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Descriptive epidemiology of ophthalmic and ocular adnexal non-Hodgkin's lymphoma

Roxana Moslehi[1,2,†], **F Bruce Coles**[1,3], and **Maria J Schymura**[1,4]

[1]Department of Epidemiology and Biostatistics, School of Public Health, State University of New York at Albany, NY, USA

[2]Cancer Research Center, State University of New York at Albany, NY, USA

[3]Epidemiology Research Institute, Wadsworth Center, New York State Department of Health, NY, USA

[4]New York State Cancer Registry, New York State Department of Health, NY, USA

Abstract

In this article, we provide an update on incidence rates of ophthalmic non-Hodgkin's lymphoma (NHL), in the context of other NHL, in the USA. We also provide population-based estimates of incidence and survival for ocular adnexal NHL, the most common form of ophthalmic NHL, for which descriptive patterns have not been previously reported. Ophthalmic and ocular adnexal NHL have unique incidence patterns, including equal rates among both genders, predominance among Asians/Pacific Islanders, and steady and rapid increases in the past few decades. Studies of international variations in the incidence of ocular adnexal NHL may provide clues as to the underlying mechanisms influencing its unique epidemiology.

Keywords

incidence and survival patterns of non-Hodgkin's lymphoma (NHL); ocular adnexal NHL; ophthalmic NHL

Ophthalmic malignancy most often presents as non-Hodgkin's lymphoma (NHL). First described in 1952, primary ophthalmic lymphoma represents 1–2% of all NHL and 5–15% of all extranodal NHL [1,2]. The two main categories of primary ophthalmic NHL are intra-ocular and ocular adnexal. Intraocular NHL can arise in the cornea, retina, choroid or ciliary body [3], and can be considered a subset of CNS lymphomas [3,4]. Ocular adnexal NHL can arise from the orbit, conjunctiva, lacrimal gland or eyelid [5]. Extranodal marginal zone B-cell lymphoma (EMZL) of mucosa-associated lymphoid tissue (MALT) type is the most common histologic type of primary ophthalmic NHL [6], accounting for 38–100% of ocular adnexal NHL [7–16]. MALT is considered the third most common form of NHL [17], accounting for 6–7% of all non-ophthalmic extranodal NHL cases [18–21]. The incidence of

[†]Author for correspondence: Tel.: +1 518 591 7225, Fax: +1 518 591 7201, rmoslehi@uamail.albany.edu.

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MALT lymphoma at all sites has been reported as increasing steadily over the last two decades since its discovery in the 1980s [22].

Descriptive trends of ophthalmic NHL are sparse in the literature, as are population-based rates and trends of ocular adnexal NHL. A 2006 study of population-based incidence data for 1992–2001 from 12 USA cancer registries participating in the Surveillance Epidemiology and End Results (SEER) Program found equal rates of ophthalmic NHL among both genders, as well as a higher incidence among Asians/Pacific Islanders based on 27 cases of intraocular and 831 cases of ocular adnexal NHL; descriptive patterns of ocular adnexal NHL were not examined separately in that study [23]. This previous study also reported long-term trends for white individuals based on data from nine SEER regions for the period 1975–2001; ophthalmic NHL rates increased rapidly at 6.2 and 6.5% per year among males and females, respectively, with no evidence of peaking, while during the same time period the rates for other extranodal NHLs increased less rapidly and reached a plateau among males in the mid-1990s [23].

The purpose of this article is to provide an update of descriptive patterns of ophthalmic NHL and to provide population-based estimates of rates for its most common form, namely ocular adnexal NHL, for which descriptive patterns have not been previously reported and may provide clues into its etiology.

Methods

We used population-based incidence data from the National Cancer Institute's SEER Program [101,102] to calculate incidence and survival rates of ophthalmic and ocular adnexal NHL and compare them with rates for other extranodal and nodal NHLs. Specifically, data of 1992–2007 and 1999–2006 from 13 SEER areas were used to estimate incidence and survival rates, respectively, for ophthalmic and ocular adnexal NHL. New York State (NYS) rates are not reported to SEER; therefore, we analyzed NYS rates separately using incidence data for 1998–2007 reported to the NYS Cancer Registry (NYSCR) [103].

