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# Two case reports of pyoderma gangrenosum and systemic lupus erythematosus

## A rare but nonfortuitous association?

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### Abstract

**Rationale:** Pyoderma gangrenosum (PG), like other neutrophilic dermatosis, may be associated with a variety of systemic disorders including inflammatory bowel diseases, rheumatoid arthritis, and hematologic disorders. Conversely, the association between PG and systemic lupus erythematosus (SLE) has rarely been reported.

**Patient concerns:** We report here 2 cases of this association.

**Diagnoses:** The first case involves a 32-year-old woman who developed, 1 year after SLE diagnosis, 3 painful nodular lesions of PG on her face, and cervical area. The second case was observed in a 37-year-old woman referred for ulcerative nodular papules of PG on her legs, whereas she had been diagnosed with SLE 10 years before. SLE was inactive in the first case, whereas PG occurred during a lupus flare up in the second one.

**Interventions:** We found 23 previous cases of SLE and PG in the literature with most cases (12/20) occurring during a lupus flare.

**Outcomes:** Although rare, this association may be supported by common innate immunity dysregulation and abnormal neutrophil activation.

**Lessons:** PG and other neutrophilic diseases reported in patients with SLE may be added to the large clinical spectrum of cutaneous lesions observed in SLE.

**Abbreviations:** IL = interleukin, NDs = neutrophilic diseases, NET = neutrophil extracellular traps, PG = pyoderma gangrenosum, SGS = Sjogren syndrome, SLE = systemic lupus erythematosus, TNF = tumor necrosis factor.

**Keywords:** NETosis, neutrophilic dermatoses, pyoderma gangrenosum, systemic lupus erythematosus

## 1. Introduction

Neutrophilic diseases (NDs) are a heterogeneous group of conditions characterized by the accumulation of neutrophils in the skin that may also involve many other organs.<sup>[1,2]</sup> The pathophysiology of ND is poorly understood but this group of diseases shares clinical and pathologic aspects with auto-inflammatory syndromes. A clinicopathologic classification has been proposed, subdividing them into deep or hypodermal forms,

whose prototype is pyoderma gangrenosum (PG); plaque type or dermal forms, whose prototype is Sweet syndrome; and superficial or epidermal forms.<sup>[1]</sup> Among this spectrum, PG typically presents with single or multiple skin ulcers with undetermined erythematous-violaceous borders, but different variants have been distinguished: ulcerative PG, pustular PG, bullous PG, superficial vegetative PG, and peristomal PG.

PG like other ND may be associated with a variety of systemic disorders.<sup>[1-3]</sup> In a study on a large series of PG patients, disease associations are reported in approximately one-third of cases, including inflammatory bowel diseases (20.2%), rheumatoid arthritis (11.8%), and hematologic disorders (3.9%).<sup>[3]</sup> PG may precede, coexist, or follow the different systemic diseases. PG can also arise as a consequence of drug therapies and may be observed in autoinflammatory syndromes, especially in the PAPA syndrome (Pyogenic sterile Arthritis, Pyoderma Gangrenosum, Acne) and in the PASH syndrome (Pyoderma Gangrenosum, Acne, Suppurative Hidradenitis).<sup>[1,4]</sup>

Conversely, the association between PG and systemic lupus erythematosus (SLE) has been rarely reported. We report 2 cases of this association. We also discuss the possible pathophysiologic links between the 2 entities and have performed a literature review of previously published cases.

## 2. Case 1

Patient 1 was a 32-year-old woman with type 1 diabetes complicated by retinopathy with occlusion of the left central retinal vein in 2008, Sjogren syndrome since 2011, and SLE since

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Patients have given their informed consent for the case report to be published.

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2014. SLE diagnosis was based on the association of a malar rash, photosensitivity, alopecia, lymphopenia, antinuclear antibodies positivity at a 1/640 dilution (indirect immunofluorescence on Hep2 cells), anti-DNA antibodies identified by Farr assay at 7 IU/mL ( $N < 5$  IU) and low complement levels. No antiphospholipid antibodies were detected. Her SLE was treated with hydroxychloroquine and acetylsalicylic acid without corticosteroids.

