidomide. The immunosuppressant agents, such as cyclosporine, cyclophosphamide and chlorambucil were effective in corticosteroid-unresponsive cases. In summary, NUD seemed to be an indicator of SLE activity or a prognostic factor as all cases with NUD associated with a renal involvement. In our case, the DUN was the first symptom leading to the diagnosis of a SLE associated to a renal involvement. The corticosteroid's administration was effective.

## Conclusion

NUD remains an unusual manifestation of SLE. Different clinical courses and relationships with SLE suggest that NUD and SLND have different pathogeneses for neutrophilic inflammatory reactions. It should be recognized, especially in young people, to avoid misunderstanding about the diagnosis of SLE and timely treatment.

## Conflict of interest

None declared.

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