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CASE REPORT / OLGU SUNUMU

Recurrence of nasopharyngeal carcinoma in the mastoid region: Therapeutic approach of a rare clinical entity

Nazofarenks kanserinin mastoid bölge rekürrensi: Nadir bir klinik tablonun tedavisi

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ABSTRACT

Mastoid region recurrence is a rare entity for nasopharyngeal cancer. This paper presents a patient with an isolated mastoid recurrence following definitive treatment and discusses the therapeutic approach which may help in the treatment of such cases.

Keywords: Chemotherapy, Mastoid region recurrence, Nasopharyngeal carcinoma, Radiotherapy

ÖZET

Nazofarenks kanserinde mastoid rekürrens takipte nadir izlenen bir durumdur. Bu makale definitif tedavi sonrası izlenen izole mastoid nüksünü sunmakta ve tedavisini tartışmaktadır.

Anahtar kelimeler: Kemoterapi, Mastoid bölge nüksü, Nazofarenks kanseri, Radyoterapi

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Introduction

Nasopharyngeal carcinoma (NPC) is a highly radiosensitive tumor with a variable range of incidence due to geographic localization, for example Southern Asia and Mediterranean countries [1]. It is the second most frequently treated head and neck malignancy following laryngeal carcinoma in reference hospitals in Turkey [2]. Most of the patients present in advanced stages. Local control and survival rates have increased up to 90 percent with concomitant chemo-radiotherapy (CRT) [3]. A common pattern for the local and regional spread of recurrences is recurrence in the nasopharyngeal and parapharyngeal area; the base of skull, paranasal sinuses or orbita. Additionally, all cervical lymphatic regions are under risk of recurrence at any time during follow-up.

The mastoid region is not affected by NPC tumor cells, either initially or at the time of recurrence. There are currently a small number of reports mentioning this issue in the literature [4-6]. Here, we report a patient with an isolated mastoid recurrence following definitive treatment. This article also discusses a therapeutic approach which may help in the treatment such cases.

Case Report

A 41-year-old man presented with a right neck mass. A contrast-enhanced nasopharyngeal mass was detected by endoscopic evaluation and a computed tomography (CT) scan. The nasopharyngeal biopsy revealed a non-keratinizing type of squamous cell carcinoma of the nasopharynx. He had no cranial nerve palsy at the time of diagnosis and CT. He also showed a right middle ear effusion due to tumor obstruction in front of the Eustachian tube. Multiple lymph nodes were detected in the submental, bilateral submandibular and posterior cervical regions, the largest diameter being 4.5 cm. No distant metastasis was detected in the thorax and abdominal CT and bone scintigraphy. Therefore, the case was diagnosed with Stage III (T1N2cM0) NPC according to the 2002 AJCC staging system [7]. The patient received two courses of induction chemotherapy (cisplatin, 100 mg/m^2 and 5-fluorouracil 1000 mg/m^2, every three weeks) followed by definitive radiotherapy (70 Gy to the nasopharynx, 66 Gy to positive lymphatic region and 46 Gy to all cervical and supraclavicular regions at 2 Gy/fraction/5 fractions per week).

One course of concurrent cisplatin (80mg/m²) was administered and it was stopped due to grade 3 renal and constitutional side effects. A clinical and radiological complete response was achieved at the sixth months of follow-up.

Thirty-three months later he had a right ear discharge and pain. Facial nerve palsy was detected in the clinical examination and magnetic resonance imaging (MRI) showed a mass in the right mastoid bone, filling the mastoid air cells and extending to the right temporal lobe of the brain (Figure 1). Positron emission tomography (PET)/CT scan revealed a hypermetabolic activity in the right temporal bone region extending to the parenchyma (Figure 2). No further local or distant recurrence was detected. An open biopsy was taken from the mass in the mastoid bone and the pathology was confirmed as the same pathology of the primary tumor. A partial radiological response was achieved after three cycles of chemotherapy (docetaxel 75 mg/m^2, carboplatin 60 mg/m^2 and 5-fluorouracil 1000 mg/m^2, every three weeks). Then the patient received radiotherapy with a dose of 30 Gy in 10 fractions. This patient showed a complete clinical and radiological response in the 74th month of follow-up.

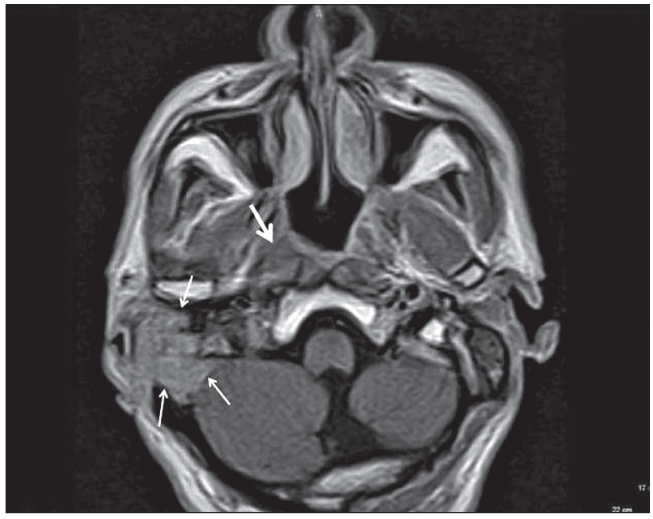

Figure 1. An axial fat saturation T1-weighted MR image with IV contrast shows the tumor with a contrast enhancement in mastoid region (thin arrows) and Eustachian tube (thick arrow).

