Small Lymphocytic Lymphoma Arising from the Nasopharynx

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Abstract

The majority of nodal or extranodal lymphomas of the head and neck are Non-Hodgkin's lymphomas (NHL). Most of them are B-cell tumors. T-cell tumors and rarely natural killer cell tumors are the other subgroups. Diffuse large cell lymphoma is the most frequently seen type of B-cell NHL and is followed by follicular lymphoma. Small lymphocytic lymphoma is another subtype of B-cell NHL, representing 5%-10% of all NHL. The treatment is performed with radiotherapy and/or chemotherapy, depending on the stage of the disease. In this study, we present the case of a 62-year-old female diagnosed with B-cell small lymphocytic lymphoma arising from the nasopharynx. The patient described as Stage 1E according to the Ann Arbor staging system was referred to the hematology/oncology clinic. Radiotherapy was selected as the treatment modality. There was no mass observed on endoscopic and radiologic examinations, which were performed 24 months after treatment.

Keywords: Small lymphocytic lymphoma, nasopharynx, treatment

INTRODUCTION

Lymphoma is the second most common neoplasm after squamous cell carcinoma in the head and neck region and corresponds to 2%–3% of malignant tumors in this region (1). While 90% of head and neck lymphoma cases show the lymph node involvement, the remainder develops from the extranodal areas such as Waldeyer's ring (2).

Small lymphocytic lymphoma/chronic lymphocytic leukemia is the most common form of adult leukemia in the United States and Europe. The incidence is 1-5.5/100.000. It is a painless systemic disease with the bone marrow and peripheral blood involvement, and/or extensive lymphadenopathy. The age at diagnosis is 64-70 years. The incidence is higher in males and in the white race (3). In cases with nodal or extranodal tissue involvement, if there is no bone marrow or blood lymphocytosis, the disease is defined as small lymphocytic lymphoma (4).

In this study, the patient with B-cell small lymphocytic lymphoma arising from the nasopharynx is presented. The characteristics of the diseases are summarized in the following.

CASE PRESENTATION

A 62-year-old female patient was admitted to a health facility 4 months before due to the hearing loss in her left ear. Nonspecific treatment was applied to the patient.

Then the nasal obstruction was added to the complaints of the patient, and she applied to a different medical center. Because of the nasopharyngeal mass, the patient was referred to our clinic.

On the otoscopic examination, it was observed that the left tympanic membrane had matt, partially retracted, and increased vascularity. The left ear had an average of 25 dB conductive hearing loss on the pure tone audiogram and Type B curve on tympanometry. On the endoscopic examination, a vegetative mass 25×30 mm in size that obliterated the nasopharyngeal air column was observed. Other findings were normal. Magnetic resonance imaging (MRI) showed a homogenous contrast-enhancing mass lesion 39x27x42 mm in size in the left nasopharyngeal region, causing partial narrowing in the axial T1-weighted cross-sectional area. It was emphasized that the mass pressured surrounding muscle structures (Figure 1). Biopsy was taken from the nasopharyngeal mass under local anesthesia in our patient.

Cite this article as:
Ülkü ÇH, Aydoğdu D, Mukhtarova E, Avunduk MC. Small Lymphocytic Lymphoma Arising from the Nasopharynx. Eur J Rhinol Allergy 2019; 2(2): 64-7.

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Received: 22.01.2019
Accepted: 09.06.2019
DOI: 10.5152/ejra.2019.147

Histopathological (Figure 2)/immunohistochemical examination revealed a diffuse lymphoid cell infiltration with a positive expression of CD 19 (Figure 3), CD20 and CD5, and a Ki67 index of 60% (Figure 4). Cells were small and monotonous and were stained as membranous with CD 20. The diagnosis was reported as small-cell lymphocytic lymphoma. The Epstein–Barr virus (EBV) IgG was positive.

The number of white blood cells was 8.500 in the hemogram. The peripheral smear was within the normal limits with 49.8% for neutrophils, 41% for lymphocytes, and 6.6% for monocytes. The patient was referred to hematology and oncology. No bone marrow biopsy was performed because there was no pathological blood value. According to the Ann Arbor staging system, the case was classified as Stage 1E. Isolated nasopharyngeal involvement was detected, and radiotherapy was planned with diagnosis of the early-stage/low-grade B-cell lymphocytic lymphoma. The patient was informed about the procedure, and a written consent form was obtained. An endoscopic examination performed 24 months after the treatment revealed no mass. A full regression was reported in the magnetic resonance axial T1-weighted contrast image (Figure 5). Patient's hemogram, otoscopic, and audiologic evaluation were within the normal limits. The patient is still followed by both clinics using a multidisciplinary approach.

