

Fig. S1. Schematic presentation of BP180 with its subdomains. The positions of the 120 kDa LAD-1, the 97 kDa LABD97 and BP180-C recombinant protein (BP180-C RP) are also shown.

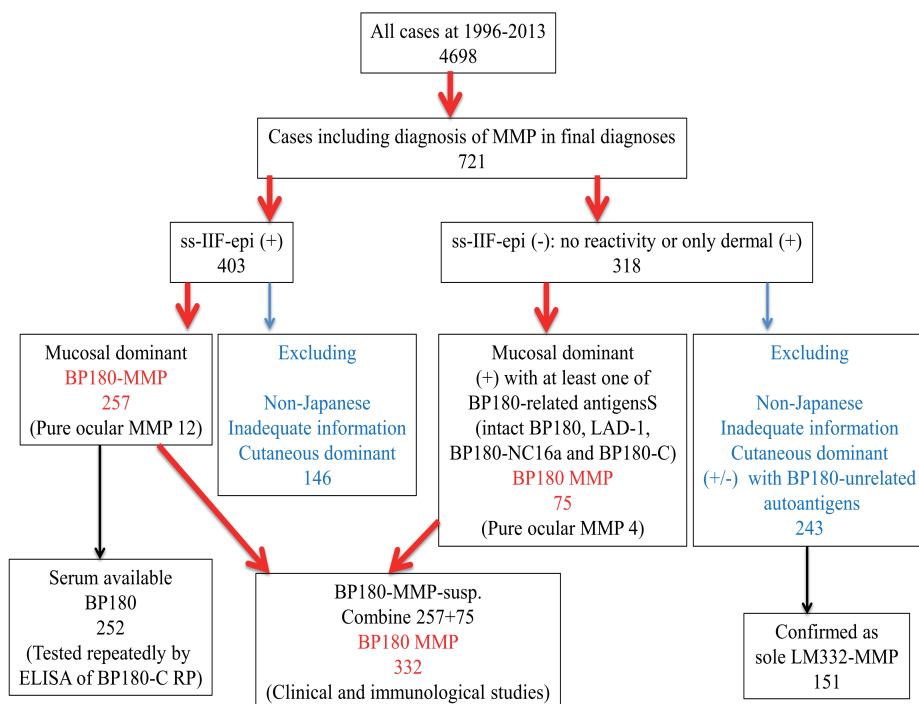


Fig. S2. Algorithm of our tentative diagnostic criteria for anti-BP180-type mucous membrane pemphigoid (MMP) using the cohort of 4,698 patients with autoimmune bullous disease studied at Kurume University. Finally, we combined 257 cases from the ss-IIF-epi(+) group and 75 cases from the ss-IIF-ep(-) group, and diagnosed 332 cases as BP180-MMP. Information for pure ocular MMP and anti-laminin-332 (LM332)-type MMP is also incorporated.

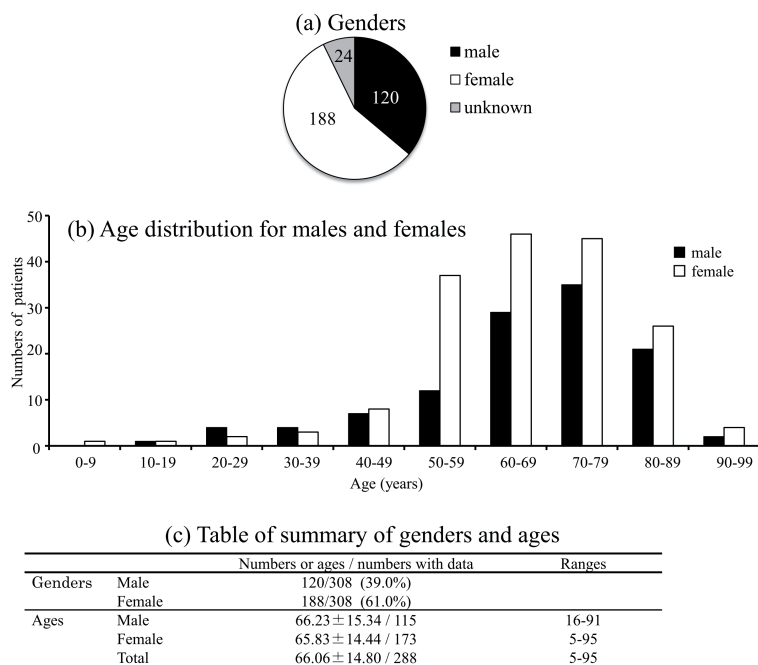


Fig. S3. Summary of the background of the 332 cases of BP180-mucous membrane pemphigoid (MMP). (a) Gender distribution. (b) Age distribution. (c) Summary of gender and age data.

