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To cite this article: T. Usenius, M. Vornanen, J. Kärjä & Y. Collan (1990) Extranodal Head and Neck Non-Hodgkin's Lymphomas in Children in Finland, Acta Oncologica, 29:4, 529-531, DOI: [10.3109/02841869009090045](https://doi.org/10.3109/02841869009090045)

To link to this article: <https://doi.org/10.3109/02841869009090045>



Published online: 08 Jul 2009.



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EXTRANODAL HEAD AND NECK NON-HODGKIN'S LYMPHOMAS IN CHILDREN IN FINLAND

T. USENIUS, M. VORNANEN, J. KÄRJÄ and Y. COLLAN

Abstract

During the 20-year period from 1961 to 1980, 17 cases of extranodal head and neck non-Hodgkin's lymphoma in children under 15 years of age were diagnosed in Finland. Eight cases had tumours in tonsils or nasopharyngeal adenoids and 9 cases lymphomas of other sites of the head and neck. The age-adjusted annual incidence rate for all cases was 0.69 and for tumours of the pharyngeal lymphatic tissue 0.35 per 10^6 children. Seven of the 17 tumours were undifferentiated lymphomas (Burkitt's and non-Burkitt's types) and 5 lymphoblastic lymphomas. The overall 5-year disease-free survival rate was 37%. Five of the 9 patients with tonsillar or adenoid lymphomas were disease-free more than 5 years.

Key words: Non-Hodgkin's lymphoma, extranodal, children.

The most common cancer types in children are leukemias, tumours of the central nervous system, and malignant lymphomas. Malignant lymphomas comprise about 10% of all malignant tumours in children under 15 years of age (1, 2). Non-Hodgkin's lymphoma (NHL) in children has a propensity to involve primarily extranodal sites, most often the gastrointestinal tract (3, 4). However, extranodal non-Hodgkin's lymphomas in the head and neck region are very uncommon.

The Finnish Cancer Registry covers the whole country. It receives reports of all cases of malignant tumours diagnosed in Finland, and it gives the possibility to obtain material for epidemiological and clinicopathological studies concerning the whole nation, which can be of special value in uncommon malignant diseases.

This report deals with the patients under 15 years of age in whom extranodal NHL of the head and neck region was diagnosed in Finland during the 20-year period from 1961 to 1980.

Material and Method

One hundred and sixty-three pediatric malignant tumours in the head and neck region were reported to the Finnish Cancer Registry during the study period. All patient information, including case histories with follow-up data was collected. The histological diagnoses cited in this paper were re-examined in sections of the original tumour biopsies and reclassified according to the formulation used by Kjeldsberg & Wilson (1) for childhood NHL. Staging was performed using Ann Arbor classification (5).

Results

Seventeen patients with extranodal NHL of the head and neck (Table) were found, the male to female ratio being 2.4:1. The age range of the patients was 1 to 13 years (mean 7.3 years).

During the period analysed, the mean number of children younger than 15 years in the whole country was 1 191 500, i.e. 25.6% of the total population. The annual incidence rate of childhood extranodal NHL of the head and neck region, age-adjusted to 'world standard population' was 0.69 per 10^6 children under 15 years of age. The corresponding figure for pediatric NHL of the tonsils and adenoids was 0.35.

Of the 17 cases, 4 had tumours in the palatine tonsils, 4 in the nasopharyngeal adenoids, and one in the lingual tonsil. Furthermore, there were 8 children with extranodal NHL in other sites of the head and neck. Two of these

Presented at the First Symposium of the Scandinavian Society for Head and Neck Oncology, Turku, Finland, April 21–22, 1989.
Accepted for publication 11 January 1990.