International Classification of Diseases for Oncology, Third Edition (ICD-O-3) SEER site recodes 33041 and 33042 were used for nodal and extranodal NHL, respectively [104]. In conjunction with the appropriate histology codes, ICD-O-3 topography codes C690–C699 were used for primary ophthalmic NHL; topography codes C690 (conjunctival), C695 (lacrimal gland), C696 (orbit) and C698 (overlapping lesions of eye and adnexa) and C699 (i.e., tumors not otherwise specified [NOS]) were used for ocular adnexal NHL. NOS tumors were included with adnexal tumors, since adnexal tumors comprise over 95% of ophthalmic NHL tumors of known topography [104]. The percentage of ophthalmic NHL tumors coded to NOS was 3.9% in the SEER 13 1992–2007 data and 4.7% in the NYS data for 1998–2007.

All rates were calculated per 100,000 person–years and were age adjusted using the 2000 US standard population. The 5-year observed and relative survival rates were calculated for cases diagnosed 1999–2006 and followed through 2007. Observed survival reflects all mortality in a cohort; it is an estimate of the probability of surviving all causes of death. The relative survival rate represents the likelihood that a patient will not die from their cancer. Relative survival is defined as the ratio of the proportion of observed survivors (all causes of death) in a cohort of cancer patients to the proportion of expected survivors in a comparable cohort of cancer-free individuals. Because the latter is generally not available, population life tables are used to estimate expected survival. Incidence rates and survival were calculated using SEER*Stat [105].

Results

During 1992–2007 in the 13 SEER areas, there were 74,359 cases of nodal NHL and 36,886 cases of extranodal NHL, including 1604 cases of primary ophthalmic and 1565 cases of ocular adnexal NHL. Male and female rates for ophthalmic and ocular adnexal NHL among all races combined were nearly equal for the entire period of 1992–2007, in contrast to non-ophthalmic extranodal and nodal NHLs, which showed predominance among males for all races (data not shown). Analysis of NYSCR rates also revealed equal gender distribution for primary ophthalmic and ocular adnexal NHL among all races combined during 1998–2007 in contrast to other extranodal (non-ophthalmic) and nodal NHL (Table 1). Analysis of SEER rates for 1992–2007 (data not shown) and NYSCR rates for 1998–2007 by ethnicity revealed higher rates of primary ophthalmic and ocular adnexal NHL among Asians/Pacific Islanders in contrast to other extranodal (non-ophthalmic) and nodal NHL sites, which were more frequent among whites (Table 1).

Based on cases diagnosed during the 1999–2006 period in 13 SEER areas and followed through to 2007, the 5-year observed and relative survival rates were calculated as 75.5 and 88.4%, respectively, for ophthalmic NHL, and 76.6 and 89.7%, respectively, for ocular adnexal NHL. Both measures of survival were found to be higher for ophthalmic and ocular adnexal NHL compared with other extranodal (non-ophthalmic) and nodal NHL among both genders and among all races combined (Table 2), as well as among white individuals only (data not provided).

Discussion

In this article, we have examined the incidence and survival rates of ophthalmic NHL in the USA; and we also report the descriptive patterns of ocular adnexal NHL, the most common manifestation of ophthalmic NHL, for which rates have not been previously reported. Our findings suggest that ophthalmic and ocular adnexal NHL have unique descriptive patterns with equal incidence among both genders and higher rates among Asians/Pacific Islanders, in contrast to other extranodal (non-ophthalmic) NHLs, which show predominance among males and white individuals. The only previous relevant population-based analysis in the USA pertained to ophthalmic NHL and reported equal rates among both genders as well as predominance among Asians/Pacific Islanders [23].

Population-based survival rates for ophthalmic and ocular adnexal NHL had not been previously reported. The significantly better survival of ophthalmic and ocular adnexal NHL, in comparison to nodal and other extranodal NHL, seen in our analysis may be related to the etiology and/or the more accessible location or histological characteristics of these conditions.

Temporal trends of ophthalmic NHL also show unique patterns that differ from trends of other extranodal (non-ophthalmic) and nodal NHL. While rates of ophthalmic NHL among white males increased in the 1970s and 1980s, non-ophthalmic extranodal NHL rates showed rapid increases nearly a decade after and peaked among males in the 1990s [23]. The AIDS epidemic is believed to account for a portion of the increase in NHL rates among both genders in most countries [24–26]. The distinctive patterns of ophthalmic NHL may be related to several factors, including diagnostic improvements and changes in classification systems of NHL, as well as causative factors involved in the etiology of this malignancy.

