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CASE REPORT

Non-Hodgkin's Malignant Lymphomas of the Palatine Tonsils: A Case Report

Z. El Hafi[1,3], W. Sidielmoctar Abdallahi[1,3], K. Hjaouj[1,3], R. Bencheikh[1,3], M.A. Benbouzid[1,3], A. Oujilal[1,3], N. Cherradi[2,3], H. EL Ouazzani[2,3] & L. Essakalli[1,3]

[1]ENT-HNS Department of Specialties Hospital – CHU Ibn Sina Rabat, Morocco
[2]Pathology Department, of Specialties Hospital – CHU Ibn Sina Rabat, Morocco
[3]Mohammed V University in Rabat; Morocco

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Abstract

The palatine tonsils are the first extra-nodal localization of cervicofacial non-Hodgkin's malignant lymphomas. It is a rare localization and most often a type B lymphoma. This case highlights the difficulty of making an early diagnosis, given the non-specificity of clinical and paraclinical signs, and the fact that only anatomopathological examination can confirm the diagnosis. Chemotherapy is the only treatment for chemo sensitive forms of the disease. Prognosis is generally good, depending on sensitivity to treatment and stage of disease.

Keywords:

- Non-Hodgkin's lymphoma
- Palatine tonsil
- Oral cavity
- Radiotherapy
- Chemotherapy
- Homeopathy

Corresponding Author:
Z. El Hafi, ENT-HNS Department of Specialties Hospital – CHU Ibn Sina Rabat, Morocco; Mohammed V University in Rabat; Morocco

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Introduction

Malignant non-Hodgkin's lymphomas (MHNL) are malignant proliferations of lymphoid tissue. They account for 5% of malignant tumors of the head and neck, and develop preferentially from extra-nodal lymphoid tissue [1,2]. Localization in Waldeyer's ring accounts for around 10% of all LMNH and more than a third of extra-nodal localizations. LMNH present themselves as different entities recognized within new classifications that incorporate data from modern immunophenotyping [4]. Histological type is more decisive in therapeutic management than location.

Clinical Case

A 57-year-old female patient with type 2 diabetes on oral antidiabetics and cardiac disease on platelet inhibitors presented with a 4-month history of ulcerative lesions of the left tonsil, associated with bilateral cervical, axillary and inguinal adenopathy

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and hypertrophy of the contralateral tonsil. On CT scan, the appearance was suggestive of a left tonsillar tumefaction with cervical adenopathy involving the entire cervical chain bilaterally. A biopsy performed at the level of the ulceration and whose anatomo-pathological examination came back in favour of a malignant non-Hodgkin's lymphoma of B phenotype. Biologically, the LDH level was elevated (over 3 mg/l). The blood count and sedimentation rate were without abnormalities. As part of the extension work-up, digestive fibroscopy and bone marrow biopsy were normal.

Patient referred for further chemotherapy with: ACVBP (Adreblastine, Endoxan, Oncovin, Bleomycin, Prednisone) and external radiotherapy.

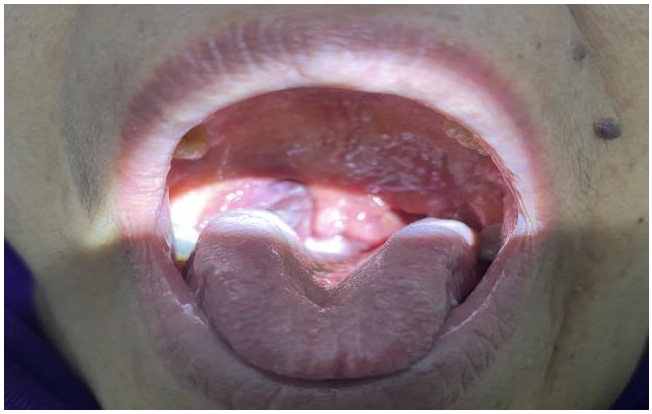

Figure 1: Oropharyngeal examination: bilateral tonsillar hypertrophy with ulcerated appearance on left side

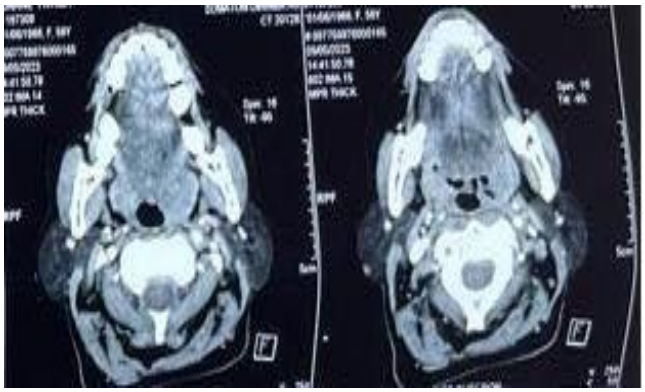

Figure 2: Cervical CT axial section: Tumor of the left palatine tonsil

Discussion

Malignant tumors of the palatine tonsil are predominantly carcinomas, while primary LMNH occupy second place with a frequency of 5 to 14% [3,5]. Among LMNH of Waldeyer's ring, the palatine tonsil is the primary site (40 to 79% of primary lesions), followed by the nasopharynx (25 to 35%) and the lingual tonsil (3 to 10%) [3]. Multiple involvement is seen in almost 5% of cases of tonsillar lymphoma. Bilateral involvement occurs in 10% of cases [4]. Epitheliomas, plasmacytomas, adenocarcinomas, melanomas, Hodgkin's disease, sarcomas and metastases are rare tumor localizations of the palatine tonsil. An increased risk of LMNH has been observed in patients with autoimmune diseases, particularly dry syndrome, which increases this risk by a factor of 40 [6]. Other factors favour the occurrence of LMNH and explain the current increase in the incidence of this disease, such as AIDS, where B lymphoma constitutes an opportunistic neoplasm, and acquired immune deficiencies following organ transplantation with immunosuppressive treatment [6,7].

