

Primary Nasopharyngeal Hodgkin's Disease: A Case Report

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Abstract

Primary nasopharyngeal Hodgkin's disease occurs in less than %1 of all Hodgkin's disease cases. With appropriate treatment, the prognosis for this particular type of Hodgkin's Lenfoma is favorable. Most documented cases of Hodgkin's Lenfoma are either stage 1 or 2 patients. A 53-year-old male patient presented with bilateral nasal obstruction. Endoscopic examination of the nasal cavity revealed a mass arising from the nasopharynx. After the excision histopathological study reported the case to be classic Hodgkin's Lenfoma, of a nodular sclerosis type. Bone marrow core biopsy, scanning of chest, abdomen and neck were normal. Further positron emission tomographic scan also showed no abnormalities. The patient was considered to have stage 1EA nasopharyngeal Hodgkin's Lenfoma and was treated with radiotherapy and 6 cycles of chemotherapy. We aim to present this uncommon case of Hodgkin's Lenfoma confined to the nasopharynx with a review of the literature.

Key Words: Hodgkin's disease; Nasopharynx; Prognosis.

Primer Nazofarengal Hodgkin Lenfoma: Olgu Sunumu

Özet

Hodgkin lenfomaların yaklaşık olarak %1'inden azını primer nazofarengal tutulum oluşturur. Bu bölgedeki Hodgkin lenfomaların uygun tedavi ile prognozları oldukça iyidir. Bildirilen Hodgkin vakalarının birçoğu evre 1 veya 2' de saptanmıştır. Bilateral burun tıkanıklığı şikayetiyle polikliniğimize başvuran 53 yaşındaki erkek hastanın yapılan endoskopik bakısında nazofarenkste kitlesi saptandı. Hastanın eksişyon sonrası patoloji sonucu 'klasik tip Hodgkin Lenfoma-nodüler sklerozan' şeklinde raporlandı. Kemik iliği biyopsisi, toraks, abdomen ve boyun bilgisayarlı tomografi (BT) sonuçları normal izlendi. Yapılan Pozitron Emisyon Tomografi (PET) normal sınırlardaydı. Evre 1EA olarak değerlendirilen bu hastaya ardışık radyoterapi ve 6 kür kemoterapi uygulandı. Biz de bu olgunun tanı ve tedavi sürecini literatür eşliğinde sunmayı amaçladık.

Anahtar Kelimeler: Hodgkin Lenfoma; Nazofarenks; Prognoz.

INTRODUCTION

Lymphomas are the second most common malignancies in and around the head and neck. Hodgkin's lymphoma (HL) constitutes 10-35% of all the lymphoma cases in the head and neck. 70-80% of HL cases in the head and neck encompass lymph nodes while extranodal HL is often seen in and around the Waldeyer ring (1). Extranodal HL anywhere except from the Waldeyer's ring in the head and neck is very rare. Primary nasopharyngeal involvement is accounted for only 1% of all HL detected cases. With proper treatment, HL around this region may provide quite satisfactory prognosis. Many reported HL cases have been determined in stage 1 or 2.

In this case report, we aim to present the treatment process of a primary nasopharyngeal HL case through a sequential combination of chemotherapy and radiotherapy along with a review of the literature.

CASE REPORT

In February 2013, a 53-year-old male patient presented with complaints of bilateral nasal obstruction that had existed for a long time. The patient did not have any

history of smoking. Without any signs of weight loss, night sweats, itching or fever, the patient's full blood count, biochemistry, and erythrocyte sedimentation rates were also within normal limits. On physical examination, we did not find any palpable lymphadenopathy in the neck but the '0' degree endoscopic examination revealed well-circumscribed mass lesion on the posterior wall of the nasopharynx that obstructed the air column. Other ear-nose-throat examination results were normal. Nasopharyngeal magnetic resonance imaging report was as follows: "a well-circumscribed lobule narrowing the air column on the nasopharynx posterior wall was identified; contoured T1A view showed a hypointense lesion; T2A examination displayed a solid, space occupying lesion appearance with intense heterogeneous contrasts following intravenous injection of contrast material (IICM) viewed as hyperintense (histopathological diagnosis is recommended)" (Figure 1).

