

Lupus Erythematosus and Neutrophilic Urticarial Dermatitis

A Retrospective Study of 7 Patients

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Abstract: Neutrophilic urticarial dermatosis (NUD) resembles urticaria clinically but is a neutrophilic dermatosis histopathologically. The majority of patients with NUD have an underlying systemic condition, mainly, autoinflammatory disorders such as cryopyrin-associated periodic syndromes, Schnitzler syndrome, and adult-onset Still disease, but a few also have systemic lupus erythematosus (LE). Here, we confirm these data and we report relevant clinical and histopathological data of 7 patients with LE and NUD.

We retrospectively retrieved the medical records of all patients with LE in whom skin biopsy showed NUD in registers of Strasbourg and Montpellier University hospitals since 2000.

All were female and aged between 13 and 45 years. Skin lesions were typically rose or red macules or slightly elevated papules occurring in a wide distribution. Individual lesions resolved within 24 hours and were not or only slightly itchy. Every patient had associated signs, most of the time polyarthritis and/or fever. NUD was the presenting mode of LE in 2 patients. NUD was misdiagnosed as a classic lupus flare and led to therapeutic intensification with the introduction of immunosuppressive drugs in 4 patients. Histopathological findings consisted of intense neutrophilic interstitial and perivascular infiltrate with leukocytoclasia and without fibrinoid necrosis of vessel walls. Direct immunofluorescence testing showed a lupus band in 4 patients. Antinuclear antibodies were always positive, anti-dsDNA antibodies were positive in 5 patients, and anti-Ro/SSA antibodies in 6 patients. Immunosuppressive drugs such as prednisone, hydroxychloroquine, mycophenolate mofetil, and methotrexate were never effective to treat NUD. Antihistamines were effective in 1 patient and dapsone or colchicine was effective in 5 patients.

NUD is not exceptional in patients with systemic LE and is easily misdiagnosed as an acute LE flare. Furthermore, we show that conventional immunosuppressive LE treatments are not efficient and we underline the major interest of dapsone and colchicine, classic neutrophil migration inhibitors, in those patients.

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Abbreviations: ACR = American College of Rheumatology, DIF = direct immunofluorescence, dsDNA = double-stranded DNA, LE = lupus erythematosus, ND = neutrophilic dermatosis, NUD = neutrophilic urticarial dermatosis, RNP = ribonucleoprotein, SLE = systemic lupus erythematosus, SLICC = Systemic Lupus International Collaborating Clinics, Sm = Smith.

INTRODUCTION

Neutrophilic urticarial dermatosis (NUD), the most recently delineated entity within the nosologic spectrum of the neutrophilic dermatoses (NDs), was first described in 2009 by Kieffer et al¹ as an eruption consisting of rose or red macules or slightly elevated plaques vanishing within 24 hours. The histopathologic findings are a dense perivascular and interstitial infiltrate of neutrophils with leukocytoclasia but without vasculitis. In this initial study, 9 patients were reported and 7 had associated systemic diseases: adult-onset Still disease (3 patients), systemic lupus erythematosus (SLE) (3 patients), and Schnitzler syndrome (1 patient). It was not surprising to find patients with adult-onset Still disease and Schnitzler syndrome, entities that are considered as acquired autoinflammatory disorders with neutrophilic tissue infiltration, but the presence of patients with a connective disease such as lupus erythematosus (LE) was unexpected. This led us to review in detail the association between ND and LE.^{1,2}

Several types of NDs have already been reported in patients with LE, such as pyoderma gangrenosum, Sweet syndrome, palisaded neutrophilic granulomatous dermatitis, amicrobial pustulosis of the folds, and recently NUD.³ Furthermore, bullous LE is a ND. The presence of neutrophilic infiltrate in early and evolving lesions of cutaneous LE is a well-known phenomenon⁴; therefore, including neutrophilic lesions in the classification of skin lesions in SLE has already been suggested.⁵

The great majority of patients with NUD have fever and joint pain. Therefore, the symptomatic set of rash, fever, and joint pain in a patient with known SLE is often mistaken for an exacerbation of LE leading to therapeutic intensification with immunosuppressors. However, the latter do usually not alleviate symptoms leading to an increase in immunosuppression, while dapsone and colchicine, classic neutrophil migration inhibitors, are generally effective to control NUD. This highlights the importance of correctly identifying this entity in lupus patients.

