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Original article

Extranodal lymphoma of the head and neck: A 67-case series

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ABSTRACT

The present study sought to describe clinical presentation in extranodal lymphoma of the head and neck (ELHN), with the aim of improving diagnostic management.
Material and methods: A single-center retrospective observational study was conducted over the period 2001–13. Age, gender, histologic type, location, type of clinical presentation, time interval between symptom onset and histologic diagnosis and presence of specific symptoms were recorded, as were the specialty of the physician initially consulted and of the physician taking the diagnostic sample.
Results: Sixty-seven cases of ELHN were diagnosed: 39 male and 28 female patients, with a median age of 68 years. B-cell lymphoma (84%) was more frequent than plasmacytoma (7%) or T-cell lymphoma (6%). Location was mainly palatine tonsil (28%), nasal fossa and sinus (19%), nasopharynx (14%) or parotid (13%). Revelation often involved a mass (33%), and only rarely any specific symptoms (9%). Time interval from symptom onset to diagnosis was short in aggressive lymphoma and longer in low-grade lymphoma (mean 4 and 10 months respectively). The physician initially consulted was an ENT specialist in 67% of cases, and an ENT specialist performed diagnostic sampling in 97% of cases.
Conclusion: ELHN is a rare pathology (5 cases per year in our department) of highly variable clinical presentation depending on location and histologic type. The ENT physician should be prepared for diagnosis regardless of anatomic location, so as to optimize diagnostic management.

1. Introduction

Hodgkin and non-Hodgkin lymphoma is the third most frequent malignant tumor of the head and neck region (12%), following squamous cell carcinoma (46%) and thyroid carcinoma (33%) [1]. Incidence has been rising for several decades. There are some 10,000 new cases of non-Hodgkin lymphoma of whatever location per year in France, slightly more than half occurring in male subjects. Twenty-three percent of non-Hodgkin and 4% of Hodgkin lymphomas of the head and neck are extranodal [2]. These lymphoid tumors mainly involve 4 sites: Waldeyer's ring, the nasal sinuses and fossae, the oral cavity and the salivary glands.

Extranodal lymphoma comprises a heterogeneous group of tumors of various histologic types and highly varied clinical presentation. Diagnosis may fail to be suspected when the clinical and/or radiological tumoral syndrome mimics epithelial tumor or infection, but needs to be considered as treatment is specific.

The present study sought to describe the characteristics of patients with extranodal lymphoma of the head and neck (ELHN), highlighting the polymorphic clinical presentation and the role of the ENT physician in the optimization of management.

2. Materials and methods

Files of all patients presenting with ELHN between January 1st, 2001 and December 31st, 2013 were analyzed retrospectively, excluding intra- and juxta-parotid nodal locations.

Definitive diagnosis was histologic. Pathology exams were analyzed according to their code on the ADICAP classification (*Association pour le Développement de l'Informatique en Cytologie et en Anatomie Pathologique*: Association for the Development of Informatics in Cytology and Anatomo-Pathology), enabling all extranodal locations (excluding code SG: [node]) of the head and neck to be selected: codes AA (amygdala), XF (face), AC (nasopharynx), XC (cervical region), AF (nasal fossae), AS (facial sinus), OE (eye), BX (oral cavity), AL (larynx), BL (tongue) and BP (parotid).

The WHO 2009 classification was used for characterizing histologic type: B-cell, T-cell or lymphocytic (LCL) lymphoma or plasmacytoma. B-cell lymphoma was classified as: MALT

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Table 1
Frequency of histologic types of ELHN.

	Histologic type	Number	Frequency (%)
B-cell lymphoma, $n = 56$	Diffuse large B-cell lymphoma (DLBCL)	36	54
	MALT B-cell lymphoma	6	9
	Lymphocytic lymphoma (LCL)	4	6
	Brain B-cell lymphoma	4	6
	Burkitt's B-cell lymphoma	3	4.5
	Centroblastic B-cell lymphoma	2	3
	Low-grade B-cell lymphoma	1	1.5
T-cell lymphoma, $n = 4$	T/NK nasal lymphoma	3	4.5
	Angioimmunoblastic T-cell lymphoma	1	1.5
Plasmacytoma		5	7
Undetermined		2	3

(mucosa-associated lymphoid tissue) lymphoma, small-cell cerebral lymphoma, Burkitt's lymphoma, CD20+ diffuse large B-cell lymphoma (DLBCL), centroblastic B-cell lymphoma or other low-grade B-cell lymphomas T-cell lymphoma was classified as L angioimmunoblastic T-cell lymphoma or angiocentric T/NK-cell nasal lymphoma.

Lymphomas were further classified as aggressive or indolent.

DLBCL, Burkitt's and T/NK-cell lymphomas are highly malignant and aggressive. Lymphoma associated with HIV is generally highly malignant. Follicular, low-grade B-cell, brain cell, LCL, MALT lymphomas and plasmacytoma are of low malignancy or indolent.

Age at diagnosis, gender, histologic type, location, type of clinical presentation, time interval between symptom onset and histologic diagnosis and known risk factors (HIV-positive status and history of immune disorder) were recorded.

The presenting symptom leading to diagnosis was recorded and counted as the clinical presentation. B symptoms and cervical adenopathies were recorded; B symptoms comprise general symptoms: fever, night sweats and weight-loss, associated with both Hodgkin and non-Hodgkin lymphoma.

