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Rare Lymphomas – Epidemiology from the Munich Cancer Registry

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Introduction

Rare cancers are defined as cancers with an incidence of less than 6/100,000. In total they represent 22% of the new cancer cases. Lymphomas in total are not rare, but they fragment into many different entities which therefore count as rare. The population-based data herein is presented to weigh in on the results of randomised clinical trials and to clarify expectations in the treatment of sporadic diagnoses.

Figure 1. Catchment area of the Munich Cancer Registry

Methods

Nine different rare lymphoma entities registered in the catchment area of the Munich Cancer Registry (MCR) 1998-2016 were selected for presentation: lymphocyte predominant Hodgkin lymphoma, hairy cell leukaemia, Mantle cell lymphoma, Burkitt's lymphoma, MALT lymphoma, M. Waldenström, primary lymphoma of the central nervous system, cutaneous T-cell lymphoma, and peripheral mature T-cell lymphoma. Age-standardised incidences (European standard population), distributions of patient and tumour characteristics, and survival were analysed per entity, sex, and period of initial diagnosis. The overall (or observed) survival was calculated by the Kaplan-Meier method. Relative survival as the quotient of overall and expected survival was calculated as an estimate for cancer specific survival.

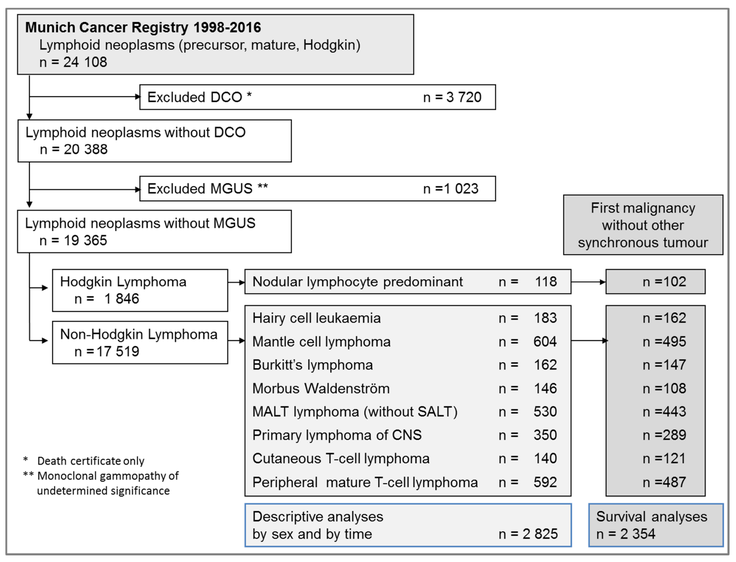

Figure 2. Study flow chart

Results

2825 patients out of 19,365 Lymphoma patients were diagnosed with the analysed rare lymphomas, accounting for 14.6% (figure 2).

Incidence

The incidences (ASR ES) ranged between 0.7/100,000 in peripheral mature T-cell lymphoma, 0.6/100,000 in Mantle cell lymphoma and 0.15/100,000 in lymphocyte predominant Hodgkin lymphoma. The male incidence was more than twice that of female incidence in most entities apart from M. Waldenström, MALT lymphoma and primary lymphomas of the central nervous system (table 1).

Patient and tumour characteristics

Men account for 62.5% of all with a wide range: in hairy cell leukaemia 81.4% and in MALT lymphoma only 48.1% were male. The median age at initial diagnosis differs between 44 years in patients with nodular lymphocyte predominant Hodgkin disease and 68 years in those with M. Waldenström (table 1). In total women where significantly older than men ($p<0.0001$) with 64.8 years (±17.5) vs. 61.5 (±16.5) on average. Over time men became significantly older at initial diagnosis with 58.8 years in 1998-2002 and 64.6 in 2013-2016 ($p<0.0001$).

Survival

The 5-year relative survival differs between 35.6% in primary CNS lymphoma and 94.7% in hairy cell leukaemia (94.2% in lymphocyte predominant Hodgkin lymphoma, 61.3% in Mantle cell lymphoma, 64.9% in Burkitt's lymphoma, 89.8% in MALT lymphoma, 82.3% in M. Waldenström, 70.7% in cutaneous T-cell lymphoma, and 42.4% in peripheral mature T-cell lymphoma) (figure 3). Regarding all rare lymphomas, a noteworthy improvement in survival over time cannot be seen (figure 4).

