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CASE REPORT

MALIGNANT LYMPHOMA WITH NASOPHARYNGEAL DEBUT - COMMENTS ON A CLINICAL CASE

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Summary

Malignant lymphoma is an issue of major importance in medical practice, with an increasing incidence in recent years, along with leukemia, being the most common malignancy among young population. Onset of the disease can be both nodal and extranodal as a quarter of extranodal lymphomas onset is located in the head and neck. This article is based on a particular clinical case of non-Hodgkin malignant lymphoma with nasopharyngeal onset. The paper highlights the importance of a multidisciplinary therapeutic management successfully used to treat a case of malignant non-Hodgkin lymphoma with nasopharyngeal location.
Key words: malignant nasopharyngeal lymphoma, multimodal treatment

Résumé

Lymphome malin à début naso-pharyngé

Le lymphome malin est une question d'importance majeure dans la pratique médicale, avec une incidence croissante ces dernières années, étant avec la leucémie le cancer le plus fréquemment rencontré parmi la population jeune. L'apparition de la maladie peut être à la fois nodale et extraganglionnaire vu un quart des lymphomes extra-ganglionnaires est situé à la tête et au cou. L'article est basé sur une présentation de cas clinique particulier de lymphome malin non-Hodgkin à début naso-pharyngé. Le document souligne l'importance d'une prise en charge thérapeutique multidisciplinaire utilisée avec succès pour traiter un cas de lymphome non hodgkinien malin à location naso-pharyngienne.
Mots-clés: lymphome malin du nasopharynx, traitement multimodal

Introduction

Lymphoma is a lymph proliferative disease, hematological par excellence. Lymphomas are divided into two categories, malignant Hodgkin lymphoma and malignant non-Hodgkin lymphoma. Malignant non-Hodgkin lymphomas can occur at both nodal and extranodal level and are most often on the stomach, liver or spleen [1].

Recent years have seen a steady increase in the incidence of malignant non-Hodgkin lymphoma, particularly among those with extranodal sites [1]. Extranodal lymphomas can appear on any organ, but head and neck localization ranks second in frequency, after the gastrointestinal sites, and generally have a reserved prognosis [2,3, 4].

Nasopharyngeal localized lymphomas are a challenge in terms of diagnosis and treatment because the patient presents late to the doctor [5]. An increase of at least 5% of the occurrence of malignant lymphoma was observed in general, especially in patients over 60 years of age and those with HIV positive [6], ranking first among hematological neoplasms [7]. An increase in terms of the extranodal localization of lymphomas was also observed [8]. Although there is no known exact cause of the disease, there are certain risk factors that can be involved in the occurrence of malignant lymphomas such as organ transplantation, Sjogren's syndrome, Hashimoto's thyroiditis, HIV or Castleman disease [9, 10]. In all of these conditions there is a depression

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of the T-cell or cell-mediated immunity and a B-cell growth in the lymphoid tissue, especially in the presence of Epstein-Barr virus. Positive diagnosis of malignant non-Hodgkin lymphoma in extranodal sites is based on pathological and immuno-histochemical examination. These examinations are also necessary for the correct choice of treatment depending on the type of the lymph proliferation (B or T) [11].

Among lymphomas occurring on the upper respiratory tract, the most common are marginal zone B cell lymphomas, MALT lymphomas, diffuse large B cell lymphomas and peripheral T-nasal type NK with cytotoxic phenotype lymphomas. Malignant lymphoma with nasopharyngeal location can be primitive or secondary and with or without medullar interest [12]. At this level the most common primary T cell lymphomas most often begin with chronic nasal obstruction, rhinorrhea and unilateral or bilateral transmission hearing loss [13].

Treatment of malignant nasopharyngeal lymphoma requires an interdisciplinary collaboration between the hematologist, oncologist, radiotherapist, otolaryngologist and pathologist. Treatment varies depending on histological type, degree of malignancy, age of the patient and especially clinical stage and presence or absence of metastases [14]. The principal means of treatment is represented by conventional chemotherapy, the most often used in the form of multi-agent chemotherapy [15]. Another mean is radiotherapy [16] which is most often associated to multi-agent chemotherapy.

There are also a number of modern therapeutic methods such as bone marrow transplantation, the use of monoclonal antibodies [17, 18] or treatment with interferon, which if used together with multi-agent chemotherapy prolong the life of the patient [19].