Changes in the classification system of lymphomas are particularly relevant to ophthalmic malignancy; until recognition and widespread use of the MALT code in the 1990s, ophthalmic involvements were often classified as benign tumors. To evaluate the impact of changes in classification systems on the incidence patterns of ophthalmic NHL, the previous

analysis of population-based data compared incidence trends of ophthalmic NHL with those of NHL at three other extranodal sites involving MALT lymphomas [23]. The results suggested that the incidence of ophthalmic NHL rose more rapidly than did NHL at other MALT-related sites during time periods before and after inclusion of MALT code in SEER, even though the percentages that were MALT were similar across sites [23]. These findings suggested that the shift from borderline to MALT may have accounted for part of the increase in incidence, but the major upward trend in the incidence of ophthalmic NHL was real and possibly related to other factors, including those involved in its etiology.

No major causative factor for ocular adnexal NHL has been identified; however, several pathogens, autoimmune processes and genetic factors have been implicated in its etiology (reviewed in [27]). There is much debate in the literature with respect to the possible causative role of pathogens in predisposition to ocular adnexal NHL (reviewed in [27]). Pathogenic infections potentially relevant to the descriptive patterns of ocular adnexal NHL involve two different species of *Chlamydia*, namely *Chlamydia trachomatis* and *Chlamydia psittaci. C. trachomatis* has proven relevance to ophthalmic involvement in the form of conjunctivitis in babies born to infected mothers [28]; this organism has also been associated with chronic conjunctivitis in adolescents and adults as a result of exposure to genital secretions, and from autoinoculation [106]. Conjunctivitis has also been noted as a symptom of *C. psittaci* infection [29]. Recently, several studies have investigated the association between *C. psittaci* and ocular adnexal NHL; while some have reported positive associations [30–35], others have failed to provide evidence of any causative role for this pathogen in ocular adnexal malignancy [7,36–41]. Several factors, such as variations in prevalence of infectious agents, variations in prevalence of other environmental modifiers of risk, and differences in genetics or other host factors, may explain the contradictory results between studies from different geographic areas and populations with respect to the potential role of *Chlamydia* and other bacterial or viral agents in the etiology of ocular adnexal NHL. Further investigations into international variations of ocular adnexal NHL rates and trends, as well as the potential underlying roles of various mechanisms in determining the distinctive descriptive patterns of this malignancy, may help shed light on factors involved in its causation.

Expert commentary

The unique incidence patterns of ophthalmic and ocular adnexal NHL cannot yet be attributed to specific underlying factors and/or mechanisms; these distinctive patterns may be partly due to changes in classification systems with the inclusion of MALT lymphomas, and partly due to the multifactorial nature of this complex condition, with a multitude of extrinsic (environmental, including infectious) and intrinsic (genetic, epigenetic and immunologic) factors playing synergistic roles in its etiology. A major etiologic risk factor for ophthalmic NHL has not yet been identified and there is equivocal evidence for the involvement of infectious agents in the etiology of ocular adnexal NHL. Clues to the etiology and underlying mechanisms driving the incidence patterns of this complex disorder may come from comprehensive descriptive investigations, including observation of international variations in rates and trends in different geographic areas and among different ethnic/racial groups.

Five-year view

Descriptive studies examining international variations in ophthalmic and ocular adnexal NHL rates and trends may be hypothesis-generating and are complementary to etiologic studies. Future multicenter comprehensive studies of ocular adnexal NHL, conducted in the context of other extranodal NHL, should ideally incorporate such aspects as descriptive

epidemiology of the disease and putative pathogens, demographic and family history characteristics of subjects, information on environmental and occupational exposures obtained through validated epidemiologic questionnaires, and biological samples for appropriate molecular studies in order to gain clues into the etiology of this malignancy.

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Key issues

- Ophthalmic and ocular adnexal non-Hodgkin's lymphomas (NHLs) depict unique gender and racial/ethnic distributions compared with other extranodal (non-ophthalmic) and nodal NHLs.
- Rates of ophthalmic and ocular adnexal NHL were found to be equal between both genders and showed predominance among Asians/Pacific Islanders, in contrast to non-ophthalmic extranodal and nodal NHL, which showed predominance among males and whites.
- Underlying etiologic factors responsible for the distinctive incidence patterns of ophthalmic and ocular adnexal NHL are not well understood.
- Information on international variations in rates and trends of ophthalmic and ocular adnexal NHL may provide clues as to the etiology of these complex conditions.
- Future comprehensive etiologic and descriptive studies incorporating epidemiology of the disease and putative pathogens, demographic and family history characteristics of subjects, information regarding environmental and occupational exposures, and biomarker distributions are needed.

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Table 1

Incidence of ophthalmic, other extranodal and nodal non-Hodgkin's lymphoma by gender in New York State, USA (1998–2007).