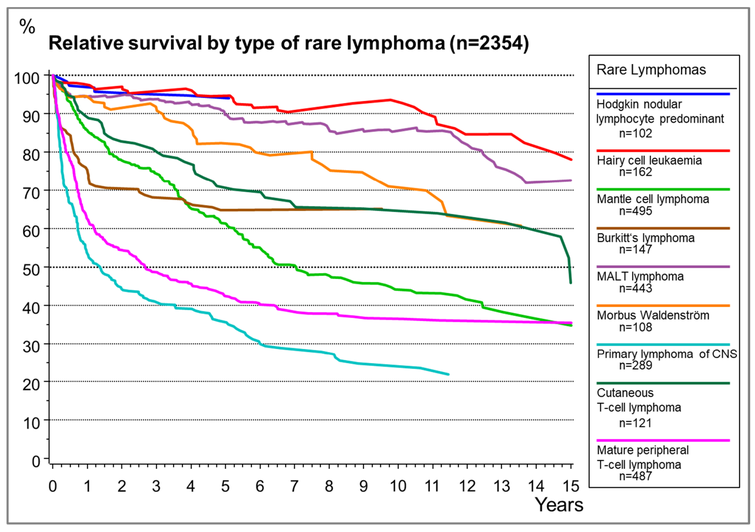

Figure 3. Relative survival by type of rare lymphoma

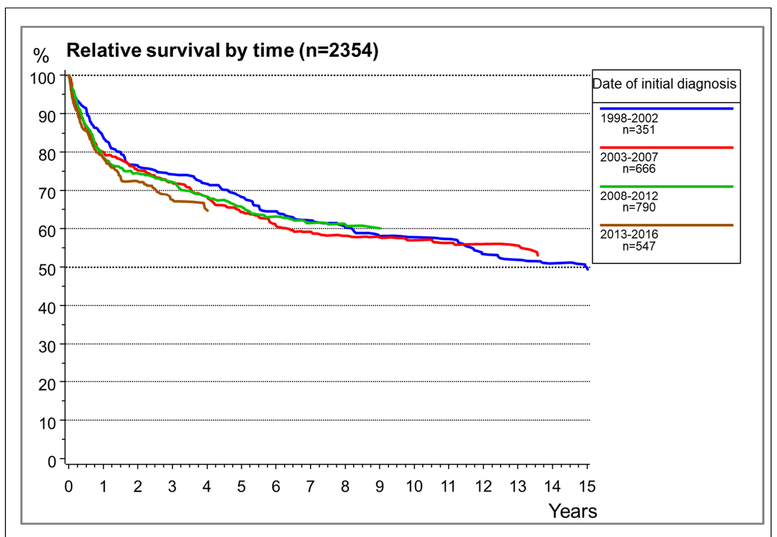

Figure 4. All rare lymphomas: relative survival by time of initial diagnosis

Merely in Mantle cell lymphoma, long-term survival, beginning at four years after initial diagnosis had improved with a 10-year relative survival of 31.1% in 1998-2002 and 46.4% 2003-2007 (figure 5).

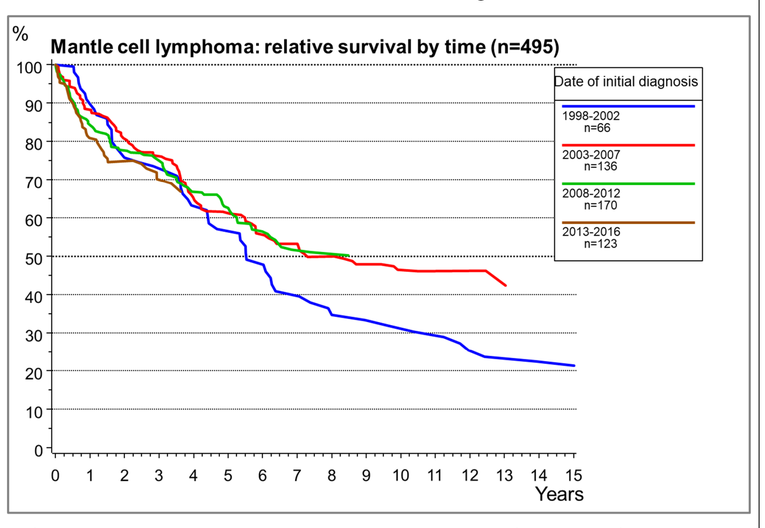

Figure 5. Mantle cell lymphoma: relative survival by time of initial diagnosis

In total, men have a better prognosis than women with a 5-year relative survival of 66.6% vs. 62.7% and a 10-year relative survival of 60.4% vs. 53.2% (figure 6). In MALT lymphomas and primary lymphomas of the CNS, differences by sex could not be seen. Only in cutaneous T-cell lymphomas, women have a better survival than men (5-year relative survival 79.3% vs. 66.3%).