The goal of surgery is firstly the diagnosis of the disease, given that accurate diagnosis is made on the pathological analysis, and secondly the reduction in size of the tumor [187] to reduce the risk of local complications or to potentiate the immune response.

Case report

We present the case of a 61 years old patient diagnosed with malignant non-Hodgkin lymphoma, localized in the nasopharynx. The patient presented bilateral chronic nasal obstruction, symptoms started one year ago. The patient was also known with hypertension and type II diabetes.

Endoscopic examination with flexible or rigid fiberoptic shows evidence of hypertrophy of lymphoid tissue in the nasopharynx (Fig. 1), from where biopsy fragments are taken to be sent to histopathology and immunohistochemistry exam that arrive to diagnose the disease as malignant non-Hodgkin lymphoma with small B cell of the marginal zone type MALT. Histological features were: CD20 +, CD5 +, CD10 +, CD79a +, BCL2 diffuse positive.

The patient was referred to the Department of Hematology, where she conducted a series of investigations that revealed elevated serum LDH and ESR, absence of nodal affecting and presence of bone marrow damage leading to the classification of the case in stage IV according to the Ann Arbor classification. Due to the investigations, six cycles of CHOP type multi-agent chemotherapy were carried out and were well tolerated by the patient.

Figure 1 - Nasal endoscopic exam

The ENT control reexamination and the flexible fiberoptic exam shows the presence of tumors in the nasopharynx. Given the poor response to chemotherapy it is decided to carry out radiotherapy with a total dose of 44 Gray. After radiotherapy there is a partial regression of the disease. The patient is admitted to the ENT clinic accusing bilateral hearing loss, sensation of blocked ears and bilateral chronic nasal obstruction.

Pure tone audiometry shows medium transmission hearing loss in her right ear and slight transmission hearing loss in her left ear (Fig. 2). Impedancemetry highlights type B tympanogram on the right ear and type C tympanogram on the left ear.

The patient underwent surgery under general anesthesia with intubation, with the primary goal of excising the tumor formation from the cavum. Subsequently, bilateral myringotomy was made with secretions aspiration from the middle ear and Diabolo type aerator tube mounted at the right tympanic membrane.

Tumor excision was performed under endoscopic control with rigid 00 fiberoptic, which facilitated access in a hard to

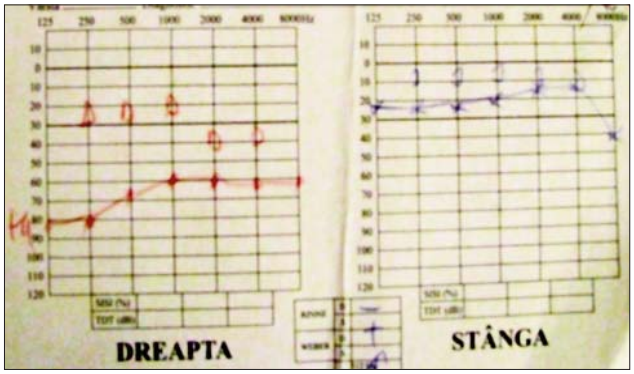

Figure 2 - Pure tone audiometry before surgery

reach region such as the nasopharynx. The intervention required the use of endoscopic instruments particularly appropriate for the removal of lymphoma tumour in areas not directly illuminated and to protect Eustachian tube openings and adjacent tissues. One of these tools is the surgical microdebrider (shaver) that grains soft tissues which it then sucks through its vacuum system.

Bilateral myringotomy is performed under otomicroscopic control by suctioning mucous secretions from the middle ear on both sides and fitting an aerator trans tympanic tube at the right tympanic membrane level (Fig. 3, 4).

Extracted pieces were sent to histopathology and immunohistochemistry that revealed fragments of mucosa that had lymphoid corium hyperplasia mostly parafolicular diffuse; germinal centers with reactive aspect; groups of monocytoid cells.

Postoperatively, the patient experienced a favorable evolution through nasal desobstruction. Liminal tonal audiometry 7 days after surgery shows a significant recovery of bilateral hearing deficiency (Fig. 5).

