

CASE AND RESEARCH LETTER

[Translated article] Elderly-Onset Generalized Pustular Psoriasis in the Elderly: A 5-Case Series



Psoriasis pustulosa generalizada en pacientes mayores de 65 años: serie de 5 casos clínicos

To the Editor,

Although psoriasis is a chronic and systemic inflammatory disease characterized by a bimodal incidence, it can manifest at any age.¹ Literature on the behavior of this disease in the elderly is scarce, despite the significant frequency of cases in this age range. While plaque psoriasis is the most common clinical sign overall, and particularly in older age,² pustular forms of psoriasis in the elderly—excluding palmoplantar pustulosis (PPP)—have been poorly characterized.^{1,3,4}

We conducted a retrospective, observational study with patients older than 65 years with a diagnosis of late-onset pustular psoriasis (PP)—onset after the age of 50—with a > 4-week history of generalized pustulosis without associated palmoplantar pustulosis, who had already been seen and/or were being followed in the psoriasis clinic at Hospital General Universitario Gregorio Marañón, Madrid, Spain from January 2000 through December 2021. Clinical, analytical, therapeutic, and evolutionary data were obtained from the electronic health records of the department.

A total of 5 patients were included (Table 1), 3 of whom were male (60%). These 3 patients exhibited plaque psoriasis lesions before the onset of pustular signs; the time from the diagnosis of plaque psoriasis to the development of generalized pustular forms was 31, 34, and 42 years, respectively. The 2 women had no personal or family history of psoriasis or other dermatological diseases. The age at diagnosis of pustular forms went from 50 up to 83 years (Table 1).

In 4 cases, the mutational status of the IL36 receptor antagonist gene (IL36RN) was analyzed, but no pathological variants were found in any of these cases.

All patients required hospitalization at some point during the course of their PP, with 2 cases requiring hospitalization at the onset of the pustular form due to fever and systemic symptoms (generalized pustular psoriasis [GPP] von Zumbusch).

The 5 patients in the series received topical corticosteroids and acitretin as first-line systemic therapy—usual therapeutic doses, 10 to 25 mg/day—which was sufficient to control the skin symptoms in only 1 case. The combination with apremilast was the control treatment in 2 patients, and in the remaining cases, biological treatment with guselkumab was used.

Compared with series on vulgar psoriasis, studies on the epidemiological and clinical-evolutionary characteristics of patients with PP are scarce,^{5–10} and even more so when talking about patients older than 60–65 years, with only 1 series specifically addressing this patient subgroup.⁵

The diagnosis of pustular forms of psoriasis is usually established around the 4th decade of life,⁶ forms in which mutations associated with PP are more prevalent.^{7–9}

In the series of 7 cases of GPP in patients older than 60 years—mean age 70.7 ± 9.2 years—⁵ no patients had a personal or family history of psoriasis, and the genetic study turned out negative in 6 of the 7 cases analyzed. Compared with a series of 32 patients with GPP without age differentiation, older patients had a greater tendency towards erythroderma.^{5,7–9}

Although rare, PP is a disease that, especially in its GPP form, can become severe, leading to patient death and frequently requiring hospitalization.^{8–10} In our setting, the need for intensive care unit admission is estimated at 6.1%, with a mortality rate of 4.8% for GPP patients overall, without considering age ranges.¹⁰ In our series, all patients required hospitalization at some point during the course of the disease. Two out of the 7 patients from the above-mentioned series⁵ required hospitalization.

Regarding treatment, elderly patients, *per se*, are a more fragile population,¹ with more comorbidities^{1,4} and, considering the potential severity of the disease, all this can complicate therapeutic management.^{2,3} That is why the use of biological drugs in the management of GPP is on the rise.^{6,8,9} Similarly, although in our series of 7 elderly patients with GPP,⁵ acitretin was the first-line therapy, 3 required biological therapies.

PP can be a late-onset condition, especially in patients without prior plaque psoriasis,^{5,8} requiring systemic treat-

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Table 1 Characteristics of the patients included in the series.