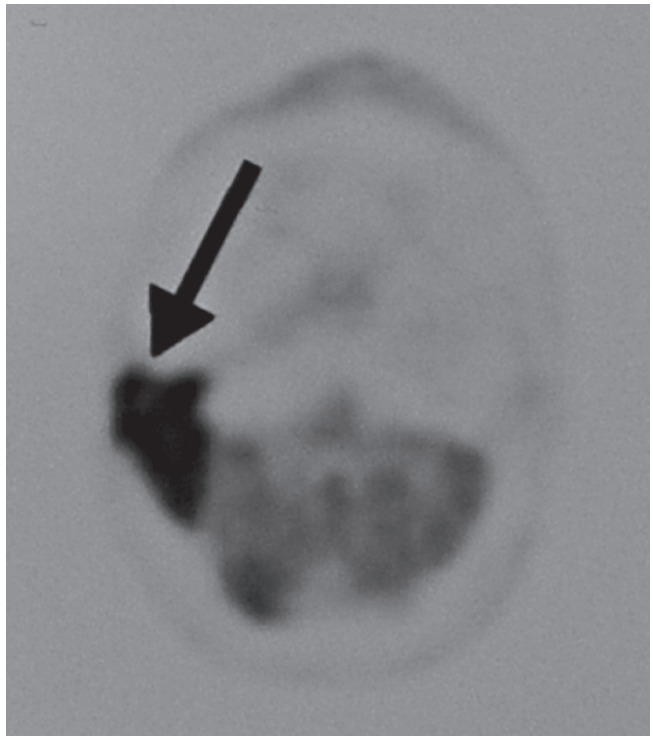

Figure 2. An axial PET/CT image shows the hyper metabolic activity in mastoid region.

Discussion

NPC cancer cells can easily disseminate locally in many ways into the nearby critical structures. Thus, the local spread of the tumor is usually observed as direct extension anteriorly to the nasal cavity and orbita; superiorly and posteriorly to the base of the skull, sphenoid sinus and clivus; inferiorly to the oropharynx and/or laterally into the lateral parapharyngeal muscles [6]. The standard treatment volumes encompass the primary tumor with its extension to these areas, the nasopharynx, almost the whole cervical lymph node levels and the supraclavicular lymphatic area. However, the middle ear and mastoid cellular region are not included in any of the radiotherapy target volumes for treatment planning.

The Eustachian tube is located beside the nasopharynx and it connects the lateral wall of the nasopharynx to the middle ear. Tumor cells can invade the Eustachian tube but surprisingly the tumor cell migration into the middle ear is not observed. We found only two reports which described mastoid region recurrence in the English literature [4,5]. Yang et al [5] reported a 44-year old male patient with right facial nerve palsy and right mastoiditis symptoms after 24 months of CRT. An MRI revealed a soft tissue mass in the right mastoid bone and temporal cavity spreading along the Eustachian tube. Surgery was performed and the patient was diagnosed with metastatic squamous cell carcinoma. Zhang et al [4] reported two cases of NPC with mastoid recurrence. The first case was a 45-year old man with NPC initially staged as T2N2M0. The primary tumor received 70 Gy conventional radiotherapy and the Eustachian tubes were included in the radiation treatment plan. Parts of the temporal bones were outside the fields. At the 12th month of follow up, the patient presented with localized redness, swelling and pain in the mastoid area. Recurrence was detected by biopsy and the tumor was irradiated with 69.96 Gy in 33 fractions. Despite the response to this treatment, multiple bone metastases were detected in the follow-up. After four cycles of vinorelbine and cisplatin-based chemotherapy, a progression free survival was achieved for 3 years. The second NPC patient was a 43-year old woman with T3N1M0 disease [4]. Radiation therapy was performed by intensity-modulated radiotherapy (72 Gy) and a part of both temporal bones received 50 Gy. Sixteen months later, a mastoid area recurrence was observed by MRI and biopsy. However, three months later she died of multiple liver and retroperitoneal lymph nodes metastases.

To our knowledge, this is the fourth NPC case presented with mastoid region recurrence. Our hypothesis is that the tumor cell migration occurred through the Eustachian tube into the mastoid

region. Despite the close anatomical relation between the Eustachian tube and the middle ear it is not clear why this issue is rare in clinical practice. Middle ear effusion is a common late side effect following radiotherapy. Radiological findings of tumor involvement can easily be confused with middle ear effusion. Therefore, we think that biopsy should be considered in all suspicious cases.

In conclusion, although middle ear effusion and mastoiditis are commonly seen in NPC patients, the tumor seeding into the middle ear is an extraordinary site for recurrence after definitive therapy. Patients should be evaluated carefully not only for radiation-induced side effects but also for an extraordinary site of tumor recurrence. In the previous reports, there is also a strong association with mastoid recurrence and distant metastasis and we think that systemic therapy should be considered for these cases and they should be multidisciplinary treated according to a tailored therapy.

Conflicts of interest

None declared.

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