One year after SLE diagnosis, 3 painful nodular lesions with surrounding inflammation and erythema developed at the inner canthus of the left eye ( $1 \times 1$  cm), at the left corner of the lips ( $2 \times 2$  cm), and in the cervical area ( $2.5 \times 3$  cm), respectively (Fig. 1A). No clinical signs of lupus activity were present.

Laboratory tests revealed normocytic anemia at 10.2 g/dL, the absence of inflammatory syndrome, normal liver, and kidney function with physiologic proteinuria. Anti-DNA antibodies were measured at 6 IU/mL by the Farr assay ( $N < 5$  IU). Complement was normal.

Histologic examination of the left cervical lesion showed a dense infiltration of dermis by leukocytoclastic or intact

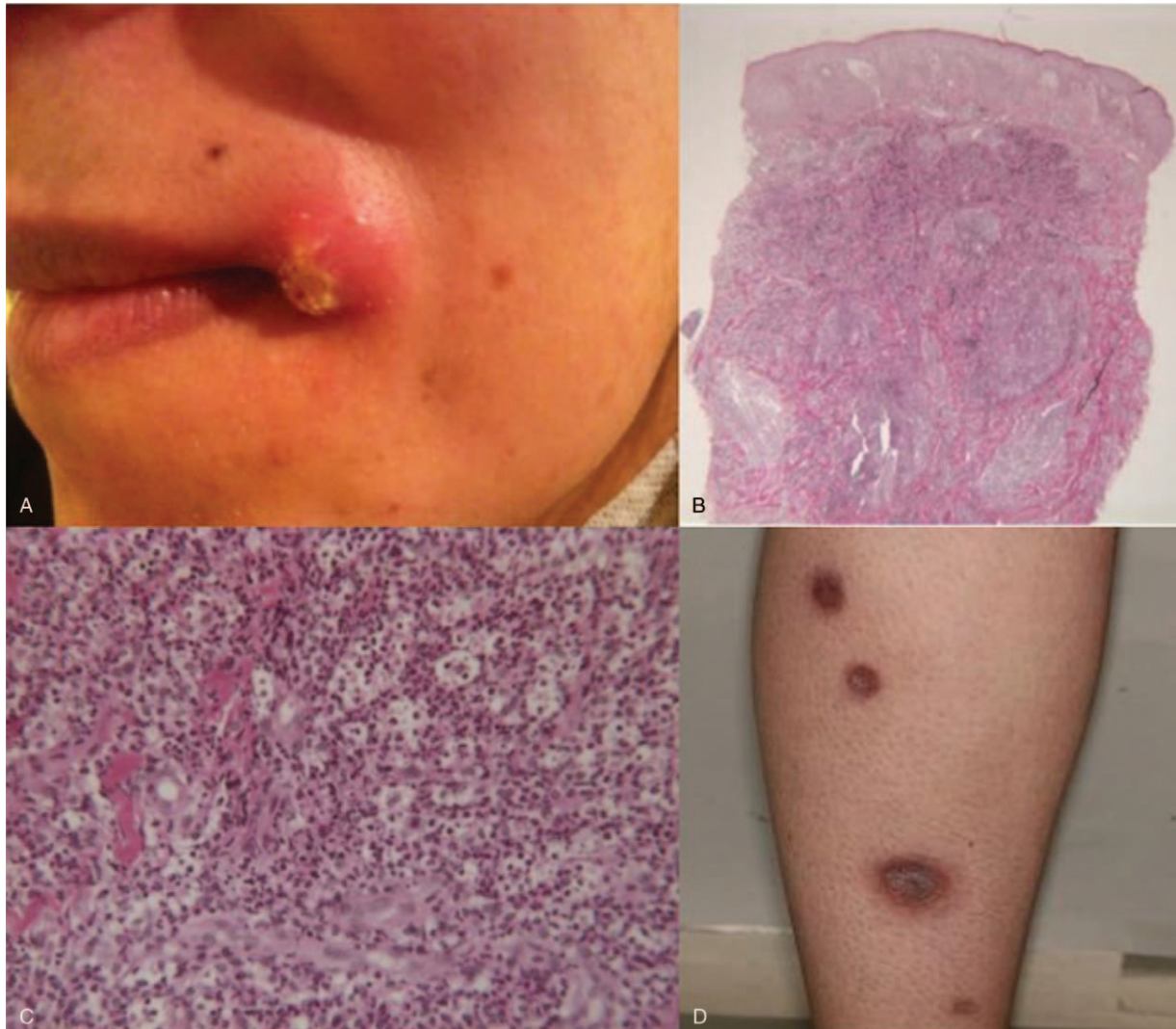
neutrophils, without any granuloma nor vascular damage (Fig. 1B, C). No pathogens including atypical mycobacteria or yeast were evidenced. The diagnosis of ulcerative variant of PG was retained.