	Case #1	Case #2	Case #3	Case #4	Case #5
Sex	Female	Male	Male	Female	Male
Current Age (years)	92	65	76	82	85
Personal history	Atrial fibrillation Anticoagulated	HIV infection HTN DL	Rectal carcinoma HTN	HTN Type 2 DM Chronic kidney disease on hemodialysis	Chronic kidney disease on hemodialysis
Age at psoriasis diagnosis (years)	50	15	40	62	52
Age at pustular psoriasis diagnosis (years)	50	57	74	62	83
Family history of psoriasis	No	No	No	No	No
Clinical presentation	Annular and circinate-type GPP	Annular and circinate-type GPP	Annular and circinate-type GPP	GPP von Zumbusch	GPP von Zumbusch
Confirmatory skin biopsy	No	No	No	No	Yes
IL36RN mutation study /Result	Yes / Negative	Yes / Negative	Yes / Negative	Yes / Negative	No
Topical corticosteroid treatment	Yes	Yes	Yes	Yes	Yes
Phototherapy	No	Yes (NB-UVB)	Yes (NB-UVB)	No	Yes (NB-UVB)
Systemic treatment	Acitretin	Acitretin	Acitretin	Methotrexate Ciclosporin Acitretin	Acitretin
Apremilast treatment	No	Yes	No	No	Yes
Biological treatment	No	No	Yes	Yes	No
Lines of biological treatment	–	–	Guselkumab	Etanercept Adalimumab Ustekinumab Ixekizumab Guselkumab	–
Control drug	Acitretin	Acitretin + Apremilast	Guselkumab	Guselkumab	Acitretin + Apremilast
Time to disease control (months)	4	6.5	5	24	1.5

DL: dyslipidemia; DM: diabetes mellitus; HTN: hypertension; IL36RN: IL36 receptor antagonist; GPP: generalized pustular psoriasis; NB-UVB: narrowband ultraviolet B; HIV: human immunodeficiency virus.

ment for its control.⁹ It can manifest severely with systemic involvement¹⁰ and does not necessarily carry mutations in IL36RN,^{5–7} especially when it is in the elderly.^{7–9} Despite the typical immunosenescence of these patients, these cases can be resistant to therapy and may require biological therapy to achieve remission of lesions and control of psoriasis.⁸

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Authors' contributions

All authors contributed to the drafting of this work, and agreed on the publication of this article.

Conflicts of interest

None declared.

References

1. Kassi K, Djeha D, Gbery IP, Kouame K, Sangaré A. Psoriasis in elderly patients in the Côte d'Ivoire: socio-demographic, clinical and therapeutic aspects and follow-up. *Int J Dermatol.* 2016;55:e83–6.
2. Phan C, Sigal ML, Estève E, Reguiat Z, Barthélémy H, Beneton N, et al. Psoriasis in the elderly: epidemiological and clinical aspects and evaluation of patients with very late onset psoriasis. *JEADV.* 2016;30:78–162.
3. Kwon HH, Kwon IH, Youn JI. Clinical study of psoriasis occurring over the age of 60 years: is elderly-onset psoriasis a distinct subtype? *Int J Dermatol.* 2012;51:53–8.
4. van Winden MEC, ter Haar ELM, Groenewoud HMM, van de Kerkhof PCM, de Jong EMGJ, Lubeek SFK. Disease and treatment characteristics in geriatric psoriasis: a patient survey comparing age groups. *Acta Derm Venereol.* 2020;100:adv00215.
5. Chao JP, Tsai TF. Elderly-onset generalized pustular psoriasis: a case series. *Clin Exp Dermatol.* 2022;14:1567–70.
6. Zheng J, Chen W, Gao Y, Chen F, Yu N, Ding Y, et al. Clinical analysis of generalized pustular psoriasis in Chinese

- patients: a retrospective study of 110 patients. *J Dermatol.* 2021;48:1336–42.
7. Twelves S, Mostafa A, Dand N, Burri E, Farkas K, Wilson R, et al. Clinical and genetic differences between pustular psoriasis subtypes. *J Allergy Clin Immunol.* 2019;143:1021–6.
 8. Ohata C, Tsuruta N, Yonekura K, Higashi Y, Saito K, Katayama E, et al. Clinical characteristics of Japanese pustular psoriasis: a multicenter observational study. *J Dermatol.* 2022;49:142–50.
 9. Wang TS, Chiu HY, Hong JB, Chan CC, Lin SJ, Tasi TF. Correlation of IL36RN mutation with different clinical features of pustular psoriasis in Chinese patients. *Arch Dermatol Res.* 2016;308:55–63.
 10. Montero-Vilchez T, Grau-Perez M, García-Doval I. Epidemiología y distribución espacial de la psoriasis pustulosa generalizada en España: un análisis poblacional a nivel nacional basado en datos de ingresos hospitalarios (2016-2020). *Actas Dermosifiliogr.* 2023;1114:97–101.
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