Here, we report 7 patients with NUD and SLE, and we have paid particular attention to the treatments undertaken and their effects.

PATIENTS AND METHODS

We performed a retrospective study and retrieved the medical records of all patients with LE whose skin biopsy showed NUD in registers of Strasbourg and Montpellier

University Hospitals (France) since 2000. Under French law, this type of study, which does not involve any invasive investigation but relies on a retrospective analysis of patient files, does not need the approval of the institutional review board.

Patients were included if they met the following criteria:

1. Diagnosis of NUD defined as recurrent or chronic cutaneous eruption consisting of macules, papules, or plaques resolving within 48 hours, pruritic or not, and histopathologic findings consisting of a diffuse neutrophilic infiltrate in the dermis with interstitial involvement with leukocytoclasia but without fibrinoid necrosis of vessel walls and without significant dermal edema.
2. Diagnosis of SLE according to American College of Rheumatology (ACR) and/or Systemic Lupus International Collaborating Clinics (SLICC) criteria or cutaneous LE based on classic clinical criteria and/or histological ascertainment of LE.

For all patients, relevant clinical data including age, gender, duration, distribution and morphology of skin lesions, history of LE, serologic data, medications at the time of diagnosis, and response to treatment were reviewed. Three patients were already described in the study by Kieffer et al¹ (patients 5–7).

RESULTS

A total of 7 patients fulfilled the criteria. Their characteristics are summarized in Table 1. A brief summary of 2 typical cases follows to illustrate the difficulty of the diagnosis and how it can be misdiagnosed as a classic lupus flare.

Patient 3

A 21-year-old female was admitted for a rash (Figure 1). She had a medical history of SLE for 4 years with glomerulonephritis type III, pancytopenia, photosensitivity, polyarthritides, and cutaneous vasculitis. Antinuclear factors were positive with anti-dsDNA, anti-Ro/SSA, anti-ribonucleoprotein (RNP), and antiribosomal antibodies. She also had low complement: C3 = 0.23 g/L ($0.75 < N < 1.40$) and C4 = 0.02 g/L ($0.10 < N < 0.34$). She had no treatment for 1 year when she developed the rash. Lesions consisted of slightly erythematous migratory annular nonitching macules and papules on the face, the trunk, and the hands (Figure 1). She also complained of joint pain and chest pain. She reported similar episodes for 2 years. Laboratory tests showed an elevated erythrocyte sedimentation rate (100 mm/h) and polyclonal hypergammaglobulinemia (15.6 g/L). She was first treated with 0.75 mg/kg of prednisone without any improvement. The treatment was strengthened successively by hydroxychloroquine, antihistaminic, and mycophenolate mofetil. Finally, a dermatological advice was required. A skin biopsy was performed that showed an interstitial dermal neutrophilic infiltrate and leukocytoclasia. Direct immunofluorescence (DIF) revealed a lupus band. A diagnosis of NUD was made. Therefore, she was first treated with colchicine without improvement and then dapsone was introduced; as a result, skin lesions disappeared within 48 hours.

Patient 6

A 22-year-old female was referred for a rash. She suffered from SLE for 2 years, with polyarthritides, pericarditis, and hemolytic anemia treated with prednisone and hydroxychloroquine.

Antinuclear factors were positive with anti-Smith, anti-RNP, and anti-Ro/SSA antibodies. Physical examination revealed slightly erythematous papules and macules of the trunk and the upper limbs. The lesions were nonpruriginous. They were associated with joint pain. She reported similar episodes twice before. The skin biopsy showed an interstitial dermal neutrophilic infiltrate with leukocytoclasia and altered collagen bundles. Direct immunofluorescence testing was negative. She was successfully treated with colchicine. The skin lesions recurred 1 year later, a few days after colchicine was stopped.

NUD Features

Clinical Findings

All patients were female aged between 13 and 45 years. The skin lesions were typically rose or red macules or slightly elevated papules occurring in a wide distribution. The lesions were annular in 1 patient. The face and the extremities were involved in 1 patient. The trunk was involved in 6 patients, upper limbs in 5 patients, and lower limbs or buttocks in 2 patients. The lesions were nonitching in 6 patients and slightly itching in 1 patient. One patient had a Koebner phenomenon. The skin lesions were never photodistributed. Associated signs were joint pain (5 patients), fever (1 patient), abdominal pain (2 patients), chest pain (1 patient), episcleritis (1 patient), vesperilio rash (3 patients), pharyngeal pain (2 patients), and paresthesia of fingers (2 patients). NUD was the first manifestation of LE in 2 cases (patients 1 and 4). The time of evolution of NUD before diagnosis ranged from 1 to 12 years. For patient 5, it occurred in the context of nephrotic syndrome with kidney failure with membranous lupus nephritis (type V according to the International Society of Nephrology/Renal Pathology Society 2003 classification of lupus nephritis).