Type of NHL	Male count†	Male rate†	Female count	Female rate†	Male/female (rate ratio)	Ethnicity rate‡ (ratio, males)	Ethnicity rate‡ (ratio, females)
Ophthalmic NHL							
All races	245	0.29	329	0.29	1.00		
White	207	0.30	264	0.29	1.03	1.00	1.00
Black	20	0.16	43	0.25	0.64	0.53	0.86
American–Indian/Alaska native	0		0				
Asian/Pacific Islander	15	0.37	17	0.30	1.23	1.23	1.03
Ocular adnexal NHL							
All races	236	0.28	317	0.28	1.00		
White	198	0.29	253	0.28	1.04	1.00	1.00
Black	20	0.16	42	0.25	0.64	0.55	0.89
American–Indian/Alaska native	0		0				
Asian/Pacific Islander	15	0.37	17	0.30	1.23	1.28	1.07
Non-ophthalmic extranodal NHL							
All races	6658	7.72	6254	5.56	1.39		
White	5545	7.92	5203	5.71	1.39	1.00	1.00
Black	780	6.18	748	4.33	1.43	0.78	0.76
American–Indian/Alaska native	4	0.94	7	1.28	0.73	0.12	0.22
Asian/Pacific Islander	199	4.40	181	3.43	1.28	0.56	0.60
Nodal NHL							
All races	14,355	16.59	12,872	11.39	1.46		
White	12,276	17.46	11,190	12.27	1.42	1.00	1.00
Black	1515	12.20	1272	7.46	1.64	0.70	0.61
American–Indian/Alaska native	10	2.10	18	4.00	0.53	0.12	0.33
Asian/Pacific Islander	388	8.56	264	5.30	1.62	0.49	0.43

† Rate is per 100,000 person-years age-adjusted using the 2000 USA standard population.

‡ White individuals used as the reference group for ethnicity rate ratio calculations.

NHL: Non-Hodgkin's lymphoma.

Table 2

ved and relative 5-year survival of ophthalmic, other extranodal and nodal non-Hodgkin's lymphoma for all races, 1999–2006[†], Surveillance, emiology, and End Results 13.

e of NHL	Male and female				Male				Female			
	Observed survival (%)	*SE (%)*	*95% LCI (%)*	*95% UCI (%)*	*Observed survival (%)*	*SE (%)*	*95% LCI (%)*	*95% UCI (%)*	*Observed survival (%)*	*SE (%)*	*95% LCI (%)*	*95% UCI (%)*
	58.4	0.2	57.9	58.9	56.7	0.3	56.0	57.4	60.4	0.4	59.7	61.1
l NHL	56.1	0.3	55.5	56.7	54.9	0.4	54.0	55.7	57.6	0.5	56.7	58.5
nodal NHL	63.0	0.4	62.1	63.8	60.4	0.6	59.3	61.6	65.8	0.6	64.6	67.0
halmic NHL	75.5	1.9	71.6	78.9	75.2	2.9	68.9	80.3	75.7	2.4	70.6	80.1
ar adnexal NHL	76.6	1.9	72.7	80.0	75.6	2.9	69.3	80.8	77.3	2.4	72.2	81.6
ophthalmic extranodal NHL	62.4	0.4	61.5	63.2	59.9	0.6	58.7	61.0	65.2	0.6	64.0	66.4
	Relative survival (%)	*SE (%)*	*95% LCI (%)*	*95% UCI (%)*	*Relative survival (%)*	*SE (%)*	*95% LCI (%)*	*95% UCI (%)*	*Relative survival (%)*	*SE (%)*	*95% LCI (%)*	*95% UCI (%)*
	68.3	0.3	67.7	68.8	66.1	0.4	65.3	66.9	70.8	0.4	70.0	71.6
l NHL	65.6	0.4	64.9	66.3	63.9	0.5	63.0	64.9	67.7	0.5	66.6	68.7
nodal NHL	73.4	0.5	72.5	74.4	70.4	0.7	69.1	71.7	76.8	0.7	75.4	78.2
halmic NHL	88.4	2.2	83.3	92.0	88.0	3.4	79.4	93.1	88.6	2.8	81.6	93.1
ar adnexal NHL	89.7	2.2	84.5	93.2	88.6	3.4	79.7	93.7	90.4	2.8	83.2	94.7
ophthalmic extranodal NHL	72.7	0.5	71.7	73.7	69.7	0.7	68.3	71.0	76.1	0.7	74.7	77.5

ence 1999–2006, follow-up through 2007.

ower confidence interval; NHL: Non-Hodgkin's lymphoma; SE: Standard error; UCI: Upper confidence interval.