A treatment with dermocorticoids and minocycline was started up, and complete resolution of PG was obtained within 6 months. Treatment was then discontinued. The patient did not present any recurrence of PG to date, with a 3-year follow-up.

### 3. Case 2

A 37-year-old woman was referred for ulcerative nodular lesions that had appeared on her legs within the last preceding weeks.

She had a medical history of peptic ulcer, Hashimoto thyroiditis, and Raynaud syndrome. In addition, she had been diagnosed with SLE 10 years before, in postpartum, given the association of lupus-specific skin lesions, nonerosive peripheral arthritis, antinuclear antibodies and anti-DNA antibodies. IgG anti-cardiolipin antibodies were positive at 21 IU (normal  $< 20$



**Figure 1.** (A) Lesion at the left angle of the mouth in patient 1. Histopathologic findings of cervical skin lesion of patient 1 revealing dermal neutrophilic infiltration without any granuloma nor vascular damage (hematoxylin-eosin-saffron stain). (B) Original magnification  $\times 25$ . (C)  $\times 200$ . (D) Multiple ulcerative papules of the posterior side of the right leg of patient 2.

IU) only once, whereas anti- $\beta$ 2 glycoprotein antibodies and lupus anticoagulant were never detected in her serum. Low dose oral prednisone (between 9 and 30mg/d) as well as hydroxychloroquine was prescribed since 10 years.

Clinical examination revealed multiple ulcerative papules of the posterior side of her legs (Fig. 1D), malar rash, inflammatory arthralgia of the hands. Histologic examination of ulcerations showed extensive neutrophilic infiltration in the entire height of the dermis. In addition, bleeding suffusions in the superficial dermis, thrombosis in several vessels of the reticular dermis, and hyperplasia of the epidermis were noticed. There was neither suppurative necrosis nor pathogens. Anti-DNA antibodies were positive and serum complement proteins were low. The diagnoses of ulcerative PG and lupus flare up were retained. Dapsone followed by colchicine and low doses of oral corticosteroids was first given but both were rapidly stopped because of side effects (cutaneous rash and gastrointestinal intolerance, respectively). Finally, oral corticosteroids (1 mg/kg/d) and methotrexate 30 mg per week allowed a complete healing of the PG and lupus lesions within months. Methotrexate was continued, while prednisolone was slowly decreased until 10 mg/d. To date, we have a 3-year follow-up, no recurrence of PG has been observed, whereas a flare up of lupus arthritis was diagnosed 2 years later.

#### 4. Discussion

We report here, 2 cases of women with SLE and ulcerative PG. No absolute criteria for the diagnosis of PG are available, but 2 groups proposed similar criteria we used. Both cases fulfilled the 3 major criteria and 2 minor criteria proposed by these groups.<sup>[1]</sup> Few cases of PG associated with SLE have been reported to date.