Histopathological Findings

We observed in all patients an intense neutrophilic interstitial and perivascular infiltrate associated with leukocytoclasia but without significant edema or fibrinoid necrosis of vessel walls. Isolated necrobiotic collagen bundles were found in 3 patients and mild vacuolar change of the basal layer was observed in 1 patient (patient 4). DIF testing performed in 6 patients detected a lupus band in 4 patients and was negative in 2 patients.

Biological Findings

Polyclonal hypergammaglobulinemia was observed in 5 patients. Erythrocytes sedimentation rate was elevated in 3 patients. C-reactive protein was elevated in 3 patients (mean value = 36 mg/L [extremes, 26–44]).

LE Features

Antinuclear antibodies were always positive; anti-dsDNA antibodies were present in 5 patients, and anti-Ro/SSA antibodies in 6 patients. The complement was low in 4 patients and noninterpretable in 1 patient because of a congenital C4 deficiency. Treatments at the time of diagnosis for the 5 patients with a known LE were hydroxychloroquine (but with poor observance in 1 patient), prednisone, and mycophenolate mofetil for 2 patients, and hydroxychloroquine and prednisone for 1 patient. Six patients had SLE according to the SLICC and ACR criteria. Three patients had kidney involvement.

TABLE 1. Summary of the 7 Patients Included in the Study

Patient No. Sex/Age	Cutaneous and Other Clinical Signs	Laboratory Findings	Autoimmunity	Histology/DIF	Prior History of LE Years/Organs Involved	Treatments for LE at Diagnosis	Treatments for NUD (Results)
1. F/45	Erythematous macules or papules, slightly itching on trunk and limbs; paresthesia of fingers; inguinal adenopathy	Polyclonal hypergammaglobulinemia	Positive ANA (1:1600); low C3 and C4; anti-Ro/SSA antibodies and anti-La/SSB antibodies	Interstitial dermal neutrophilic infiltrate; leukocytoclasia; altered collagen bundles/DIF: lupus band	None	None	Antihistamines (CR)
2. F/40	Erythematous nonitching macules and papules on trunk and limbs; joint pain; paresthesia of fingers; pharyngeal pain; asthenia; myalgia	ESR = 53 mm/h; CRP = 44 mg/L normocytic normochromic anemia; elevated alpha2globulinemia	Positive ANA (1:1280); anti-dsDNA antibodies; anti-Ro/SSA antibodies and anti-La/SSB antibodies; complement nonavailable	Interstitial neutrophilic infiltrate; leukocytoclasia/DIF: negative	12 y/arthritis; photosensitivity; Raynaud disease; livedo; xerostomia	Hydroxychloroquine	Hydroxychloroquine + prednisone (NC) hydroxychloroquine + prednisone + methotrexate (NC)/ hydroxychloroquine + prednisone + methotrexate + dapsone (CR) Prednisone (NC)/ hydroxychloroquine (NC)/ hydroxychloroquine + antihistamines (NC) prednisone + mycophenolate mofetil + hydroxychloroquine (NC)/prednisone + mycophenolate mofetil + hydroxychloroquine + colchicine (NC)/ prednisone + mycophenolate mofetil + hydroxychloroquine + dapsone (CR)
3. F/21	Slightly erythematous migratory annular nonitching macules and papules on face, trunk, hands nonitching; joint pain; chest pain	Panycytopenia; ESR = 44 mm/h; polyclonal hypergammaglobulinemia	Positive ANA (1:2500); anti-dsDNA antibodies; anti-Ro/SSA antibodies; anti-Sm antibodies; anti-RNP antibodies; anti-ribosomal antibodies; anti-CCP antibodies; low C3 and C4	Interstitial dermal neutrophilic infiltrate; leukocytoclasia/DIF: lupus band	4 y/glomerulonephritis type III; pancytopenia; photosensitivity; polyarthritis; cutaneous vasculitis	None	Hydroxychloroquine (NC)/ hydroxychloroquine + antihistamines (NC) prednisone + mycophenolate mofetil + hydroxychloroquine (NC)/prednisone + mycophenolate mofetil + hydroxychloroquine + colchicine (NC)/ prednisone + mycophenolate mofetil + hydroxychloroquine + dapsone (CR)
4. F/20	Erythematous migratory papules and macules nonitching on the trunk; joint pain; diarrhea and abdominal pain; episcleritis	Polyclonal hypergammaglobulinemia; leukopenia; hemolytic anemia; CRP = 26 mg/L; congenital partial C4 deficit	Positive ANA (1:2048); anti-dsDNA antibodies; anti-cardiolipin antibodies; anti-Ro/SSA antibodies and anti-La/SSB antibodies; complement nonavailable	Interstitial dermal neutrophilic infiltrate; leukocytoclasia; interface changes/DIF: lupus band	None/digestive involvement; polyarthritis; skin; hematologic	None	Hydroxychloroquine (NC)/prednisone + azathioprine + hydroxychloroquine (NC)/prednisone + mycophenolate mofetil + hydroxychloroquine (NC)/prednisone + mycophenolate mofetil + hydroxychloroquine + dapsone (PR)
5. F/22	Few episodes of slightly erythematous wandering macules and papules nonitching on trunk and upper limbs; palpebral and lower limbs edema (nephrotic syndrome); vesperitilo rash	Hypoprotidemia and hypoalbuminemia	Positive ANA (1:320); anti-dsDNA antibodies; low C3 and C4	Interstitial dermal neutrophilic infiltrate; leukocytoclasia/DIF: lupus band	11 y/membranoproliferative glomerulonephritis type IV; cutaneous vasculitis	Mycophenolate mofetil + prednisone	0 (unknown)

