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Pyoderma gangrenosum associated with hidradenitis suppurativa

A. Ah-Weng, J. A. A. Langtry,* S. Velangi,* C. D. Evans and W. S. Douglas

*Departments of Dermatology, Monklands Hospital, Lanarkshire and *Sunderland Royal Hospital, Sunderland, UK*

Summary

Pyoderma gangrenosum (PG) is associated with a number of systemic diseases. PG in association with hidradenitis suppurativa (HS) has been rarely reported. We describe six patients (three men, three women; aged 35–51 years), who developed PG on a background of HS. The onset of PG occurred only after HS had been present for at least two decades. No relationship in disease activity between the two conditions was observed. Three patients described previous severe adolescent acne vulgaris, one had concurrent systemic lupus erythematosus and another had chronic iron-deficiency anaemia. The course of PG was severe and refractory in four patients, who required treatment including high-dose oral corticosteroids, ciclosporin, intravenous immunoglobulin and intravenous cyclophosphamide.

Report

Pyoderma gangrenosum (PG) typically presents as painful ulceration with a dusky erythematous undermined edge. The most common site is the lower leg, but lesions can occur at any site. In 1930, Brunsting *et al.* described five patients with necrotic, enlarging skin ulcers, for which the term pyoderma (ecthyma) gangrenosum was introduced.[1] Four of the patients had ulcerative colitis, and subsequent reports have documented the association of PG with other systemic diseases, including Crohn's disease, monoclonal gammopathy and myeloma.[2] However, the presentation of hidradenitis suppurativa (HS) in patients with PG is rarely reported. We describe six patients with HS who later developed coexistent and, in some, refractory PG.

The details of each of the six patients are illustrated in Table 1. There were three women and three men, in whom PG presented from the third to fifth decades of life. The duration of onset ranged from 1 week to 6 years, and developed after HS had been present from 18 to 30 years, median 21. In our series, two patients developed ulcerative PG on their legs, one each over the chest, and one over the back of the hand. Of the two patients with superficial granulomatous PG, one presented with PG over her lower abdomen and thighs. In the other, superficial pyodermatous ulceration developed within the axillary vaults and groin folds, which resembled an exacerbation of HS (Fig. 1). In this case diagnostic confusion could have occurred as a result of ulceration within flexural sites typically affected by HS.

There was no observed correlation in disease activity between the two conditions at presentation or after treatment of PG. Three of the patients had suffered from severe acne vulgaris in adolescence, one had active systemic lupus erythematosus at the outset, and another presented with chronic iron-deficiency anaemia.

Except for patient 3, who had developed typical ulceration of pyoderma gangrenosum (Fig. 2), all other patients had biopsies from which histological staining and culture found no evidence of fungal or atypical mycobacterial infection. All the patients had haematological, biochemical and myeloma screens, and autoantibody profiles to investigate for underlying systemic conditions associated with PG.

The association of PG with numerous systemic disorders is well established. However, we found only nine cases of coexisting PG and HS from a search of the English-language, peer-reviewed literature, as outlined in Table 2.[2–7] In a review of 86 patients with PG, arthritis was present in 37% of patients and inflammatory bowel disease in 36%, while 10% had a

Correspondence: A. Ah-Weng, Department of Dermatology, Sunderland Royal Hospital, Kayll Road, Sunderland SR4 7TP, UK.
E-mail: allanahweng@yahoo.co.uk

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Table 1 Summary of patients with pyoderma gangrenosum and hidradenitis suppurativa in the current series (2003).

Patient ID	Age/sex	Duration of HS (years)	Pyoderma gangrenosum				
			Onset	Site	Type	Treatment dosage/day (outcome)	Other associated disease
1	45/F	22	2 years	Chest and axillae	Ulcerative	Prednisolone 40 mg (NR), azathioprine 150mg (NR), IVIG 2 mg/kg monthly (R), i.v. cyclophosphamide 500 mg 3-weekly (R), (hydroxychloroquine 200 mg)	SLE and glomerulonephritis
2	51/M	30	1 week	Back of hands	Ulcerative	Prednisolone 60 mg (R)	Severe acne vulgaris
3	35/F	18	3 years	Lower legs	Ulcerative	Prednisolone 12.5 mg (NR), ciclosporin A 4.5 mg/kg (R)	psoriasis, paranoid schizophrenia
4	42/M	20	6 years	Back of thigh, lower abdomen	Superficial	Ciclosporin A 5 mg/kg (NR), trimethoprim 200 mg	Severe acne vulgaris
5	44/M	20	4 months	Lower legs	Ulcerative	Minocycline 200 mg (NR) dapsone 100 mg (NR), ciclosporin A 4 mg /kg (R)	Severe acne vulgaris, HS Left axillae excised
6	57/F	25	1 month	Groin and axillae	Superficial	Prednisolone 30 mg (R)	Iron-deficiency anaemia

Treatment response in PG: R, responsive, NR, not responsive. IVIG, intravenous immunoglobulin, SLE, systemic lupus erythematosus.

