

Dermatology Reports

The efficacy of apremilast in pemphigus: a systematic review of case reports

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doi:10.4081/dr.2025.10245

SUPPLEMENTARY MATERIAL

Supplementary Table 1. Baseline characteristics of the included cases.

Study	Study design	Age	Gender	Diagnosis	Baseline disease severity	Affected area	Prior treatments	Comorbidities
Zhang <i>et al.</i> , China (2024a) ¹¹	Case report	54	Female	PF	Mild (PDAI: 7; NRS: 8)	Trunk	Topical mometasone furoate cream	Diabetes mellitus and atrophic gastritis
Zhou <i>et al.</i> , China (2023) ¹⁷	Case report	28	Female	IGAP	Moderate-to-severe (PDAI: 24; VAS for pruritus: 9) and refractory	Trunk and extremities	Prednisolone, tetracycline, sulfasalazine	None
Meier <i>et al.</i> , Germany (2020) ¹⁵	Case report	62	Female	PV	Severe (ABSIS: 38) and refractory	Oral cavity	Azathioprine, mycophenolate mofetil, dapsone, intravenous immunoglobulins, systemic steroids, rituximab	Not reported
Delvaux <i>et al.</i> , France (2024) ¹⁸	Case report	67	Female	PV	Severe mucosal involvement (NRS for pain: 8)	Mucosal (ocular, oral, pharyngeal, laryngeal, nasal, and esophageal involvement)	Colchicine, prednisolone mouthwashes	Hypertension, type 2 diabetes mellitus, dyslipidemia
Zhang <i>et al.</i> , China (2024b) ¹⁶	Case series of 3 patients	Case 1: 56 Case 2: 74 Case 3: 26	Case 1: Female Case 2: Male Case 3: Female	Case 1: PF Case 2: PV Case 3: PV	Moderate-to-severe (PDAI: 63, 21 and 29, respectively)	Not reported	Case 1: Intravenous prednisolone, cyclophosphamide shock therapy Case 2: Intravenous immunoglobulin and mycophenolate mofetil Case 3: Prednisolone	Case 1: Hypertension, diabetes mellitus, hepatitis B, sepsis Case 2: Hypertension, cataract, cellulitis Case 3: Syphilis

PF, pemphigus foliaceus; PV, pemphigus vulgaris; IGAP, IgA pemphigus; PDAI, Pemphigus Disease Area Index; VAS, Visual Analogue Scale; ABSIS, Autoimmune Bullous Skin Disorder Intensity Score; NRS, Numerical Rating Score.

Supplementary Table 2. Treatment characteristics and outcomes in the included studies.

Treatment characteristics or outcomes	Zhang <i>et al.</i> , China (2024a) ¹¹	Zhou <i>et al.</i> , China (2023) ¹⁷	Meier <i>et al.</i> , Germany (2020) ¹⁵	Delvaux <i>et al.</i> , France (2024) ¹⁸	Zhang <i>et al.</i> , China (2024b) ¹⁶
Initial apremilast dose	60 mg (30 mg twice daily)	-	-	Gradual increase	-
Maintenance apremilast dose	30 mg once daily	60 mg/day	60 mg (30 mg twice daily)	60 mg (30 mg twice daily)	60 mg (30 mg twice daily)
Concomitant treatment	-	-	Prednisone (reduced from 20 mg to 5 mg) and mycophenolate mofetil (2 g)	Prednisone (1 mg/kg/day) and rituximab (1g at two-week intervals) during the last 5 weeks of treatment with apremilast	Case 1: Tapered prednisone Case 2: Prednisolone (0.4mg/kg/day) Case 3: Prednisolone (0.5 mg/kg/day)
Treatment duration	38 weeks	Not reported	32 weeks	11 weeks followed by a 2-week discontinuation then reintroduction for 9 weeks	8 weeks
Time to improvement	4 weeks	4 weeks	12 weeks	3 weeks	No improvement noted after 8 weeks
Efficacy outcomes	Significant clinical improvement (the PDAI decreased from 7 to 1, and the NRS from 8 to 0) Serum anti-Dsg1 antibodies decreased from 28.18 to 1.48 kU/L Treg and iTreg cells increased, reaching 8.15% and 9.59% respectively	Significant clinical improvement (the PDAI decreased from 24 to 0, and the VAS for pruritus decreased from 9 to 3)	Significant clinical improvement (the ABSIS decreased from 38 to 0) Serum anti-Dsg1 antibodies decreased from 200 to 139 kU/L and anti-Dsg3 from 117 to 8 U/mL Increase in Treg (from 9% to 17%) and Treg cells (from 7.5% to 15%) No change on the Th and Tfh inflammatory subsets	Significant clinical improvement (fewer lesions, cessation of new lesions, decrease in NRS from 8 to 3, and weight stabilization) Serum anti-Dsg3 antibodies decreased from 129 to 14 U/mL	No response
Adverse events	None	None	Not reported	Not reported	None
Relapse	None	None	None	Rapid relapse upon apremilast discontinuation; improvement resumed after reintroduction	-

PDAI, Pemphigus Disease Area Index; VAS, Visual Analogue Scale; ABSIS, Autoimmune Bullous Skin Disorder Intensity-Score; NRS, Numerical Rating Score; Dsg, desmoglein; Treg, T regulatory; iTreg, induced T regulatory; Tfr, T follicular regulatory; Th, T helper; Tfh, T follicular helper.