

The chronicity and disease burden of generalized pustular psoriasis

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Background



- GPP is a rare, inflammatory skin disease characterized by sudden, widespread eruption or flares of small sterile pustules, often with systemic symptoms¹
- Many patients experience chronic symptoms between flares and may require continuous management²
- Here, we describe the long-term burden of GPP and its effects on QoL in 10 patients who presented to US hospitals or outpatient clinical practices with GPP flare

Conclusions

- GPP is associated with both recurrent flares and chronic symptoms between flares, leading to decreased patient QoL, increased morbidity, and risk of complications
- Safe and effective long-term treatment options for GPP are needed to mitigate disease burden



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Key themes



Case presentations

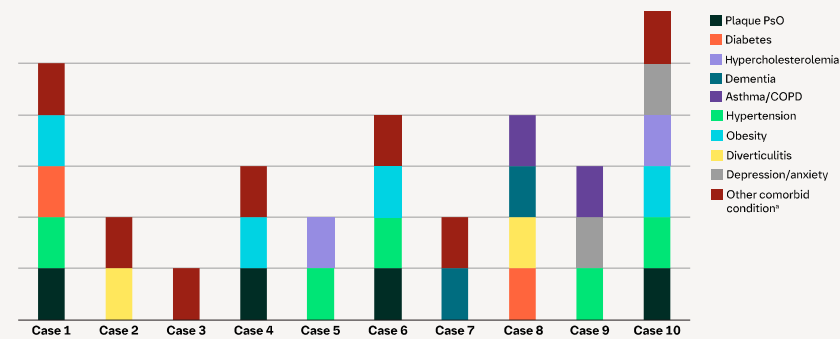
- 10 patients aged 18–92 years had heterogeneous presentations of GPP flare in addition to several comorbid conditions (Table 1, Figure 1)
- All patients experienced negative QoL burden related to GPP (Figure 2)

Table 1. Summary of patient cases

Case Report	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8	Case 9	Case 10
Patient demographics	36-year-old American Indian male	58-year-old White female	40-year-old White female	18-year-old Asian male	73-year-old White male	62-year-old White female	92-year-old White female	72-year-old Black female	47-year-old Black female	39-year-old White male
GPP presentation	Severe GPP presentation with systemic symptoms	Moderate to severe flare of sterile pustules that progressed to whole body, with no systemic symptoms	Pustular plaques on the extremities and trunk with systemic symptoms	Plaques studded with pustules of moderate skin severity affecting 15–20% of BSA	Numerous annular and arcuate erythematous plaques studded with pustules, and with trailing scale	Severe, diffuse erythematous rash with sheets of desquamation and sensation of heat in skin; fever, and chills	Erythematous, scaly papules coalescing into plaques on trunk and extremities; fever, arthralgias, and altered mental status	Severe erythroderma and widespread pustules, skin pain, fever, and chills	Erythema, swelling, and pustules on trunk and extremities; severe joint pain and muscle aches	Localized erythematous eczematous rash to anterior neck and trunk, with no systemic symptoms
Flare trigger	IV and PO corticosteroid	HCQ	HCQ	Possibly COVID	HCQ, corticosteroid withdrawal	Possibly COVID	Corticosteroid withdrawal	Stress	Unknown	Stress
Hospitalized?	Yes	No	Yes	No	No	Yes	Yes	Yes	No	No
Initial diagnosis	GPP	AGEP	AGEP	Guttate PsO	Nummular dermatitis	GPP	GPP	GPP	RA and lupus	Contact dermatitis

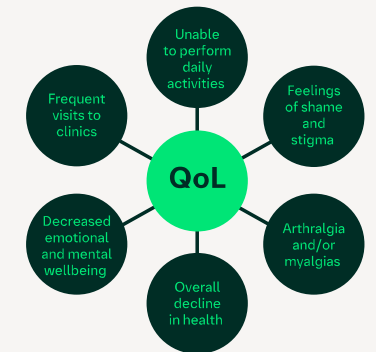
- All patients had comorbid conditions; hypertension was the most common, followed by plaque PsO (Figure 1)
- 4 patients had a history of GPP
- 6 patients were initially misdiagnosed, leading to delays in treatment
- 5 patients were hospitalized due to the severity of their GPP flare, and multiorgan failure was reported in 2 patients
- Patients reported GPP decreased their QoL, with impaired confidence, mood, and ability to carry out activities of daily living (Figure 2)
 - Some patients had limited mobility; Case 1 required a wheelchair, and Case 9 was initially bedridden and then required a walker
 - Case 4 reported being bullied due to the appearance of his skin, while Case 10 felt that his disease was difficult to manage after failing multiple therapies
- After failure of off-label therapies, all 10 patients received targeted GPP therapy with IV spesolimab

Figure 1. Burden of GPP and comorbid conditions



*Others included obstructive sleep apnea, non-specific arthritis, Raynaud's, granuloma annulare, autoimmune hepatitis, autism, SHOX gene duplication, tachycardia, colorectal cancer, osteomyelitis, history of allergic contact dermatitis, and history of cerebral vascular accident

Figure 2. Burden of GPP on patient QoL



Abbreviations
BSA: body surface area; COVID: coronavirus disease 2019; COPD: chronic obstructive pulmonary disease; GPP: generalized pustular psoriasis; HCQ: hydroxychloroquine; IV: intravenous; PO: by mouth; PsO: psoriasis; QoL: quality of life; RA: rheumatoid arthritis

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2. Eivaski B, Lenzini MS. *Clin Cosmet Invest Dermatol* 2024;7:2407-740.

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