TABLE 1. (Continued)

Patient No. Sex/Age	Cutaneous and Other Clinical Signs	Laboratory Findings	Autoimmunity	Histology/DIF	Prior History of LE Years/Organs Involvement	Treatments for LE at Diagnosis	Treatments for NUD (Results)
6. F/22	3 episodes of transient nonitching rose macules and papules on trunk and upper limb; joint pain	Hemolytic anemia	Positive ANA (1:1280); anti-Sm antibodies; anti-RNP antibodies; anti-Ro/SSA antibodies; low C3 and C4	Interstitial dermal neutrophilic infiltrate; leukocytoclasia; altered collagen bundles/DIF negative	2 y/hemolytic anemia; polyarthritides; pericarditis	Hydroxychloroquine + prednisone	Azathioprine (NC)/colchicine (CR)
7. F/13	2 episodes of nonitching migratory pink papules on buttocks and limbs; vesperitillo rash; fever; joint pain; pharyngeal and abdominal pain	ESR = 69 mm/h; CRP = 40 mg/L; inflammatory anemia and thrombocytopenia; polyclonal hypergammaglobulinemia	Positive ANA (1:1280); anti-dsDNA; anti-Ro/SSA; antibodies anticardiolipin antibodies; anti-beta2GPI antibodies; type 2 cryoglobulinemia; anti-Sm antibodies; anti-RNP antibodies complement non available	Interstitial dermal neutrophilic infiltrate; leukocytoclasia; altered collagen bundles/no DIF	1 y/hemolytic anemia; hematuria; proteinuria; polyarthritides; Raynaud disease	Mycophenolate mofetil + prednisone	Colchicine (CR)

ANA = antinuclear antibodies, CCP = cyclic citrullinated peptide, CR = complete remission, CRP = C-reactive protein, DIF = direct immunofluorescence, dsDNA = double-stranded DNA, ESR = erythrocyte sedimentation rate, F = female, LE = lupus erythematosus, NC = no change, NUD = neutrophilic urticarial dermatosis, PR = partial remission, RNP = ribonucleoprotein, Sm = Smith.



FIGURE 1. Red pale macules of the arm (patient 3).

Treatment

The symptomatology of NUD lead to a therapeutic intensification in 4 patients with the introduction of immunosuppressive drugs. Those treatments including prednisone, methotrexate, hydroxychloroquine, mycophenolate mofetil, and azathioprine were always ineffective on skin lesions but could be effective on joint pain (patient 2: methotrexate; patient 4: azathioprine and mycophenolate mofetil). Antihistamines were effective in 1 patient with a delay of 3 weeks. Dapsone or colchicine was effective in 5 patients with no major side effect.

