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Supplementary

Supporting information appendix

ICD and SNOMED codes used to identify cohort of mastocytosis patients

Diagnosis	ICD-9	ICD-10	SNOMED
Cutaneous mastocytosis		Q822	97401
- Urticaria pigmentosa		Q822A	
- DCM		Q822B	
- Mastocytoma	238F	Q822C	
- TMEP		Q822D	
- Mastocytosis unspec		Q822X	
Systemic mastocytosis		Q822	97411, 97413
Malignant mastocytosis	202G		
Mast cell leukemia		C943	97423
Malignant mast cell tumor, Aggressive SM, SMAHN		C962	97413
Mast cell tumor of unknown nature, ISM		D470	97411
Mast cell sarcoma		C962	97403

Registries used for our dataset

Register	Data content
Swedish Cancer Register	Notification of cancer diagnosis, site and date, according to SNOMED.

Patient Register	In- and outpatient Registers, diagnosis according to ICD, revision 7-10.
Cause of Death Register	Date and underlying/contributing causes of death, according to ICD-codes.
Register of Total Population	Data of the composition, family relationships and identities of the Swedish population.
Longitudinal database on socioeconomic factors (LISA)	Socioeconomic factors, including income, civil status, type of employment and income

Supplementary tables

Table 1. Demographics, all subjects between index year of 2001-2018

	All subjects	
All subjects, n /%	4438	100
Data sources	n	%
		% of all subjects in NPR
NPR, outpatient care, ICD-codes	4292	outpatient
<i>C943</i>	13	0.3
<i>C962</i>	96	2.2
<i>D470</i>	405	9.4
<i>Q822</i>	1059	24.6
<i>Q822A</i>	1313	30.5
<i>Q822B</i>	57	1.3
<i>Q822C</i>	977	22.7
<i>Q822D</i>	99	2.3
<i>Q822X</i>	273	6.3
		% of all subjects in NPR
NPR, Inpatient care, ICD-codes	390	inpatient
<i>238F</i>	1	0.0
<i>C943</i>	6	0.7
<i>C962</i>	42	10.7
<i>D470</i>	79	20.2
<i>Q822</i>	137	35.1
<i>Q822A</i>	59	15.1
<i>Q822B</i>	32	8.2
<i>Q822C</i>	4	0.1
<i>Q822D</i>	3	0.1
<i>Q822X</i>	27	6.9
		% of all subjects in SCR
Cancer registry, SNOMED-codes	186	
<i>97401</i>	26	13.9
<i>97403</i>	2	1.0

<i>97411</i>	105	56.4
<i>97413</i>	50	26.8
<i>97423</i>	3	1.6
<hr/>		
Sex		
<hr/>		
Male	2245	50.6
Female	2193	49.4
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Age at index date		
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Median/IQR	12	1-47
Mean/sd	25	26.6
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Age group	n	%
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0-1 (infants)	1237	27.9
2-9	923	20.8
10-19	238	5.4
20-29	284	6.4
30-39	328	7.4
40-49	388	8.7
50-59	365	8.2
60-69	345	7.8
70-79	227	5.1
80-89	91	2.1
90+	12	0.3

Table 2. Distribution of subtypes by register source in individuals (≥ 20 years) with a mastocytosis diagnosis

Data source	n	%
NPR, outpatient care, ICD-codes	1912	% of all subjects in NPR outpatient
<i>C943</i>	9	0.5
<i>C962</i>	61	3.1
<i>D470</i>	296	15.4
<i>Q822</i>	447	23.3
<i>Q822A</i>	804	42.0
<i>Q822B</i>	28	1.5
<i>Q822C</i>	56	2.9
<i>Q822D</i>	88	4.6
<i>Q822X</i>	123	6.4
NPR, Inpatient care, ICD-codes	330	% of all subjects in NPR inpatient
<i>238F</i>	1	0.0
<i>C943</i>	3	0.1
<i>C962</i>	39	11.8
<i>D470</i>	71	21.5
<i>Q822</i>	108	32.7
<i>Q822A</i>	49	14.8
<i>Q822B</i>	31	9.3
<i>Q822C</i>	1	0.0
<i>Q822D</i>	3	0.1
<i>Q822X</i>	24	7.3
Cancer Register, SNOMED codes	175	% of all subjects in SCR
<i>97401</i>	19	10.8
<i>97403</i>	2	1.1
<i>97411</i>	101	57.7
<i>97413</i>	50	28.6
<i>97423</i>	3	1.7

Table 3. CCI and comorbidities by subtype

	Advanced n=159	Benign n=1110	Mixed n=771	p-value
Charlson Comorbidity Index (CCI)				<0.001
0	101 (63.5)	960 (86.5)	562 (72.9)	
1	10 (6.3)	71 (6.4)	86 (11.2)	
2+	48 (30.2)	79 (7.1)	123 (16.0)	
Morbidities within CCI				
Myocardial Infarction	4 (2.5)	14 (1.3)	16 (2.1)	0.273
Congestive Heart Failure	3 (1.9)	12 (1.1)	19 (2.5)	0.068
Peripheral Vascular Disease	3 (1.9)	6 (0.5)	13 (1.7)	0.036
Cerebrovascular Disease	2 (1.3)	20 (1.8)	42 (5.4)	<0.001
Dementia	0 (0.0)	2 (0.2)	4 (0.5)	0.319
Chronic Pulmonary Disease	5 (3.1)	25 (2.3)	39 (5.1)	0.004
Connective Tissue Disease-Rheumatic Disease	2 (1.3)	5 (0.5)	5 (0.6)	0.443
Peptic Ulcer Disease	1 (0.6)	6 (0.5)	9 (1.2)	0.309
Mild Liver Disease	0 (0.0)	6 (0.5)	5 (0.6)	0.596
Diabetes without complications	6 (3.8)	31 (2.8)	39 (5.1)	0.039
Diabetes with complications	1 (0.6)	3 (0.3)	8 (1.0)	0.101
Paraplegia and Hemiplegia	0 (0.0)	3 (0.3)	17 (2.2)	<0.001
Renal Disease	4 (2.5)	3 (0.3)	14 (1.8)	0.001
Cancer	38 (23.9)	55 (5.0)	80 (10.4)	<0.001
Moderate or Severe Liver Disease	0 (0.0)	0 (0.0)	1 (0.1)	0.439
Metastatic Carcinoma	9 (5.7)	5 (0.5)	8 (1.0)	<0.001
AIDS/HIV	0 (0.0)	3 (0.3)	0 (0.0)	0.284

Figure 1. Overall survival by mastocytosis subtype with their matched comparators (controls).