We found 22 cases of SLE and PG association in the literature, 1 PG/SLE/SGS association and 4 PG/SGS association.<sup>[5-24]</sup> Main features of the 23 cases of PG associated with SLE and our 2 additional patients are recapitulated in Table 1.

PG affects individuals of all ages, with a peak incidence between 20 and 50 years of age, and affects men and women almost equally.<sup>[1,3]</sup> However, there is a female predominance in nonmalignancy-associated PG.<sup>[1]</sup> Among the reported cases of patients with PG in a context of SLE, a large majority were women (18/22 cases for which gender was reported).<sup>[5-20]</sup> These patients with SLE were younger (mean age  $40.8 \pm 14.7$  years), than those with idiopathic PG or with PG associated with other conditions (mean age of 50.3, 59, and 61.5 years, respectively, in 3 different series of PG in the literature).<sup>[3,25,26]</sup>

Our 2 patients had ulcerative PG but the variant of PG was rarely specified in the 23 previously published.<sup>[5-20]</sup> No histologic difference was observed between SLE-associated PG and PG related to other conditions.<sup>[1,3]</sup>

Among the 23 cases of SLE-associated PG reported in the literature, most of the patients (16/23, 69%) had been diagnosed with SLE before the onset of PG, whereas PG was observed before the first symptom of SLE in only 3/23 cases.<sup>[5-20]</sup> Both diseases appeared simultaneously in 4 cases. In both cases described here, diagnosis of SLE preceded the development of PG, as observed in the majority of the published cases of such association.

Concerning the possible link between lupus activity and PG development, we analyzed in the literature, the clinical and biologic markers of lupus activity for the 20 patients diagnosed with SLE when PG occurred.<sup>[5-20]</sup> We found clinical symptoms of active lupus and/or complement consumption at the time of PG onset in 12/20 cases, no sign of lupus activity in 6 cases. In 2 cases,

**Table 1**  
Main features of the 25 cases of pyoderma gangrenosum associated with systemic lupus erythematosus.

Reference	Gender/age	Location of the lesion (number)	Time of PG onset compared to SLE diagnosis	Lupus activity at the time of PG onset
Case 1	F/32	Face (3)	1 year later	No activity
Case 2	F/37	Leg (mp)	10 years later	Signs of activity
Gonzalez-Moreno (2015) (5)	M/46	Leg (1)	After	Signs of activity
Gonzalez-Moreno (2015) (5)	F/63	Foot (1)	After	No activity
Gonzalez-Moreno (2015) (5)	F/42	Leg (1)	After	No activity
Gonzalez-Moreno (2015) (5)	F/46	Leg (mp)	After	No activity
Gonzalez-Moreno (2015) (5)	M/36	Foot (mp)	After	Signs of activity
Olson (1971) (6)	F/15	Leg (mp)	1 year before	Na
Peterson (1984) (7)	F/48	Intergluteal and inguinal folds (6)	Simultaneous	Signs of activity
Pinto (1991) (8)	F/35	Leg (1)	15 years later	Signs of activity
Hostetler (1993) (9)	F/27	Trunk, chest, knees (mp)	13 years later	No activity
Roger (1993) (10)	F/25	Foot (1)	1 month later	Signs of activity
Holbrook (1996) (11)	F/57	Leg (1)	2 years later	No activity
Schmid (1998) (12)	F/64	Leg (2)	11 years later	Signs of activity
Sakamoto (2002) (13)	F/55	Trunk, chest, shoulders (mp)	3 years later	Signs of activity
Waldman (2005) (14)	F/35	Leg (mp)	5 years before	Na
Reddy (2007) (15)	M/34	Legs (mp)	After	Signs of activity
Hind (2008) (16)	F/14month	Face, ends (mp)	Simultaneous	Signs of activity
Masattlioglu (2009) (17)	F/35	Leg (1)	Simultaneous	Signs of activity
Masattlioglu (2009) (17)	F/47	Thigh (1)	10 months before	Na
Canas (2010) (18)	ns/48	md (1)	2 years later	Md
Canas (2010) (18)	ns/28	md (1)	4 years later	Signs of activity
Canas (2010) (18)	ns/50	md (1)	Simultaneous	Md
Husein-ElAhmed (2010) (19)	M/36	Foot (1)	8 years later	No activity
Hamzi (2013) (20)	F/35	Leg (1)	18 years later	No activity

F=female, M=male, md=missing data, mp=multiple, na=not applicable, PG=pyoderma gangrenosum, SLE=systemic lupus erythematosus.

the clinical and biologic descriptions were not sufficient to determine lupus activity.