DISCUSSION

We report 7 patients with NUD and SLE, showing that this association is not rare. NUD is characterized by rash, joint pain, and fever and thus was often initially misdiagnosed as a classic lupus flare, leading to introduction of immunosuppressive drugs, a measure that was potentially harmful and inefficient. However, when correct diagnosis was established and treatment with dapsone or colchicine was introduced, control of not only skin lesions but also associated signs was obtained in the majority of cases. Thus, recognition of NUD is important and relevant in clinical practice. Based on the lupus patients followed-up in our institutions, it can be estimated that a little <5% of lupus patients seen at a tertiary dermatology referral center have NUD.

NDs in the setting of LE are frequently encountered. As illustrated in Table 2 and Appendix Table 2, <http://links.lww.com/MD/A125>, there are already many clinical observations reported in the literature, mainly as case reports: Sweet syndrome (N=20), pyoderma gangrenosum (N=26), palisaded neutrophilic granulomatous dermatitis (N=17), amicrobial pustulosis of the folds (N=22), and bullous LE (N > 85). More recently, Larson and Granter³ reviewed NDs associated with LE including Sweet-like syndrome, nonbullous neutrophilic LE, NUD, and autoimmunity-related ND: 38 patients were reported under these names including the 3 patients of the Kieffer et al¹ study. We believe all those observations belong to the same nosologic spectrum, delineating a subset of lupus patients to

TABLE 2. Neutrophilic Dermatitis and Lupus Erythematosus

Entity	Clinical Features	Histopathology	Number of Cases of ND With LE
Sweet syndrome	Erythematous to violaceous tender papules or nodules that often coalesce to form irregular plaques Fever/general malaise/joint pain/episcleritis/aphthous stomatitis	Dense infiltrate of mature neutrophils in the upper half of the dermis Neutrophils may extend throughout the dermis and even into the subcutis Marked edema of the papillary dermis Leukocytoclasia but no vasculitis or fibrinoid extravasation	20
Pyoderma gangrenosum	Rapid progression of painful, necrolytic ulceration with an irregular, undermined, violaceous border—usually with a preceding papule, pustule, or bulla, and pain out of proportion to the size of the ulcerated area	Histopathology depends on variant (ulcerative, pustular, bullous, vegetative, superficial granulomatous, . . .) Early lesion: folliculitis Late lesion: necrosis of epidermis and superficial dermis and ulcer with base showing mixed inflammatory cells with abscess formation Advancing edge of lesion: perivascular infiltrate of lymphocytes and plasma cells with endothelial swelling, fibrinoid extravasation, and numerous neutrophils	26
Palisaded neutrophilic granulomatous dermatitis	Erythematous urticated plaques or papules; firm, violaceous, or waxy plaques; skin-colored or erythematous papular or nodular lesions, sometimes annular and sometimes with ulceration or crusting; tender subcutaneous nodules; and firm linear subcutaneous cords or bands	Interstitial (and variably perivascular) granuloma annulare-like or necrotizing extravascular granulomatous process with basophilic degenerate collagen; intense tissue neutrophilia, leukocytoclasia, small vessel leukocytoclastic vasculitis (in about 50%) and sometimes an admixture with eosinophils	17
Amicrobial pustulosis of the folds	Pustulosis involving ≥1 major folds, ≥1 minor folds, and the anogenital area	Intraepidermal spongiform pustules and a mainly neutrophilic dermal infiltrate	22
Bullous LE	Bullous lesions predominantly on the face, neck, and upper trunk, but may be more widespread. May heal with milia formation. One-third have mouth lesions. Photosensitivity may occur	Subepidermal “smooth-edged” splitting; papillary microabscesses; nuclear dust prominent in papillae and superficial vessels; neutrophils can extend deep in dermis	>85
Neutrophilic urticarial dermatosis	Rose or red macules or slightly elevated plaques vanishing within 24 h Arthralgia/fever	Perivascular and interstitial infiltrate of neutrophils with leukocytoclasia but without vasculitis	42

LE = lupus erythematosus, ND = neutrophilic dermatosis.

which we previously referred as neutrophilic cutaneous LE.⁶ Those patients have exaggerated innate immune response and it is yet not known if this portends prognostic value for the SLE per se (see below). But the neutrophilic part of the disease needs a specific management, with drugs usually not prescribed for similar symptoms—rash and joint pain—occurring in patients with SLE. Thus, all of those observations exhibit the frequency of ND in the setting of LE and support the idea of including them in a classification of skin involvement of LE.⁵ As already stated, they are important to recognize because they can be the first manifestation of LE and their management is very peculiar.