Table

Extranodal non-Hodgkin's lymphomas of the head in Finnish children under 15 years of age during the 20-year period from 1961 to 1980

Primary site/ type of lymphoma	Number of cases		
	Male	Female	Total
Tonsils, nasopharyngeal adenoids			
Lymphoblastic lymphoma		1	1
Undifferentiated lymphoma ^a	4	1	5
Large cell lymphoma	2		2
Not specified	1		1
Extranodal, other			
Lymphoblastic lymphoma	2	2	4
Undifferentiated lymphoma ^a	1	1	2
Large cell lymphoma	1		1
Not specified	1		1
Total	12	5	17

^a Burkitt's and non-Burkitt's types

tumours appeared in the maxilla, one in the mandible, one in the parotid gland, and 4 in other sites of the head.

The distribution of the different tumour types is seen in the Table. Seven tumours were undifferentiated lymphomas (Burkitt's and non-Burkitt's types). Lymphoblastic lymphoma was present in 5 patients.

Using the Ann Arbor staging method, 5 children were found to have stage IE disease, 7 stage IIE and 7 stage IV.

The stage IE and IIE patients received radical radiotherapy or a combination of radiotherapy and chemotherapy, the stage V patients chemotherapy.

One patient was lost from follow-up. The 5-year disease-free survival rate for the 16 patients was 37% (6 patients). The corresponding rate for stage IE cases was 75%. None of the stage IV patients survived longer than three years. Five of the 9 patients with tonsillar disease remained free of disease but only one of the 8 patients with other extranodal tumours survived for at least 5 years.

Discussion

The incidence and distribution of malignant tumours in Finnish children is similar to that observed in many other white populations (6). Lymphomas comprise about 10% of the tumours and 30–40% of these are non-Hodgkin's lymphomas (1, 2). The head and neck region is an uncommon presentation site for childhood NHL, even though childhood lymphomas have a propensity for extranodal involvement.

Cobleigh & Kennedy (7) reviewed 2 173 patients with NHL of the upper aerodigestive tract and the salivary glands. Fewer than 1% of the cases occurred before age 20 years. Makepeace et al. (8) presented 51 patients with NHL arising in the tonsil; only one patients was younger

than 15 years of age. Barton et al. (9) reviewed 65 cases with NHL in the tonsil. All the patients were older than 19 years of age. Schwarze et al. (10) have presented a material collected at the Lymph Node Registry in Kiel. In the period from 1965 to 1979, 11 children younger than 16 years were found to have NHL of the oropharynx.

NHL in children also differs from adult lymphoma with regard to histopathological type. Lymphomas with a follicular pattern, which constitute about a half of the tumours found in adults, are very rare in children. In addition, childhood NHL is limited to three major histological types: lymphoblastic lymphoma, undifferentiated lymphoma (Burkitt's and non-Burkitt's type), and large-cell ('histiocytic') lymphoma (1). The types most often found in our series were the undifferentiated lymphomas (Burkitt's type) and lymphoblastic lymphomas (7 and 5 of 17 patients respectively). Our findings are in accord with the report by Kjeldsberg & Wilson (1).

The treatment of our patients was heterogenous because the patients came from different hospitals and the study period was very long (1961–1980). During this period the treatment models have changed. Stage IE and IIE patients received radical radiotherapy or a combination of radiotherapy and chemotherapy. The 5-year disease-free survival rate in the whole material was 38%. Five of the 9 patients with tonsillar or adenoid tumours remained disease-free for more than 5 years. The results are comparable to those reported by other authors (8, 9, 11).

The etiology of NHL is not known. African Burkitt's lymphoma may result from interaction of Epstein-Barr virus (EBV) and malarial infections. EBV might play a greater role in the pathogenesis of pediatric cancer than is known at present, e.g. we have found DNA sequences of EBV in adenocarcinoma of the parotid gland (12) using in situ DNA hybridization technique. We are continuing our studies to investigate the possible viral etiology of childhood lymphomas.

ACKNOWLEDGEMENTS

We wish to thank the Finnish Cancer Registry for placing its records at our disposal, and Finnish hospitals for providing us with the clinical data and biopsy material. This investigation was supported by Paavo Koistinen Säätiö. The authors would like to extend their thanks to the foundation.

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