The specialization (ENT or other) of the physician seen for the presenting symptom leading to diagnosis and of the physician taking the diagnostic sample was recorded in each case. The type of sample enabling histologic diagnosis was classified as surgical specimen (total or partial organ resection) or surgical biopsy. Fine-needle aspiration ahead of diagnostic biopsy was also recorded.

All patients underwent local and remote imaging. Fig. 1A and B shows two examples (CT and MRI) of a temporal plasmacytoma and a thyroid lymphoma. The images are not pathology-specific and contrasted with the respective clinical presentations.

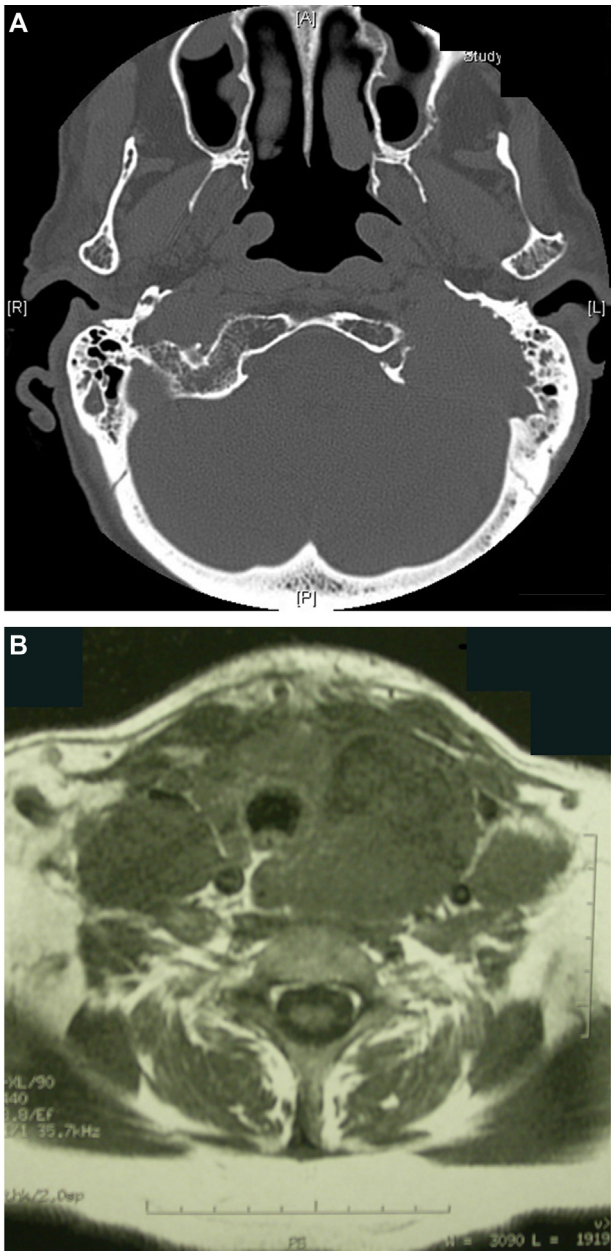

Fig. 1. A: Temporal bone CT scan, axial slice. Temporal bone plasmacytoma: severe bone lysis, in contrast to relatively silent symptomatology: hearing loss and vestibular hyper-reflexivity of progressive onset over several months. B: cervical MRI, axial slice: thyroid DLBCL: left thyroid mass pushing back and invading trachea. Inspiratory dyspnea requiring intensive care.

3. Results

Sixty-seven ELHNs were diagnosed in the department between January 1st, 2001 and December 31st, 2013: 39 male and 28 female patients (M/F sex ratio, 1.4), with a mean age of 65 years (range, 17–97 years). Fig. 2A shows incidence according to age.

B-cell lymphoma ($n = 56$, 84%) was more frequent than plasmacytoma ($n = 5$, 7%) or T-cell lymphoma ($n = 4$, 6%). Table 1 shows frequency according to histologic type.

The most frequent histologic type was DLBCL, a high-grade B-cell lymphoma ($n = 36$). MALT B-cell lymphoma was systematically parotid ($n = 6$) and LCL systematically tonsillar ($n = 4$). There were no cases of Hodgkin lymphoma.

T-cell lymphoma comprised T/NK nasal lymphoma ($n = 3$) and angioimmunoblastic T-cell lymphoma ($n = 1$). T/NK lymphoma was systematically EBV-positive, with sinonasal location; the angioimmunoblastic T-cell lymphoma was nasopharyngeal.

The most frequent ELHN locations were: palatine tonsil ($n = 19$, 27%), nasal fossa and sinus ($n = 13$, 19%), nasopharynx ($n = 10$, 14%) and parotid gland ($n = 9$, 13%). Other locations comprised tongue, soft palate, face, orbit, thyroid, lingual tonsil, temporal bone and larynx. Four patients had 2 extranodal locations of simultaneous or consecutive onset: 1 hypophyseal LCL with concomitant tonsillar LCL; 1 basilingual DLBCL 9 years after rhinopharyngeal DLBCL; 1 left parotid MALT lymphoma 6 years after right parotid MALT lymphoma; and 1 tonsillar DLBCL 4 years after sinonasal DLBCL. ELHN locations are shown in Fig. 2B.