DISCUSSION

In 90% of the cases, head and neck lymphoma is observed as a regional lymph node involvement, while the rest affects extranodal areas such as Waldeyer's ring (2). Majority of the nodal or extranodal head and neck lymphomas are NHL. Waldeyer's ring lymphoid tissues, including the nasopharynx, are thought to be relatively resistant to Hodgkin's lymphoma (5).

The Waldeyer's ring involvement corresponds to 7% of all NHL cases (6). Jacops et al. (7) analyzed 156 extranodal NHL cases affecting the head and neck region. In 66% (n=103) of the patients, the tonsil, nasopharynx, or tongue base (Waldeyer's ring components) involvement, and in 34% (n=53), the extralymphatic area involvement such as the salivary glands,

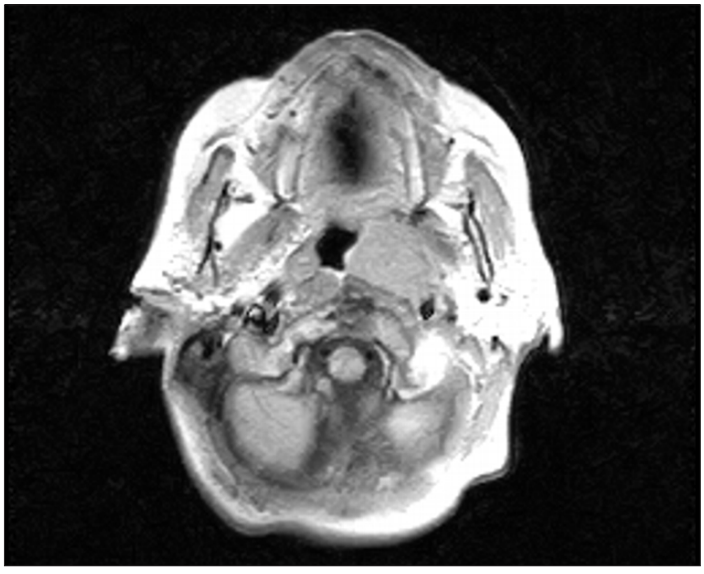

Figure 1. Axial view of the nasopharynx and the mass on T1-weighted MRI (before treatment)

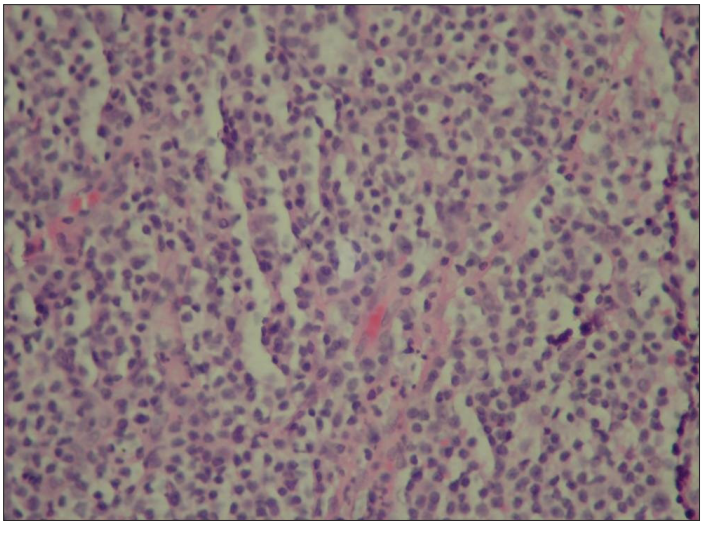

Figure 2. Histopathological view (Hematoxylin eosin)