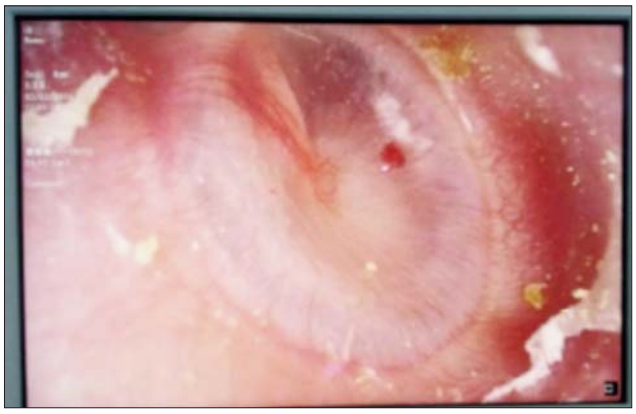

Figure 3 - Serous otitis before the miringotomy

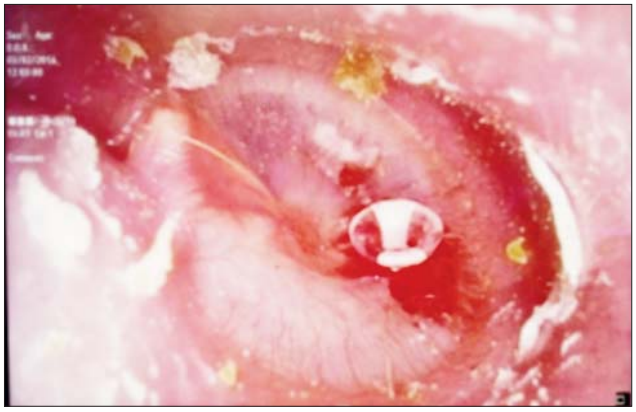

Figure 4 - The right ear myringotomy with Diabolo tube

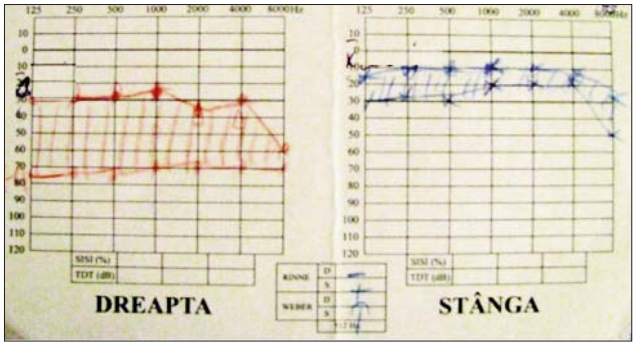

Figure 5 - Postoperative pure tone audiometry

Results and discussion

Among lymphoma category with extranodal onset, a significant group is represented by nasopharyngeal localized malignant lymphoma. Although malignant non-Hodgkin lymphoma is par excellence a hematological disease and overall treatment is multi-agent chemotherapy and radiotherapy, we tried to highlight a special clinical situation in which surgical approach has an important role in disease treatment.

The goals of the performed surgery were supporting the diagnosis by taking biopsy fragments, nasopharyngeal tumor excision for nasal desobstruction and performing miringotomy with transtympanic nasal tube mounting on the right ear.

Reducing the volume of tumor formation has proved helpful to potentiate the immune response and increase the effect of radiotherapy and chemotherapy but also in hearing deficit recovery.

The role of surgical treatment for malignant lymphoma may be found both in biopsy and in ablation, especially given, as regards this case, that multi-agent chemotherapy and radiotherapy response to treatment was minimum. Negative prognosis of the disease and symptomatology, along with the decrease in hearing, imposed performing surgery and miringotomy with transtympanic aerator tube fitting in the right ear. Another role of the surgery was to prevent histological conversion of the disease, taking into account the fact that the patient was in stage IV, having spinal damage and that the malignant lymphoma was an aggressive histological type.

Conclusions

Endoscopic surgical approach in this case has facilitated access in an area difficult to access with conventional surgery and the use of specific tools helped perform surgical gestures with greater precision and accuracy. The double video endoscopic transnasal and transoral approach has allowed a better exposure of the lesion and of the cavum in general without damaging adjacent healthy tissue and with reduced bleeding of the wound. Also, using an operating microscope has allowed interventions to be more precisely performed on the tympanic membrane.

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