Lymphomas of The Head and Neck

An Essay

Submitted for partial Fulfillment of master degree of Oto-Rhino-Laryngology Ain-Shams University

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2007

"بسم الله الرحمن الرحيم"

".. و يسئلونك عن الروح قل الروح من أمر ربى و ما أوتيتو من العلم إلا قليلا"

"حدق الله العظيم"

سورة الإسراء: من الآية 85

To my mother,

my wife,

and my daughter Malak

Alaa El-Din

Achnowledgement

I wish to express my deep gratitude and thanks to Prof. Dr. Osama Mahmoud, professor of otolaryngology-Ain Shams university for providing me with the initial stimulus which aroused my interest in this work.

Also, I want to express my deep thanks to Prof. Dr. Hisham El-Halaby professor of otolaryngology-Ain Shams university for his valuable help & guidance.

and finally I wish to express every deep thanks to Dr. Mohammad Amir Lecturer of otolaryngology-Ain Shams university for spending a lot of his valuable time guiding & directing me through all parts of this work.

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List of abbreviations

)	AHNSAmerican Head and Neck Surgeon
•	AIDS Aquired Immunodeficiency Syndrome
•	AJCC American Joint Committee on Cancer
•	BLBurkitt's Lymphoma
•	BLL Burkitt's Like Lymphoma
•	BNLI British National Lymphoma Investigation
•	CS Clinical Stage
•	EBV Epstien-Barr Virus
•	EORTC European Organization for Research and Treatment of Cancer
•	FDG-PET (Flouro-Deoxy-Glucose)-(Positron-Emission-Tomography)
•	HD Hodgkin's Disease
•	HIV Human Immunodeficiency Virus
•	HTLV-1 Human T-cell Lymphotropic Virus type 1
•	IPI The International Prognostic Index
•	LDHLactate Dehydrogenase
•	LDHDLymphocyte Depleted Hodgkin's Disease
•	LESA Lymphoepithelial Sialadenitis
•	LPHD Lymphocyte Predominant Hodgkin's Disease
•	MALT Mucosa Associated Lymphoid Tissue
•	MCHD Mixed Cellularity Hodgkin's Disease
•	NCI National Cancer Institute
•	NHL
•	NK-cells Natural Killer cells
•	NSHD Nodular Scelerosing Hodgkin's Disease
•	PCR Polymerase Chain Reaction
•	PET-CT Positron-Emission-Tomography Computed Tomography
•	PS Pathological Stage
•	REAL Revised European American Lymphoma classification
•	RS cells Reed-Sternberg cells
•	SCC Squamous Cell Carcinoma
•	SPECT Single-Photon-Emission Computed Tomography
•	SWOG Southwest Oncology Group
)	TLI Total Lymphoid Irradiation
)	WF The Working Formulation
•	WHOThe World Health Organization

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CHAPTER 1

Introduction & Aim Of The Work

Introduction

Billroth first used the term "malignant lymphoma" in 1871(Billroth T. 1871). Of course, this is now somewhat redundant, as there is no "benign lymphoma". Lymphomas are a group of complex and diverse lymphoid malignancies (Quraishi et al.2000). They can be considered a subset of the larger group lymphoproliferative disorders. This group includes: benign reactive lymphoproliferative disorders, histiocytosis X (more recently termed Langerhans-cell histiocytosis), plasma cell neoplasms, as well as Hodgkin's disease (HD) and non-Hodgkin's lymphomas (NHL) (Grogan T.M. 1995).

Lymphomas are a diverse group of neoplasms that comprise 4% of all new cancers annually (Decherd M., and Pou A.M. 2002).

The etiology of lymphoma is unknown but viral exposure is associated with Burkitt's lymphoma and there is a three fold increase in Hodgkin's lymphoma in adults exposed to Epstien-Barr virus (Ott G., et al. 1997). There may also be increasing numbers of human immunodeficiency virus (HIV)-related lymphomas as this disease spreads. (Parkin D.M., et al. 2000).

Cervical lympadenopathy is the most common head and neck presentation for both diseases. However, extranodal site involvement is also common in the head and neck, as 10% of lymphomas occur in areas such as Waldeyer's ring (tonsil, nasopharynx and base of the tongue), paranasal sinuses, nasal cavity, larynx, oral cavity, salivary glands(parotid), thyroid and orbit (Jacob C. 1993). Hodgkin's disease presents at younger age and is less common than NHL. Associated mediastinal lymphadenopathy were more common with HD., and abdominal adenopathy with NHL. Constitutional symptoms were more common with HD. More advanced disease with a decreased overall survival rate was seen with NHL (Urquhart A., and Berg R. 2001).

According to the *American Cancer Society 2001*, the estimated new-case rate in 2001 for NHL was 56200, for HD 7400, for oral cavity, oropharynx, and pharynx tumors 30100, for larynx tumors 10000, and for thyroid cancers 19500. These data indicate that NHL and HD pose a threat of similar magnitude to the rest of common head and neck malignancies.

