

Table SI. Patients' treatment scheme from baseline to month 12

Pat. No.	Age, years/Sex	Lesions	Weight, kg	Baseline	Month 6	Month 12
1	32/F	Cutaneous+mucosal	86	100	5 + AZA	5
2	52/M	Cutaneous+mucosal	89	100	5 + CP	5
3	46/F	Mucosal	88	90	5	5
4	38/F	Cutaneous	82	90	5	5
5	32/F	Mucosal	65	70	5	5
6	36/F	Mucosal	75	80	5	5
7	70/F	Cutaneous+mucosal	80	80	5	5
8	25/M	Mucosal	83	100	5	5
9	66/M	Cutaneous+mucosal	84	90	5	5
10	41/F	Mucosal	73	80	5 + AZA	5
11	55/M	Cutaneous+mucosal	95	100	5 + AZA	5
12	26/F	Cutaneous+mucosal	52	60	5 + AZA	5
13	51/M	Cutaneous+mucosal	110	120	5 + AZA	5
14	26/M	Cutaneous+mucosal	60	80	5 + AZA	5
15	38/M	Cutaneous+mucosal	105	120	5 + AZA	5
16	45/M	Cutaneous+mucosal	90	100	5 + AZA	5
17	43/M	Mucosal	67	80	5 + AZA	5
18	38/F	Mucosal	70	80	5 + AZA	5
19	34/F	Cutaneous+mucosal	115	120	5	5
20	41/M	Cutaneous+mucosal	82	90	5 + CP	5
21	66/F	Cutaneous+mucosal	60	70	5 + AZA	5
22	43/M	Cutaneous+mucosal	93	100	5 + AZA	5
23	79/F	Cutaneous+mucosal	65	70	5	5
24	58/F	Cutaneous	73	80	5	5
25	40/F	Mucosal	63	70	5 + AZA	5
26	76/F	Cutaneous	72	80	5	5
27	52/M	Cutaneous	85	90	5	5
28	30/M	Cutaneous	84	90	5	5
29	38/M	Cutaneous+mucosal	87	90	5	5
30	78/F	Cutaneous	80	80	5	5
31	74/M	Mucosal	80	80	5 + AZA	5
32	37/F	Mucosal	58	60	5 + AZA	5
33	33/F	Mucosal	54	60	5	5
34	72/M	Cutaneous+mucosal	50	80	5	5
35	64/M	Cutaneous	65	70	5	5

AZA: azathioprine, CP: cyclophosphamide, all therapies in mg of prednisolone

Table SII. Descriptive statistics of clinical scores, anti-desmoglein-1 (anti-DSG-1) autoantibodies and anti-DSG-3 autoantibodies for each subgroup of patients at baseline, month 6 and month 12

Time-point	Clinical characteristics	Subgroup A	Subgroup B	Subgroup C
		(n=7) Median (min–max)	(n=11) Median (min–max)	(n=17) Median (min–max)
Baseline	ABSIS (Total)	13.0 (1.5–94.5)	19.0 (9.5–54.0)	39.5 (11.5–168.0)
	Skin involvement score	13.0 (1.5–94.5)	0.0 (0.0–0.0)	24.0 (4.5–112.0)
	Mucosal extent score	0.0 (0.0–0.0)	3.0 (1.0–10.0)	3.0 (1.0–11.0)
	Mucosal severity score	0.0 (0.0–0.0)	16.0 (8.5–44.0)	16.0 (4.5–45.0)
	PDAI (Total)	10.0 (3.0–51.0)	20.0 (2.0–32.0)	25.0 (9.0–158.0)
	Cutaneous lesions score	10.0 (3.0–51.0)	0.0 (0.0–0.0)	13.0 (3.0–79.0)
	Mucous membranes score	0.0 (0.0–0.0)	20.0 (2.0–32.0)	4.0 (2.0–79.0)
	DSG-1	142.6 (31.3–214.6)	6.5 (3.5–22.2)	115.4 (3.8–204.7)
Month 6	DSG-3	6.1 (2.1–143.9)	154.7 (23.3–192.7)	133.2 (21.2–181.3)
	ABSIS (Total)	0.0 (0.0–0.0)	0.0 (0.0–12.0)	0.0 (0.0–75.0)
	Skin involvement score	0.0 (0.0–0.0)	0.0 (0.0–0.0)	0.0 (0.0–48.0)
	Mucosal extent score	0.0 (0.0–0.0)	0.0 (0.0–3.0)	0.0 (0.0–5.0)
	Mucosal severity score	0.0 (0.0–0.0)	0.0 (0.0–9.0)	0.0 (0.0–22.0)
	PDAI (Total)	0.0 (0.0–0.0)	0.0 (0.0–6.0)	0.0 (0.0–34.0)
	Cutaneous lesions score	0.0 (0.0–0.0)	0.0 (0.0–0.0)	0.0 (0.0–27.0)
	Mucous membranes score	0.0 (0.0–0.0)	0.0 (0.0–6.0)	0.0 (0.0–7.0)
Month 12	DSG-1	10.5 (4.3–16.9)	4.8 (3.2–10.5)	8.9 (0.7–134.8)
	DSG-3	6.5 (1.6–13.9)	81.4 (4.7–159.0)	14.5 (2.1–170.2)
	ABSIS (Total)	0.0 (0.0–0.0)	0.0 (0.0–37.0)	0.0 (0.0–30.5)
	Skin involvement score	0.0 (0.0–0.0)	0.0 (0.0–0.0)	0.0 (0.0–10.5)
	Mucosal extent score	0.0 (0.0–0.0)	0.0 (0.0–7.0)	0.0 (0.0–3.0)
	Mucosal severity score	0.0 (0.0–0.0)	0.0 (0.0–30.0)	0.0 (0.0–17.0)
	PDAI (Total)	0.0 (0.0–0.0)	0.0 (0.0–30.0)	0.0 (0.0–15.0)
	Cutaneous lesions score	0.0 (0.0–0.0)	0.0 (0.0–0.0)	0.0 (0.0–12.0)
Month 12	Mucous membranes score	0.0 (0.0–0.0)	0.0 (0.0–30.0)	0.0 (0.0–11.0)
	DSG-1	5.5 (3.1–8.9)	4.7 (0.7–15.3)	6.9 (0.5–33.8)
	DSG-3	4.8 (1.1–7.1)	51.6 (3.6–144.6)	6.7 (1.1–144.5)

