

# Annual Report of Nation-Wide Survey of Cutaneous Lymphomas in Japan (2022)

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## Result 1. Number of newly diagnosed primary cutaneous lymphoma patients in 2022

	Total		Neoplasm category	Male No.	Female No.	M/F	Age		
	No.	%					Median	Average±sd	Range
<b>Total</b>	<b>500</b>		<b>%</b>	<b>284</b>	<b>216</b>	<b>1.31</b>	<b>71</b>	<b>66.7 ± 16.7</b>	<b>97 - 14</b>
<b>Mature T- and NK-cell neoplasms</b>	<b>412</b>	<b>82.4</b>	<b>100</b>	<b>230</b>	<b>182</b>	<b>1.26</b>	<b>69</b>	<b>65.3 ± 16.8</b>	<b>97 - 14</b>
Mycosis fungoides	252	50.4	61.2	142	110	1.29	68	64.9 ± 16.3	93 - 14
Sézary syndrome	8	1.6	1.9	5	3	1.67	80	70.0 ± 17.5	83 - 33
Primary cutaneous CD30+ T-cell lymphoproliferative disorders	50	10.0	12.1	30	20	1.50	62.5	63.2 ± 16.3	94 - 22
Primary cutaneous anaplastic large cell lymphoma	34	6.8	8.3	24	10	2.40	63	66.3 ± 15.8	94 - 38
Lymphomatoid papulosis	16	3.2	3.9	6	10	0.60	62.5	56.4 ± 15.2	78 - 22
Adult T-cell leukemia/lymphoma (smoldering type)	27	5.4	6.6	13	14	0.93	74	73.6 ± 9.0	91 - 57
Hydroa vacciniforme LPD	0	0.0	0.0	0	0	-	-	-	-
Extranodal NK-/T-cell lymphoma, nasal type	18	3.6	4.4	13	5	2.60	70	67.8 ± 14.8	90 - 27
Subcutaneous panniculitis-like T-cell lymphoma	3	0.6	0.7	1	2	0.50	22	36.7 ± 25.8	73 - 15
Primary cutaneous $\gamma\delta$ T-cell lymphoma	0	0.0	0.0	0	0	-	-	-	-
Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma	2	0.4	0.5	0	2	0.00	79	79.0 ± 13.0	92 - 66
Primary cutaneous acral CD8+ T-cell lymphoma	1	0.2	0.2	0	1	0.00	62	-	-
Primary cutaneous CD4+ small/medium T-cell lymphoproliferative disorder	8	1.6	1.9	4	4	1.00	67	62.3 ± 17.9	83 - 34
Peripheral T-cell lymphoma, NOS (primary cutaneous)	43	8.6	10.4	22	21	1.05	67	64.7 ± 19.7	97 - 20
<b>Mature B-cell neoplasms</b>	<b>79</b>	<b>15.8</b>	<b>100</b>	<b>46</b>	<b>33</b>	<b>1.39</b>	<b>77</b>	<b>74.2 ± 13.6</b>	<b>93 - 26</b>
Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue	18	3.6	22.8	11	7	1.57	69.5	65.2 ± 16.4	91 - 26
Primary cutaneous follicle center lymphoma	13	2.6	16.5	8	5	1.60	72	69.8 ± 11.4	87 - 46
Primary cutaneous DLBCL, leg type	35	7.0	44.3	19	16	1.19	82	80.5 ± 10.5	93 - 32
EBV+ mucocutaneous ulcer	2	0.4	2.5	1	1	1.00	68	68.0 ± 8.0	76 - 60
other	11	2.2	13.9	7	4	1.75	74	75.5 ± 9.1	91 - 60
<b>Blastic plasmacytoid dendritic cell neoplasm</b>	<b>9</b>	<b>1.8</b>		<b>8</b>	<b>1</b>	<b>8.00</b>	<b>73</b>	<b>65.4 ± 21.2</b>	<b>91 - 17</b>