Classification of lymphoid neoplasms remains one the most challenging and controversial subjects in pathology. Rappaport conceived one of the first useful systems in 1956. Subsequently other systems arose, including the Kiel, Lukes-Collins, British National Lymphoma Investigation (BNLI), World Health Organization (WHO), and Dorfman classification systems (Decherd M.E., and Pou A.M. 2002). This resulted in some degree of confusion as trials and outcomes from one system were difficult to apply to another. Of these, the Kiel classification remained active the longest, particularly in Europe and Asia.

In 1994 the International Lymphoma Study Group proposed the Revised European-American Lymphoma classification (REAL). Currently, the World Health Organization is working on an updated system based largely on the (REAL) classification (Harris N.L., et al. 1994).

Staging can be defined as determining the extent of the malignant disease within a given patient. For lymphomas, traditional TNM staging is difficult to apply, as one is often not able to distinguish between T, N, and M. The Ann Arbor system, proposed in 1971, has been the gold standard for both HD and NHLs since that time. Staging is important for assessing prognosis and determining the best course of treatment (Jacob J.R., and Negendank W.G., et al. 1991).

Treatment options for both HD and NHLs include observation, involved-field radiation, subtotal lymphoid radiation, chemotherapy with or without radiation, and bone marrow transplant.

Aim of the work

Once the diagnosis of lymphoma is established, most care is rendered by radiation oncologists and medical oncologists. The role of the otolaryngologist is usually limited to biopsy. However, for many patients the otolaryngologist serves as the entry point into the medical system.

So, the aim of the work is to

- 1. Awareness of the current knowledge of lymphomas of the head and neck.
- 2. Awareness of the proper workup of lymphomas of the head and neck.

This is of importance for the otolaryngologist to assure timely care of these cancer patients and to prevent the otolaryngologist from being a mere technician in the multidisciplinary care of these patients.

CHAPTER 2

Anatomy, Physiology & Immunology Reviews

Anatomy

The Lymph Glands of the Head and neck (Figure, 1), (Gray H.1967).

The lymph glands of the head are arranged in the following groups:

Occipital.	Posterior Auricular
Anterior Auricular	Parotid.
Facial.	Deep Facial
Lingual	Retropharyngeal.

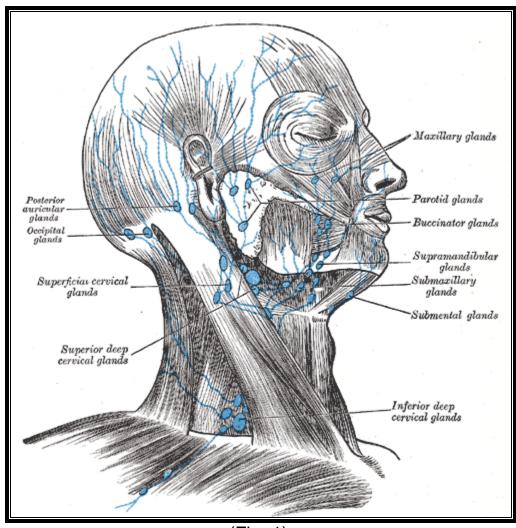
The occipital glands, one to three in number, placed on the back of the head close to the margin of the trapezius and resting on the insertion of the semispinalis capitis. Their afferent vessels drain the occipital region of the scalp, while their efferents pass to the superior deep cervical glands.

The posterior auricular glands (mastoid glands), usually two in number, are situated on the mastoid insertion of the sternocleidomastoid, beneath the auricularis posterior. Their afferent vessels drain the posterior part of the temporoparietal region, the upper part of the cranial surface of the auricle or pinna, and the back of the external acoustic meatus; their efferents pass to the superior deep cervical glands.

The anterior auricular glands (preauricular glands), from one to three in number, lie immediately in front of the tragus. Their afferents drain the lateral surface of the auricle and the skin of the adjacent part of the temporal region; their efferents pass to the superior deep cervical glands.

The parotid glands, form two groups in relation with the parotid salivary gland, a group embedded in the substance of the gland, and a group of subparotid glands lying on the lateral wall of the pharynx. Occasionally small glands are found in the subcutaneous tissue over the parotid gland. Their afferent vessels drain the root of the nose, the eyelids, the frontotemporal region, the external acoustic meatus and the tympanic cavity, possibly

also the posterior parts of the palate and the floor of the nasal cavity. The efferents of these glands pass to the superior deep cervical glands. The afferents of the subparotid glands drain the nasal part of the pharynx and the posterior parts of the nasal cavities; their efferents pass to the superior deep cervical glands.



(Fig. 1), lymph glands of the head & neck Quoted from (Gray H.1967)