Table SI. Underlying diseases in 332 patients with BP180-mucous membrane pemphigoid (MMP)

Underlying diseases	Patients, <i>n</i>
Hypertension	22
Malignant tumours	21
Malignant lymphomas	3
Prostate cancer	3
Lung cancer	3
Liver cancer	2
Ovarian cancer	2
Pancreatic cancer	2
Metastatic cancer (unknown primary cancers)	2
Thyroid cancer	1
Uterine cancer	1
Oesophageal cancer	1
Gastric cancer	1
Diabetes mellitus	14
Heart diseases	13
Hyperlipidaemia	10
Rheumatoid diseases	7
Hyperuricaemia	7
Hepatitis C	5
Sjögren syndrome	4
Asthma	4
Kidney disease	6
Cerebrovascular disorder	3
Allergic rhinitis	3
Mental disorder	3
Metal allergy	2
Osteoporosis	2
Gastritis	2
Others <sup>a</sup>	One each

<sup>a</sup>Stevens-Johnson syndrome, bronchiolitis obliterans, Parkinson's disease, Ménière's disease, Hashimoto disease, hypothyroidism, collagen disease, allergic conjunctivitis, cataract, glaucoma, hypochromic anaemia, idiopathic thrombocytopenic purpura, alkaptonuria, spinal stenosis, hip osteoarthritis, chronic urticaria, meningioma, stomach ulcer, osteomyelitis of the jaw.

Table SII. Summary of sites of mucocutaneous lesions in 332 patients with BP180-mucous membrane pemphigoid (MMP)

	Patients, <i>n</i>	Restricted to the sites, <i>n</i>
Mucous lesions	332	200
Oral	284	106
Lips	33	0
Gingival	93	26
Buccal	60	5
Palate	60	1
Tongue	23	0
Ocular	97	16
Nasal	11	0
Laryngeal	35	2
Pharyngeal	42	1
Oesophageal	14	0
Genital	32	2
Others	6	0
Skin lesions	132	0
Trunk	98	0
Extremities	94	0

Table SIII. Treatments reported in consultation letters at the time of our serological studies

Treatments	Patients, <i>n</i>	Treated as monotherapy, <i>n</i>
Topical steroids	39	14
Systemic steroids	97	60
Minocycline	25	0
Anti-histamines	9	0
Tetracycline	14	0
Immunosuppressive agents <sup>a</sup>	10	0
Niacinamide	26	0
Diaminodiphenyl sulfone (DDS)	26	1 (not effective)
Macrolides <sup>b</sup>	3	1 (roxithromycin, effective)
Chinese herbal drug <sup>c</sup>	2	0
Steroid pulse therapy	7	0
Intravenous immunoglobulin	3	0
Plasmapheresis	5	0
Before initiation of treatment	35	
Unknown	148	

<sup>a</sup>Immunosuppressive agents: cyclosporine 5, azathioprine 3, mizoribine 1, cyclophosphamide 1. <sup>b</sup>Roxithromycin 1, clarithromycin 1, erythromycin 1. <sup>c</sup>Chinese herbal drug: Tsumura-Saireito.

Table SIV. Results of direct immunofluorescence (IF) in patients with BP180-mucous membrane pemphigoid (MMP)

Sites of deposition	Immunoglobulins and complement C3	+			Unknown
		+	+/-	-	
BMZ	IgG	102	6	24	200
	IgA	43	4	24	261
	IgM	9	5	26	292
	C3	105	5	17	205
Cell surfaces	IgG	6	1	57	268
	IgA	1	0	44	287
	IgM	0	0	26	306
	C3	4	1	53	274

BMZ: basement membrane zone.

Table SV. Results of indirect immunofluorescence (IIF) studies in patients with BP180-mucous membrane pemphigoid (MMP)

		Positive					Total	Negative	Unknown	
		×10	×20	×40	×80	<×160				
Indirect IF of normal human skin										
BMZ	IgG	37	1	42	3	44	5	132	187	13
	IgA	9	1	7	0	5	4	26	259	47
Cell surfaces	IgG	1	0	1	0	5	0	7	187	138
	IgA	0	0	0	0	0	0	0	248	84
ss-IIF										
IgG	Epidermal side	96	2	105			4	207	110	15
	Dermal side	34	0	24			4	62	255	15
IgA	Epidermal side	87	1	56			7	151	131	50
	Dermal side	3	0	1			0	4	278	50

BMZ: basement membrane zone.

Table SVI. Results of immunoblotting (IB) and enzyme-linked immunosorbent assays (ELISA) in all patients with BP180-mucous membrane pemphigoid (MMP)

Methods	Substrates	IgG			IgA		
		+	-	Positive rate	+	-	Positive rate
IB	BP180-NC16a	86	230	27.2%	4	237	1.7%
	BP180-C	118	156	43.1%	17	204	7.7%
	BP180-NC16a and/or BP180-C	159	160	49.8%	21	227	8.5%
ELISA	BP180-NC16a	79	206	27.7%	0	7	0%
	BP180-C	139	187	42.6%	89	237	27.3%
	BP180-NC16a and/or BP180-C	173	158	52.1%	89	237	27.3%

Table SVII. Final serological diagnoses in 332 cases of BP180-mucous membrane pemphigoid (MMP)

Serological diagnoses	<i>n</i>
Sole BP180-MMP (excluding 16 cases of pure ocular MMP)	238
BP180-MMP + LM332-MMP	51
BP180-MMP + LM332-MMP + EBA	1
BP180-MMP + LM332-MMP + PV	2
BP180-MMP + integrin MMP	1
BP180-MMP + EBA	2
BP180-MMP + EBA + PNP	1
BP180-MMP + LM-gamma-1 pemphigoid	1
BP180-MMP + PV	10
BP180-MMP + PF	2
BP180-MMP + PNP	6
BP180-MMP + Stevens-Johnson syndrome	1
Pure ocular MMP	16
Total	332

MMP: mucous membrane pemphigoid; EBA: epidermolysis bullosa acquisita; PV: pemphigus vulgaris; PF: pemphigus foliaceus; PNP: paraneoplastic pemphigus.