A punch biopsy was performed during the endoscopy and because the pathology result was 'fibroinflammatory proliferation, we excised the mass in the nasopharynx completely. Following the excision, the decisive pathology results was reported as "classical type-nodular sclerosing Hodgkin's disease" (Figure 2).



Figure 1. Transverse, sagittal, and coronal sections in the MR examination of the nasopharynx show a well-circumscribed lobule in the form of a contoured, solid lesion on the posterior wall.

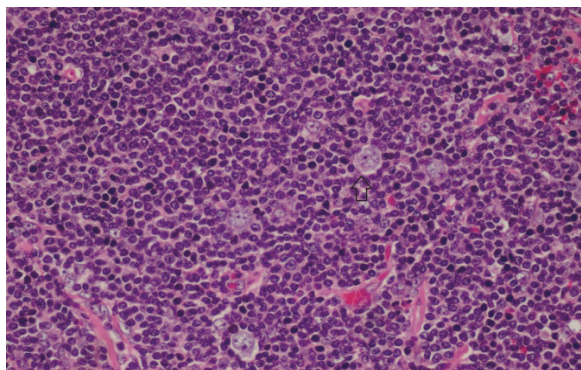


Figure 2. Histopathological results show wide tumour cells with visible nuclei, mono nuclei, and bilobule nuclei (Reed-Sternberg cell).

Bone marrow biopsy, and the computed tomography (CT) of the chest, abdomen, and neck were normal. The Positron Emission Tomography (PET) results were within normal limits. The disease was evaluated as stage 1EA according to Ann-Arbor classification. We applied 6 cycles of ABVD (Doxorubicin, bleomycin, vinblastine, dacarbazine) that was repeated once in every 30 days, Waldeyer's ring, and a total of 36 Gy of radiotherapy that would include the bilateral cervical lymph nodes. It has been two months since we started the treatment and our patient did not show any signs of recurrence during the follow-up.

DISCUSSION

Lymphomas of the head and neck region cover both nodal and extranodal lymphomas. Compared with Hodgkin's lymphoma (HL), non-Hodgkin's lymphoma (HLA) is more commonly seen in the head and neck regions (1). Whereas HL in the head and neck regions is frequently observed in nodal tissues. Extranodal involvement is very rare (2). Quite uncommon in HL patients, primary nasopharyngeal involvement constitutes only less than 1% of all the reported cases of HL. Eavey and Goodman have identified nasopharyngeal

HL in only two patients in their study of 500 patients with HL in the head and neck regions (3). Similarly, Anselmo et al. report only 7 nasopharyngeal HL cases in their 24-year-long study on 2150 patients (4). Bjorklund et al., after performing nasopharynx biopsy on 45 patients with systematical Hodgkin's disease, have identified 7 cases with nasopharyngeal involvement (16%). Therefore, they argue that nasopharyngeal biopsy should be considered as a part of HL staging (5).

All over the world, literature reports less than 90 nasopharyngeal involvement cases in HL patients and only 20 of these cases primarily cover the nasopharyngeal region (6). HL usually occur in men and its most common histological subtype is mixed cellularity (4, 7). Nasopharyngeal HL should be distinguished from EBV-related lymphoproliferation because EBV was positive in a large proportion of the reported HL cases.

To confirm the diagnosis, pathological verification is needed; however, final diagnosis should follow further immunohistochemical analysis. CD 30 (+) and/or CD 15 (+) Reed-Stenberg cells are seen in the majority of cases. Besides, CD 20, CD 3, and CD-4 have negative expression.

The treatment of isolated nasopharyngeal involvement in HL is not different from other HLA localisations. The general treatment protocol is sequential chemotherapy and radiotherapy (8). We think that 6 cycles of ABVD (Doxorubicin, bleomycin, vinblastine, dacarbazine) is the most appropriate treatment along with Waldeyer's ring and 25-40 Gy of radiotherapy targeted at cervical lymph nodes (9).

Briefly, the nasopharynx HL is very atypical and involves a rare localisation among HL. It should definitely be considered in differential diagnosis of nasopharyngeal masses. Most cases are stage 1 or 2 and can be cured through appropriate treatment. Combined chemoradiotherapy is the ideal method of treatment.

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