The rash observed in patients with NUD is very characteristic and should be suspected in patients with pale red macules or only slightly raised nonitching papules vanishing within 24 hours. In this particular case, a dermatological look as well as skin biopsy is crucial, which shows the typical neutrophilic dermal infiltrate. This is all the more important since effective nonimmunosuppressive treatment exists, namely, dapsone or colchicine. Indeed, dapsone, a classic neutrophil migration inhibitor is often used in dermatology to treat other NDs such as dermatitis herpetiformis or pyoderma gangrenosum.⁷ It has already been reported effective in other LE-associated ND such as bullous LE⁸ or amicrobial pustulosis of the folds.⁹ Therapeutic dosages of dapsone range from 50 to 200 mg/d. The lowest effective dose should be given to minimize the possible side effects such as methemoglobinemia, hemolytic anemia, or hypersensitivity syndrome (also referred

to as dapsone syndrome¹⁰). Before starting the treatment, glucose-6-phosphate dehydrogenase deficiency should be excluded. Regular controls of blood cell count, methemoglobinemia, and liver functions should be done. By analogy with autoinflammatory syndromes,¹¹ pyoderma gangrenosum,¹² Sweet syndrome,¹³ or leukocytoclastic vasculitis,¹⁴ colchicine had been used to treat NUD and other LE-associated ND with a good efficacy. It had been effective in 2 of the patients reported here. Therapeutic doses for NUD range from 0.5 to 1 mg/d.¹⁵ It may cause nausea, vomiting, diarrhea, and abdominal pain but is most of the time well tolerated.

In our study, dapsone or colchicine was associated with immunosuppressive drugs in 4 patients and colchicine was the only treatment in only 1 patient. Therefore, the real effect of neutrophil migration inhibitors alone is difficult to appreciate. Their use as monotherapy in patients with SLE having NUD needs further study to elucidate their efficacy as sole treatment.

Characteristics of LE in our patients with NUD are heterogeneous. Some patients had only LE-specific skin and/or joint involvement, while others had systemic involvement. Kidney was the most frequently involved organ. It is so far not known if NUD is a prognostic factor or an indicator of systemic disease activity of SLE. In our study, expression of LE varied considerably. In 1 case (patient 5), NUD seemed to be associated with systemic activity leading to renal failure. A kidney biopsy was performed at the time that showed no neutrophil infiltration

but a typical lupus glomerulonephritis type V. NUD was the presenting mode of LE in 2 cases. Patient 1 had no systemic involvement to this day. Patient 4 developed a systemic lupus flare with rash, joint pain, and digestive involvement leading to introduction of prednisone. In the study of Pavlidakey et al,¹⁶ NUD was the presenting mode in 6 of 7 patients with LE.

Interestingly, the anti-Ro/SSA and/or anti-La/SSB antibodies were positive in 6 of our patients and status was unknown in 1. Regarding the literature, anti-Ro/SSA antibodies were positive in 8 patients, negative in 9 patients, and their status was not specified in 18 patients.^{16–20} This raises the question whether anti-Ro/SSA antibodies could be related in anyway with NUD and potentially link innate and acquired immunity.

Thus, it will be a challenge in the future to determine if NUD portends a prognostic importance for the specific manifestations of SLE. Meanwhile, its recognition and appropriate treatment to avoid immunosuppressive overtreatment and relieve patients with treatments targeting neutrophils is essential. The association of NUD and SLE is another example of the expanding spectrum of disorders combining autoinflammatory and autoimmune mechanisms and possibly also immune deficiencies although this was not apparent in this series of patients.

CONCLUSION

We describe 7 more new cases of NUD occurring in the setting of LE, a situation that is not exceptional and easily misdiagnosed as an acute lupus flare. We show that conventional immunosuppressive LE treatments are not efficient, at least not alone, and we underline the major interest of dapsone and colchicine.

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