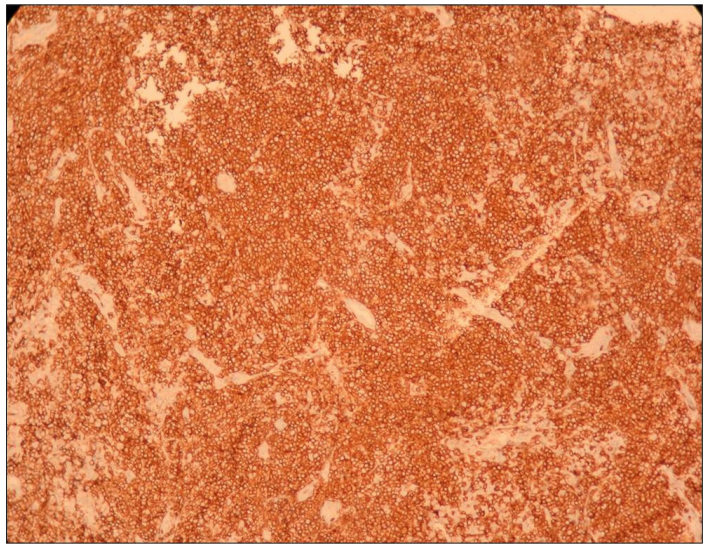

Figure 3. Immunohistochemical / Histopathological view (Positive expression with CD 19)

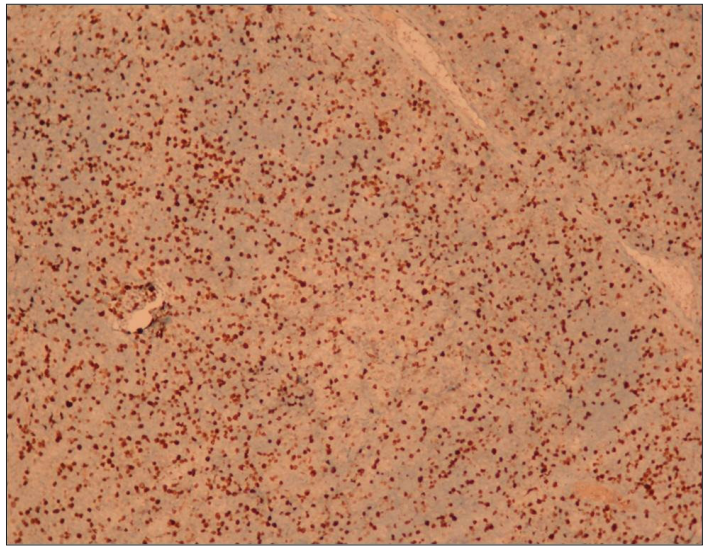

Figure 4. Immunohistochemical / Histopathological view (Ki67)

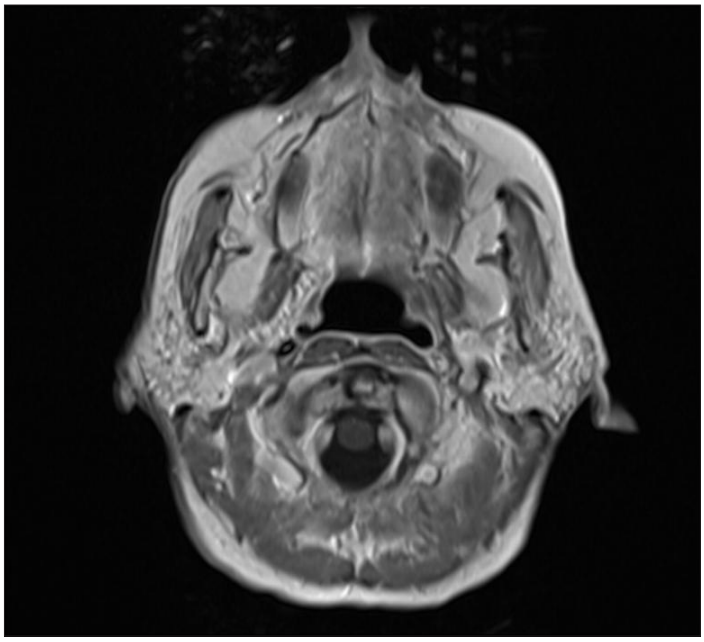

Figure 5. Axial view of the nasopharynx on T1-weighted MRI (after treatment)

paranasal sinuses, oral cavity, and larynx have been reported. In the same study, the tonsils were the most commonly involved anatomical site with a rate of 38%. It was followed by the nasopharynx (16%), paranasal sinuses (13%), salivary glands (13%), and tongue base (12%).

Non-Hodgkin's lymphomas are mostly B-cell (80%–90%) tumors, followed by T-cell and rarely natural killer cell tumors. Among B-cell lymphomas, while diffuse wide-cell lymphoma is the most common (30%-40%), follicular lymphoma is seen in 20%–30%. Small lymphocytic lymphoma is another subtype of B-cell lymphomas and constitutes 5%–10% of all NHLs. It usually occurs in persons aged >60 years and more frequently in males (8).