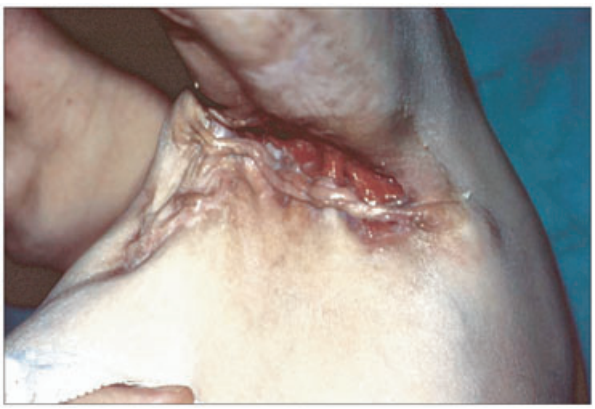

Figure 1 Superficial pyoderma gangrenosum in the left axillary vault over the site of hidradenitis suppurativa (patient 6).

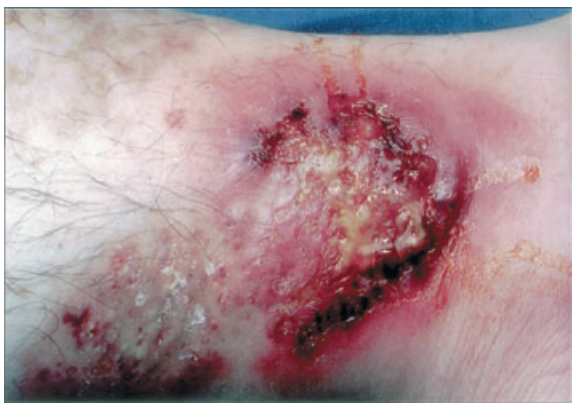

Figure 2 Sloughy ulcer with raised dusky red edge and adjacent cribriform scarring over the medial aspect of the left ankle (patient 3).

monoclonal gammopathy and 5% (4 patients) had associated HS.[2] A similar survey of disease associated with PG revealed one patient with HS.[7] Of 10 patients reviewed with HS and/or acne conglobata followed up for spondyloarthropathy, one patient was reported to have associated PG.[3] In one case report, PG was a problem in the management of a patient with seronegative polyarthropathy, cystic acne and HS.[4] For another, HS responded to ciclosporin together with improvement of PG during treatment.[5] In a patient with Behçet's disease treated with colchicine, concurrent axillary HS, bullous PG and perianal fistulae were also present.[6]

In HS, acute inflammation of the apocrine glands results in sinuses, abscess formation and scarring. The initiating trigger appears to be an occluding infundibulofolliculitis with secondary apocrine gland involvement.[8] However, its pathogenesis is unclear and an underlying immune abnormality has not been established, although dysfunctional neutrophils have been implicated.[9] Other aetiological factors include an inherited tendency and hormonal influence.[10]

The pathogenesis of PG also remains unknown, although numerous defects of the immune system have been implicated including defective neutrophil chemotaxis and phagocytosis, reduced lymphokine production and migration.[11] In some cases, cutaneous anergy to *Candida albicans*, streptokinase and dinitrochlorobenzene has been described.[9] In a series of 65 patients, 36 (55%) had immunoglobulin and complement deposition along

Table 2 Summary of patients with pyoderma gangrenosum and hidradenitis suppurativa.

Ref ID	No. of patients	Age/sex	Duration of HS (years)	Pyoderma gangrenosum			Treatment dosage/day (outcome)	Other associated disease
				Onset	Site	Type		
von den Driesch[7]	1	–	–	–	Groin and axillae	Superficial	Oral corticosteroids	–
Shenefelt[4]	1	42/M	12	3 months	Lower legs	Ulcerative	Prednisolone 40 mg (R), intralesional triamcinolone (R), sulfasalazine 500 mg three times daily (R), minocycline 100 mg twice daily (R)	Cystic acne, seronegative arthritis
Buckley *et al.*[5]	1	48/M	8	4 months	Right leg	Ulcerative	Minocycline (NR), ciclosporin A 4.5 mg/kg (R)	HS, responded to ciclosporin
Powell FC *et al.*[2]	4	–	–	–	–	–	–	–
Rosner *et al.*[3]	1	24/M	–	–	–	–	–	Acne conglobata, peripheral and axial arthropathy
Raynor *et al.*[6]	1	33/F	<1	1 month	Lower legs	Bullous	Colchicine 0.6 mg twice daily (R)	PG, HS and perianal fistula-complicated Behçet's disease

Treatment response in PG: R, responsive, NR, not responsive. –Information not available.

the endothelial wall, which indicated a vasculitic aetiology.[12] However, the only suggested common link for both HS and PG is defective neutrophil function, which has been described in both conditions but there are no studies to support impaired neutrophil activity as a common aetiological pathway.

Conclusions

We would like to highlight the observed association of PG and HS in a series of patients. PG appears to develop after long-standing HS. There was no correlation of disease activity observed between these two conditions.

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