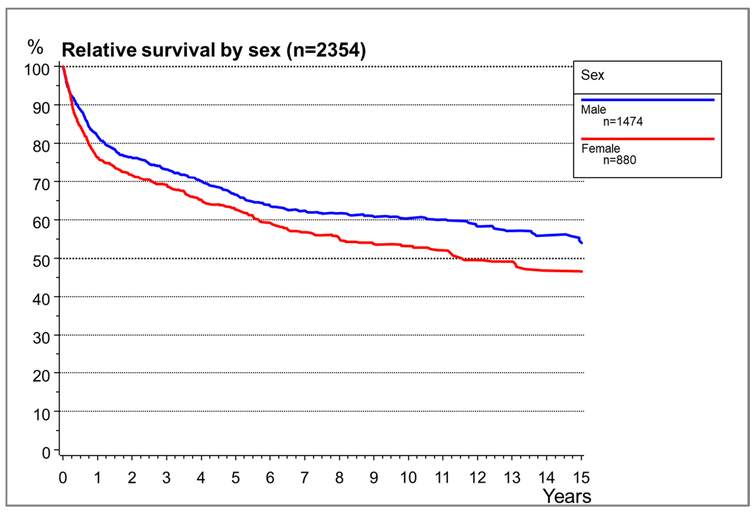

Figure 6. All rare lymphomas: relative survival by sex

Table 1. Incidences, age at diagnosis and death, 5-year relative survival by type of rare lymphoma

Rare Lymphomas	N 1998-2016	Proportion of all documented lymphoma * %	Incidence male ASR (ES)	Incidence female ASR (ES)	Incidence total ASR (ES)	Pro-portion male %	Age at initial diagnosis Mean ± SD	Age at lymphoma-caused death Mean ± SD	5-year relative survival %
HL nodular lymphocyte predominant	118	0.61	0.21	0.08	0.15	72.9	43.9 ± 20.7	68.2 ± 18.2	94.2
Hairy cell leukaemia	183	0.95	0.41	0.08	0.23	81.4	62.0 ± 12.8	74.2 ± 12.6	94.7
Mantle cell lymphoma	604	3.12	0.93	0.31	0.60	72.2	67.9 ± 11.4	72.8 ± 10.1	61.3
Burkitt lymphoma	162	0.84	0.33	0.13	0.23	72.2	43.9 ± 25.5	59.8 ± 22.4	64.9
Morbus Waldenström	146	0.75	0.19	0.13	0.16	56.9	68.4 ± 9.9	74.9 ± 10.6	82.3
MALT lymphoma	530	2.74	0.59	0.52	0.54	48.1	65.7 ± 14.0	76.0 ± 11.4	89.8
Primary lymphoma of CNS	350	1.81	0.39	0.29	0.34	52.0	65.4 ± 12.7	68.8 ± 11.4	35.6
Cutaneous T-cell lymphoma	140	0.72	0.26	0.11	0.18	67.9	55.6 ± 20.8	70.1 ± 15.3	70.7
Peripheral mature T-cell lymphoma	592	3.06	0.99	0.50	0.72	61.2	62.7 ± 18.7	69.6 ± 14.1	42.4
Total	2825	14.59	4.30	2.15	3.15	62.5	62.7 ± 17.2	70.8 ± 13.3	65.2

* N=19,365 without Death certificate only-cases and monoclonal gammopathy of undetermined significance (MGUS)
SD Standard deviation ASR Age standardised rate ES European standard population

Conclusions

Incidence, age, and outcome in rare lymphomas vary extremely.

Conclusive survival improvements over time can be seen in Mantle cell lymphoma. In total, survival improvements between 1998 and 2016 were not seen in a population-based setting. Maybe, newer chemotherapy regimen, targeted therapies and stem cell transplantation have not found their way into broad application or cannot be applicated so easily in all other patient groups beyond randomized clinical trials.

The treatment of rare diseases such as these lymphomas should be expanded by the knowledge of "real-life" population-based data to adjust and deliver the patients' expectations.

The authors have declared no conflicts of interest.

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