## Result 2. All patients with primary cutaneous lymphoma registered in 2022

	Total		Neoplasm category	Male No.	Female No.	M/F	Age		
	No.	%					Median	Average±sd	Range
<b>Total</b>	<b>2486</b>		<b>%</b>	<b>1351</b>	<b>1135</b>	<b>1.19</b>	<b>70</b>	<b>66.3 ± 16.1</b>	<b>98 - 5</b>
<b>Mature T- and NK-cell neoplasms</b>	<b>2246</b>	<b>90.3</b>	<b>100</b>	<b>1209</b>	<b>1037</b>	<b>1.17</b>	<b>69</b>	<b>65.6 ± 16.1</b>	<b>97 - 5</b>
Mycosis fungoides	1640	66.0	73.0	886	754	1.18	70	66.2 ± 15.5	97 - 9
Sézary syndrome	38	1.5	1.7	26	12	2.17	70.5	67.5 ± 15.5	87 - 32
Primary cutaneous CD30+ T-cell lymphoproliferative disorders	267	10.7	11.9	138	129	1.07	63	61.0 ± 17.9	94 - 5
Primary cutaneous anaplastic large cell lymphoma	165	6.6	7.3	93	72	1.29	66	64.4 ± 16.5	94 - 14
Lymphomatoid papulosis	102	4.1	4.1	45	57	0.79	56	55.6 ± 18.7	90 - 5
Adult T-cell leukemia/lymphoma (smoldering type)	115	4.6	5.1	53	62	0.85	73	72.2 ± 9.9	93 - 45
Hydroa vacciniforme LPD	2	0.1	0.1	1	1	1.00	43.5	43.5 ± 26.5	70 - 17
Extranodal NK-/T-cell lymphoma, nasal type	28	1.1	1.2	19	9	2.11	69.5	64.7 ± 19.6	90 - 7
Subcutaneous panniculitis-like T-cell lymphoma	22	0.9	1.0	7	15	0.47	50	49.5 ± 21.1	82 - 15
Primary cutaneous $\gamma\delta$ T-cell lymphoma	2	0.1	0.1	0	2	0.00	74	74.0 ± 7.0	81 - 67
Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma	3	0.1	0.1	1	2	0.50	82	80.0 ± 10.7	92 - 66
Primary cutaneous acral CD8+ T-cell lymphoma	2	0.1	0.1	1	1	1.00	68.5	68.5 ± 6.5	75 - 62
Primary cutaneous CD4+ small/medium T-cell lymphoproliferative disorder	22	0.9	1.0	10	12	0.83	61.5	57.9 ± 19.9	83 - 12
Peripheral T-cell lymphoma, NOS (primary cutaneous)	105	4.2	4.7	67	38	1.76	71	65.6 ± 18.7	97 - 15
<b>Mature B-cell neoplasms</b>	<b>228</b>	<b>9.2</b>	<b>100</b>	<b>132</b>	<b>96</b>	<b>1.38</b>	<b>75</b>	<b>72.7 ± 14.3</b>	<b>98 - 21</b>
Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue	81	3.3	35.5	43	38	1.13	66.0	65.1 ± 15.6	91 - 21
Primary cutaneous follicle center lymphoma	54	2.2	23.7	39	15	2.60	73.0	74.1 ± 11.7	98 - 41
Primary cutaneous DLBCL, leg type	71	2.9	31.1	37	34	1.09	82.0	80.1 ± 10.8	97 - 32
EBV+ mucocutaneous ulcer	4	0.2	1.8	2	2	1.00	68.0	65.8 ± 13.8	91 - 60
other	18	0.7	7.9	11	7	1.57	75.0	74.9 ± 8.1	91 - 60
<b>Blastic plasmacytoid dendritic cell neoplasm</b>	<b>12</b>	<b>0.5</b>		<b>10</b>	<b>2</b>	<b>5.00</b>	<b>75</b>	<b>68.4 ± 19.1</b>	<b>91 - 17</b>

## Result 3. Number of secondary cutaneous lymphoma patients registered in 2022

	Newly-diagnosed		Male/Female		M/F	All registerd		Male/Female		M/F
	No.	%	No.	No.		No.	%	No.	No.	
<b>Total</b>	<b>215</b>	<b>100</b>	<b>111</b>	<b>104</b>	<b>1.07</b>	<b>406</b>	<b>100</b>	<b>225</b>	<b>181</b>	<b>1.24</b>
Diffuse large B-cell lymphoma	72	33.5	36	36	1.00	113	27.8	62	51	1.22
Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (except primary cutaneous type)	3	1.4	2	1	2.00	9	2.2	5	4	1.25
Follicular lymphoma	13	6.0	5	8	0.63	18	4.4	8	10	0.80
Mantle cell lymphoma	2	0.9	1	1	1.00	2	0.5	1	1	1.00
Plasmablastic lymphoma	1	0.5	1	0	-	2	0.5	2	0	-
Intravascular lymphoma	43	20.0	23	20	1.15	47	11.6	26	21	1.24
Adult T-cell leukemia/lymphoma (except smoldering type)	36	16.7	17	19	0.89	96	23.6	56	40	1.40
Angioimmunoblastic T-cell lymphoma	6	2.8	3	3	1.00	11	2.7	7	4	1.75
Anaplastic large cell lymphoma, ALK-positive	4	1.9	1	3	0.33	5	1.2	1	4	0.25
Anaplastic large cell lymphoma, ALK-negative	8	3.7	6	2	3.00	24	5.9	16	8	2.00
Peripheral T-cell lymphoma, NOS (except primary cutaneous type)	5	2.3	3	2	1.50	14	3.4	6	8	0.75
Hodgkin lymphomas	1	0.5	1	0	-	4	1.0	3	1	3.00
Other iatrogenic immunodeficiency-associated lymphoproliferative disorders	7	3.3	5	2	2.50	18	4.4	8	10	0.80
Langerhans cell histiocytosis	2	0.9	0	2	0.00	13	3.2	4	9	0.44
Other	12	5.6	7	5	1.40	30	7.4	20	10	2.00

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