Hereditary and genetic factors have been noted in the development of chronic lymphocytic leukemia/small lymphocytic lymphoma. There is no single environmental risk factor. It was thought that the exposure to pesticides, sunlight, ionizing radiation, carcinogens, diet, alkalizing agents, hepatitis C, and EBV could pose a risk, but a consistent relation has not been detected (3). It is a low-grade and slow-growth-prone NHL subtype (3, 8). It is often not noticed by the patient until a diagnosis is made by the high number of lymphocytes in routine control examinations. The most common symptom is lymphadenopathy, while the difficulty in exercising and fatigue are common complaints (3).

Waldeyer's ring is often a reservoir area for EBV, and some authors indicate that EBV for both carcinoma and NHL may be a causal factor (5, 9). However, the data on nasopharyngeal Hodgkin lymphoma and EBV are controversial (5).

The symptoms and signs of NHLs developing in the nasopharynx are nonspecific and may mimic nasopharyngeal carcinoma (5, 8).

In the present case, the main complaints of the patient were the hearing loss in the left ear and nasal obstruction. On the endoscopic examination, a vegetative mass that obliterated the nasopharyngeal air column was observed. The left ear had conductive hearing loss on pure tone audiogram and Type B curve on tympanometry. MRI reported a mass lesion in the left nasopharyngeal region that caused partial narrowing in the lumen and a homogenous contrast enhancement. It was emphasized that the mass pressured the surrounding muscle structures.

In a study of 114 extranodal NHL cases developed in the head and neck region by Shima et al. (8), the mean age was 60.5, and the female-to-male ratio was reported as 1:1.5. The most commonly affected area was Waldeyer's ring, followed by the oral cavity, paranasal sinuses, nasal cavity, and larynx. 75% of the cases were defined as Stages I and II, and 89% as B-cell and 11% as T-cell origin.

The definitive diagnosis of the disease is made by biopsy. For the appropriate treatment protocol, it is important to determine the subgroup with immunohistochemical examination and perform to being staging (8).

In approximately 10%-20% of chronic lymphocytic leukemia/small lymphocytic lymphomas, second primary malignancies develop, and death occurs in 2 out of 3 of cases (10).

A biopsy sample was taken from the nasopharyngeal mass under local anesthesia in our patient. The histopathological/immunohistochemical examination revealed B-cell lymphocytic lymphoma. The EBV IgG was found to be positive. Blood values were normal. The patient was described as Stage 1E according to the Ann Arbor staging system.

When all NHLs are evaluated, the 5-year survival rate is 54%. Gender, histological grade, T/B phenotype, clinical stage, and the area of origin are the factors affecting survival. The presence of low-grade, T-cell, and advanced stage is among the poor prognosis markers. The 5-year survival rate was reported to be 46% in patients with NHL arising from Waldeyer's ring (8).

Symptomatic, early-stage, and low-grade nasopharyngeal small lymphocytic lymphomas can be treated with radiotherapy and the prognosis is relatively good (8). In the United States, the 5-year survival rate in patients with chronic lymphocytic leukemia/small lymphocytic lymphoma aged <65 years is 83%, while this rate is 68% in patients aged >65 years (3).

CONCLUSION

In this study, following biopsy and systemic evaluation, the patient diagnosed with early-stage and low-grade B-cell lymphocytic lymphoma was referred to the hematology/oncology clinic, and radiotherapy was applied. An endoscopic and radiological examination performed 24 months post-treatment did not reveal any mass. Patient's hemogram and otoscopic and audiologic evaluations were within the normal limits. The patient is still followed by both clinics using the multidisciplinary approach.

Here, an ENT (ear nose and throat) specialist was responsible for early diagnosis, proper orientation, and post-treatment endoscopic control examinations.

Informed Consent: Written informed consent was obtained from the patients who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - Ç.H.Ü., D.A., E.M., M.C.A.; Design - Ç.H.Ü., D.A., E.M., M.C.A.; Supervision - Ç.H.Ü., D.A.; Materials - Ç.H.Ü., M.C.A.; Data Collection and/or Processing - E.M., M.C.A.; Analysis and/or Interpretation - E.M., M.C.A.; Literature Search - Ç.H.Ü., D.A., E.M.; Writing Manuscript - Ç.H.Ü., D.A.; Critical Review - Ç.H.Ü., D.A., E.M., M.C.A.

Conflict of Interest: The authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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