Table SIII. Rh- and p-values of autoantibodies titres and clinical scores from baseline to month 12

Time point	Clinical characteristics	Subgroup A (n=7)		Subgroup B (n=11)		Subgroup C (n=17)	
		Anti-DSG-1 Rh/p-value	Anti-DSG-3 Rh/p-value	Anti-DSG-1 Rh/p-value	Anti-DSG-3 Rh/p-value	Anti-DSG-1 Rh/p-value	Anti-DSG-3 Rh/p-value
Baseline	ABSIS (Total)	0.536/0.215	-0.214/0.645	-0.023/0.947	0.236/0.484	0.608/0.010	0.174/0.505
	Skin involvement score	0.739/0.058	-0.072/0.878	N/A	N/A	0.650/0.005	0.134/0.609
	Mucosal extent score	N/A	N/A	0.174/0.608	0.422/0.196	0.133/0.610	0.325/0.203
	Mucosal severity score	N/A	N/A	-0.050/0.883	0.219/0.517	0.456/0.066	0.327/0.200
	PDAI (Total)	0.739/0.058	-0.072/0.878	0.162/0.635	0.355/0.284	0.591/0.013	0.326/0.202
	Cutaneous lesions score	0.739/0.058	-0.072/0.878	N/A	N/A	0.736/0.001	0.314/0.219
	Mucous membranes score	N/A	N/A	0.162/0.635	0.355/0.284	0.186/0.475	0.339/0.184
Month 6	ABSIS (Total)	N/A	N/A	0.662/0.026	0.378/0.252	0.637/0.006	0.442/0.076
	Skin involvement score	N/A	N/A	N/A	N/A	0.637/0.006	0.442/0.076
	Mucosal extent score	N/A	N/A	0.672/0.023	0.373/0.259	0.408/0.104	0.408/0.104
	Mucosal severity score	N/A	N/A	0.662/0.026	0.378/0.252	0.408/0.104	0.408/0.104
	PDAI (Total)	N/A	N/A	0.662/0.026	0.378/0.252	0.637/0.006	0.442/0.076
	Cutaneous lesions score	N/A	N/A	N/A	N/A	0.637/0.006	0.442/0.076
	Mucous membranes score	N/A	N/A	0.662/0.026	0.378/0.252	0.408/0.104	0.408/0.104
Month 12	ABSIS (Total)	N/A	N/A	0.475/0.140	0.755/0.005	0.551/0.022	0.735/0.001
	Skin involvement score	N/A	N/A	N/A	N/A	0.665/0.004	0.637/0.006
	Mucosal extent score	N/A	N/A	0.475/0.140	0.755/0.005	0.366/0.149	0.570/0.017
	Mucosal severity score	N/A	N/A	0.676/0.022	0.661/0.027	0.366/0.149	0.570/0.017
	PDAI (Total)	N/A	N/A	0.475/0.140	0.755/0.005	0.548/0.023	0.731/0.001
	Cutaneous lesions score	N/A	N/A	N/A	N/A	0.662/0.004	0.634/0.006
	Mucous membranes score	N/A	N/A	0.475/0.140	0.755/0.005	0.514/0.035	0.715/0.001