

Annual Report of Nation-Wide Survey of Cutaneous Lymphomas in Japan (2021)

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Result 1. Number of newly diagnosed primary cutaneous lymphoma patients in 2021

	Total		Neoplasm category	Male No.	Female No.	M/F	Age		
	No.	%					Median	Average±sd	Range
Total	447		%	235	212	1.11	69	66.0 ± 17.9	98 - 9
Mature T- and NK-cell neoplasms	365	81.7	100	193	172	1.12	68	64.6 ± 17.6	98 - 9
Mycosis fungoides	212	47.4	58.1	115	97	1.19	69	65.9 ± 15.9	98 - 20
Sézary syndrome	8	1.8	2.2	5	3	1.67	75	74.8 ± 8.8	86 - 58
Primary cutaneous CD30+ T-cell lymphoproliferative disorders	65	14.5	17.8	32	33	0.97	63	62.2 ± 19.9	95 - 9
Primary cutaneous anaplastic large cell lymphoma	43	9.6	11.8	20	23	0.87	68	64.6 ± 18.7	95 - 13
Lymphomatoid papulosis	22	4.9	4.9	12	10	1.20	61	57.6 ± 21.3	89 - 9
Adulut T-cell leukemia/lymphoma (smoldering type)	18	4.0	4.9	9	9	1.00	70	71.1 ± 11.2	89 - 48
Hydroa vacciniforme LPD	0	0.0	0.0	0	0	-	-	-	-
Extranodal NK-/T-cell lymphoma, nasal type	8	1.8	2.2	1	7	0.14	52	51.9 ± 21.3	78 - 24
Subcutaneous panniculitis-like T-cell lymphoma	10	2.2	2.7	3	7	0.43	44	42.6 ± 18.0	66 - 21
Primary cutaneous γδ T-cell lymphoma	0	0.0	0.0	0	0	-	-	-	-
Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma	0	0.0	0.0	0	0	-	-	-	-
Primary cutaneous acral CD8+ T-cell lymphoma	2	0.4	0.5	0	2	0.00	45.5	45.5 ± 1.5	47 - 44
Primary cutaneous CD4+ small/medium T-cell lymphoproliferative disorder	7	1.6	1.9	3	4	0.75	63.0	56.9 ± 20.0	80 - 11
Peripheral T-cell lymphoma, NOS (primary cutaneous)	35	7.8	9.6	25	10	2.50	72	67.2 ± 17.7	88 - 26
Mature B-cell neoplasms	74	16.6	100	35	39	0.90	74.5	73.1 ± 17.0	96 - 21
Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue	13	2.9	17.6	4	9	0.44	61	55.8 ± 19.5	83 - 21
Primary cutaneous follicle center lymphoma	17	3.8	23.0	10	7	1.43	74	73.9 ± 11.9	92 - 45
Primary cutaneous DLBCL, leg type	37	8.3	50.0	17	20	0.85	80	79.7 ± 12.8	96 - 42
EBV+ mucocutaneous ulcer	1	0.2	1.4	0	1	0.00	44	-	-
other	6	1.3	8.1	4	2	2.00	73	72.2 ± 17.3	92 - 40
Blastic plasmacytoid dendritic cell neoplasm	8	1.8		7	1	7.0	79	64.7 ± 27.1	94 - 10