LMNH can occur at any age, but the average age of onset is around 59. More than 80% of Waldeyer's ring NHLs occur after the age of 50 [7,9]. Most series in the literature describe an even distribution of the disease according to sex. Occasionally, there is a slight male predominance [10].

LMNH of the tonsil is rapidly progressive, with nonspecific presenting signs such as tonsillar asymmetry, odynophagia, upper dysphagia and cervical swelling, as in the case of our patient. These signs can lead to confusion with simple angina. In such cases, the diagnosis is often overlooked and delayed [11]. It is the persistence of the symptomatology, despite well-managed treatment, that prompts the diagnosis. Bilateral tonsillar involvement is rare, accounting for 10% of cases. Systemic signs, uncommon and present in only 25% of cases, are helpful in diagnosis [7,12]. Associated lymph node involvement is common, occurring in around 2/3 of cases. It is often homolateral to the mobile, painless lesion, quite different from lymph node metastases of squamous cell carcinoma [4]. At the time of diagnosis, It is firm, evolving beneath a non-ulcerated mucosa, contrasting with the absence of pain. This may suggest a peritonsillar phlegmon, but the absence of infectious signs, pain, trismus and uvula edema should alert the examiner. Unfortunately, it is not uncommon for these lesions to be wrongly incised in the first instance [4]. Imaging does not contribute to a positive diagnosis, especially as a small asymmetry in size between the two tonsils is considered normal. However, imaging can be useful in assessing tumour volume and detecting non-palpable adenopathies, particularly in the retro pharyngeal region. The absence of invasion of adjacent structures of the tonsil, despite the size of the tumor mass, is in favor of a lymphomatous localization [12]. In cases of clinically suspected tonsillar lymphoma, the diagnosis can only be confirmed by biopsy. Histological examination includes: a simple morphological analysis of the architecture of the tissue sampled (diffuse or follicular tumor invasion); immunohistochemistry, which confirms the lymphoid nature of the tumor mass and establishes its B or T phenotype, looking for other markers specific to each sub-type; and finally, cytogenetic analysis (looking for chromosomal abnormalities). The patient's workup includes myocardial function and, in addition to standard biology, HIV, HTLV-1, EBV and hepatitis C serologies, in order to search for a viral context that may have favored the onset of lymphoma. Lastly, a stomatological evaluation must be carried out with a view to irradiation. At the end of this work-up, a number of pejorative prognostic elements can be identified [14,15]: age over 60, site of involvement, volume greather than 5 cm3. Stage III or IV, involvement of the spinal cord or neurogical site, elevated LD or 2 microglobulin level, Abnormalities of chromosomes 6, 7 and 17, HIV infection.

The International Prognostic Index (IPI), which takes into account five of these factors (age greater than 60 years, PS greater than 2, stage III or IV, high LDH level, presence of more than one extra-nodal location), makes it possible to separate patients with a

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different evolutionary profile and to adapt therapy for aggressive lymphomas [7]. NHLs of the palatine tonsil are almost always of B phenotype, with a predominance of diffuse large-cell forms over follicular and mantle lymphomas. There are rare cases of anaplastic T lymphomas [16]. These diffuse large-cell lymphomas are aggressive tumors, characterized by rapid spontaneous evolution, and as a result, the initial presentation is localized (stages I and II) in just over a third of cases [4,12]. First-line treatment is adapted to the initial prognostic factors described above. It is based on chemotherapy, which alone can give a chance of cure. Radiotherapy can only play an adjuvant role [4]. According to Fisher et al [17], the reference chemotherapy is protocol This stage of the work-up enables the disease to be classified according to the Ann Arbor stages: - Stage I: involving only the site that led to the diagnosis. CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone), which is better tolerated than third-generation protocols. For some authors, exclusive radiotherapy appears to be a suitable treatment for low-grade stage I and II lymphomas [18]. Combining chemotherapy with irradiation of affected lymph nodes appears to significantly improve prognosis. The 5-year survival rate for NHL confined to the palatine tonsil is 86%. This figure drops to 41% in the presence of adenopathy [8]. Relapses are frequent and indicate a poor prognosis. They occur mainly in the first two years after treatment. They occur at a distance from the primary site, particularly in non-irradiated lymph nodes and in the gastrointestinal tract [13].

Conclusion

Primary non-Hodgkin's malignant lymphomas of the palatine tonsil are rare. Their diagnosis is evoked by unilateral tonsillar hypertrophy and confirmed by biopsy. Early diagnosis is difficult (intraparenchymal nodular stage). Therapeutic decisions can only be taken once the histological diagnosis has been clearly established, followed by an assessment of the disease and the patient. Prognosis depends on stage. The International Prognostic Index (IPI) can be used to separate patients with different evolutionary profiles, and to adapt management.

Conflicts of interest: There is no potential competing interests

Ethical Consideration: None

Acknowledgements: None

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