No guidelines are available concerning PG treatment in the setting of lupus. The therapeutic strategy in the general context of neutrophilic dermatosis consists in modulating activation, maturation or migration of neutrophils (with colchicine, dapsone, corticosteroids, and maybe minocycline). Interestingly, drugs able to modulate connective tissue diseases activity like SLE (such as hydroxychloroquine, methotrexate, mycophenolate mofetil, and cyclophosphamide) were reported effective on neutrophilic skin lesions in the literature.<sup>[5–21]</sup> This observation suggests that SLE and PG may share some common points in their pathogenesis.

The pathophysiology of PG is still unclear. An increasing body of evidence supports the role of proinflammatory cytokines like interleukin (IL)-1-beta, IL-17, and tumor necrosis factor (TNF)-alpha in the pathophysiology of NDs similarly to classic monogenic autoinflammatory diseases.<sup>[1,27]</sup> Multiple proinflammatory genes have been previously reported as highly upregulated in lesional skin of PG: STAT1, STAT3, TYK2, IL8, CCL5, IL-β, IL1R1, CD40, and CD40LG.<sup>[27]</sup> Overexpression of some Pattern Recognition Receptors, like Toll-like receptors, has also been recently reported in lesional skin.<sup>[28]</sup>

In SLE, the role of innate immunity has been a rapidly expanding area of research over the last decade. Immune complexes can activate the NLRP3 inflammasome, and SLE-derived macrophages are hyper-responsive to innate immune stimuli, leading to enhanced activation of the inflammasome and production of inflammatory cytokines.<sup>[29]</sup> Recent data indicate also that among innate immune cells, neutrophils could particularly take part to the pathogenesis of SLE.<sup>[30]</sup> Under various stimulations, neutrophils are able to generate extracellular structures, termed “NETs” (Neutrophil Extracellular Traps), notably containing DNA and histones. NETs display bacteriostatic function and participate in pathogens elimination. Growing evidences suggest that aberrant NETs generation may occur in noninfectious conditions such as autoimmune diseases and may contribute to the breaking of tolerance toward nuclear antigens exposed on traps. In patients with SLE, an imbalance between NETs formation and clearance has been evidenced that may participate in tolerance breaking, initiation of autoimmunity as well as in organ damages.<sup>[30]</sup> Taken together, these recent data may lead us to think that the association of PG and SLE may not be fortuitous.

Interestingly, other NDs have been observed in the setting of SLE like neutrophilic urticarial dermatosis, Sweet syndrome, palisaded neutrophilic granulomatous dermatitis, microbial pustulosis of the folds, and bullous lupus erythematosus.<sup>[31–33]</sup> All those observations have lead some physicians to delineate a new nosologic group of cutaneous lesions in SLE, referred as neutrophilic cutaneous lupus erythematosus.<sup>[34]</sup>

## 5. Conclusion

Although rare, the association of SLE and NDs like PG may be supported by common innate immunity dysregulation and abnormal neutrophil activation. PG and other NDs reported in patients with SLE may be added to the large clinical spectrum of neutrophilic cutaneous lupus erythematosus lesions.<sup>[34]</sup>

## Author contributions

Writing – original draft: Delphine Lebrun.

Writing – review & editing: Ailsa Robbins, Maxime Hentzien, Ségolène Toquet, Julie Plee, Anne Durlach, Jean-David Bouaziz, Firouzé Bani-Sadr, Amélie Servettaz.

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