Result 2. All patients with primary cutaneous lymphoma registered in 2021

	Total		Neoplasm category	Male No.	Female No.	M/F	Age		
	No.	%					Median	Average±sd	Range
Total	2155		%	1168	987	1.18	70	66.3 ± 16.0	99 - 6
Mature T- and NK-cell neoplasms	1957	90.8	100	1060	897	1.18	69	65.9 ± 15.8	98 - 6
Mycosis fungoides	1433	66.5	73.2	778	655	1.19	69	66.4 ± 15.0	98 - 17
Sézary syndrome	32	1.5	1.6	22	10	2.20	72	69.1 ± 14.9	87 - 31
Primary cutaneous CD30+ T-cell lymphoproliferative disorders	256	11.9	13.1	134	122	1.10	67	63.6 ± 17.4	95 - 9
Primary cutaneous anaplastic large cell lymphoma	159	7.4	8.1	89	70	1.27	70	66.5 ± 16.6	95 - 13
Lymphomatoid papulosis	97	4.5	5.0	45	52	0.87	59	59.0 ± 17.7	91 - 9
Adulut T-cell leukemia/lymphoma (smoldering type)	88	4.1	4.5	47	41	1.15	72	70.9 ± 10.9	92 - 43
Hydroa vacciniforme LPD	1	0.0	0.1	1	0	-	69	69.0	-
Extranodal NK-/T-cell lymphoma, nasal type	15	0.7	0.8	7	8	0.88	63	53.5 ± 23.9	86 - 6
Subcutaneous panniculitis-like T-cell lymphoma	30	1.4	1.5	11	19	0.58	49	48.8 ± 21.0	93 - 19
Primary cutaneous γδ T-cell lymphoma	3	0.1	0.2	2	1	2.00	68	68.3 ± 7.8	78 - 59
Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma	0	0.0	0.0	0	0	-	-	-	-
Primary cutaneous acral CD8+ T-cell lymphoma	2	0.1	0.1	0	2	0.00	45.5	45.5 ± 1.5	47 - 44
Primary cutaneous CD4+ small/medium T-cell lymphoproliferative disorder	22	1.0	1.1	12	10	1.20	66.5	61.2 ± 19.9	89 - 11
Peripheral T-cell lymphoma, NOS (primary cutaneous)	75	3.5	3.8	46	29	1.59	73	67.9 ± 18.2	91 - 14
Mature B-cell neoplasms	186	8.6	100	97	89	1.09	73	70.5 ± 16.5	99 - 21
Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue	60	2.8	32.3	29	31	0.94	63	61.2 ± 17.0	99 - 21
Primary cutaneous follicle center lymphoma	53	2.5	28.5	32	21	1.52	74	73.3 ± 13.3	97 - 40
Primary cutaneous DLBCL, leg type	63	2.9	33.9	31	32	0.97	79	78.3 ± 13.1	97 - 41
EBV+ mucocutaneous ulcer	3	0.1	1.6	1	2	0.50	44	55.3 ± 18.2	81 - 41
other	7	0.3	3.8	4	3	1.33	70	65.6 ± 14.7	84 - 40
Blastic plasmacytoid dendritic cell neoplasm	12	0.6		11	1	11.0	74	67.5 ± 21.1	94 - 10

Result 3. Number of secondary cutaneous lymphoma patients registered in 2021

	Newly-diagnosed		Male/Female		M/F	All registered		Male/Female		M/F
	No.	%	No.	No.		No.	%	No.	No.	
Total	192	100	100/	92	1.09	393	100	220/	173	1.27
Diffuse large B-cell lymphoma	47	24.5	23	24	0.96	101	25.7	57	44	1.30
Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (except primary cutaneous type)	1	0.5	0	1	0.00	15	3.8	9	6	1.50
Follicular lymphoma	13	6.8	5	8	0.63	20	5.1	8	12	0.67
Mantle cell lymphoma	1	0.5	1	0	-	2	0.5	2	0	-
Plasmablastic lymphoma	0	0.0	0	0	-	0	0.0	0	0	-
Intravascular lymphoma	34	17.7	22	12	1.83	40	10.2	25	15	1.67
Adulut T-cell leukemia/lymphoma (except smoldering type)	43	22.4	25	18	1.39	102	26.0	61	41	1.49
Angioimmunoblastic T-cell lymphoma	9	4.7	4	5	0.80	16	4.1	8	8	1.00
Anaplastic large cell lymphoma, ALK-positive	1	0.5	1	0	-	4	1.0	2	2	1.00
Anaplastic large cell lymphoma, ALK-negative	10	5.2	6	4	1.50	16	4.1	10	6	1.67
Peripheral T-cell lymphoma, NOS (except primary cutaneous type)	4	2.1	3	1	3.00	11	2.8	6	5	1.20
Hodgkin lymphomas	2	1.0	2	0	-	9	2.3	8	1	8.00
Other iatrogenic immunodeficiency-associated lymphoproliferative disorders	14	7.3	2	12	0.17	23	5.9	3	20	0.15
Langerhans cell histiocytosis	4	2.1	1	3	0.33	11	2.8	4	7	0.57
Other	9	4.7	5	4	1.25	23	5.9	17	6	2.83