



## A Case Report of Nasopharyngeal Neuroendocrine Carcinoma

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### Abstract

**Background:** The purpose of this article is to report the treatment outcome of a patient with nasopharyngeal neuroendocrine carcinoma.

**Method:** The patient received concurrent Chemoradiotherapy (CRT) for the primary lesion and microwave ablation for isolated hepatic metastasis later.

**Results:** Complete Remission (CR) was achieved after the completion of concurrent CRT. However, Prophylactic Cranial Irradiation (PCI) was not performed. Isolated hepatic metastasis was diagnosed 3 months after the completion of CRT. Then microwave ablation for isolated hepatic metastasis was performed. Finally, unlike the most common death caused by brain metastasis in literature retrieval, the patient died of hepatic function failure caused by multiple hepatic metastasis. The Overall Survival (OS) of the patient is about 18 months.

**Conclusion:** Standardized concurrent CRT also has a good therapeutic effect on nasopharyngeal neuroendocrine carcinoma. The administration of prophylactic cranial irradiation may not be necessary for nasopharyngeal neuroendocrine carcinoma, which remains to be confirmed.

**Keywords:** Nasopharyngeal carcinoma; Neuroendocrine tumors; Concurrent chemoradiation; Prophylactic cranial irradiation

### Introduction

Neuroendocrine carcinoma is a highly malignant tumor, which often arise in lung while relatively rare in the nasopharyngeal region. Limited information of treatment for nasopharyngeal neuroendocrine carcinoma have been reported because of its rarity. This is a case report about the treatment outcome of a patient with nasopharyngeal neuroendocrine carcinoma who was treated with CRT and microwave ablation for Isolated hepatic metastasis later.

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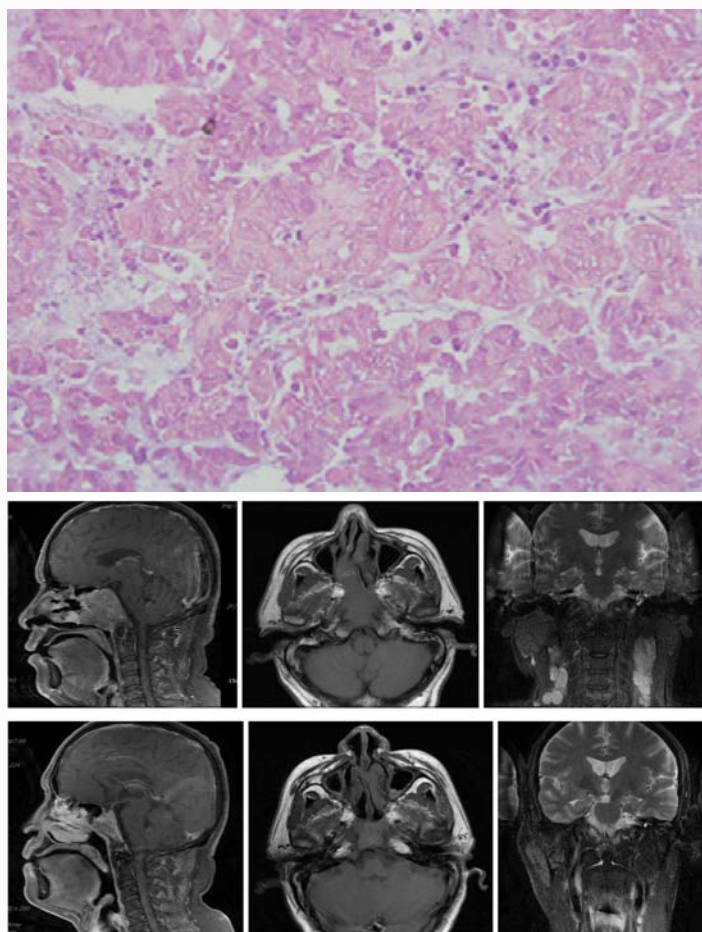
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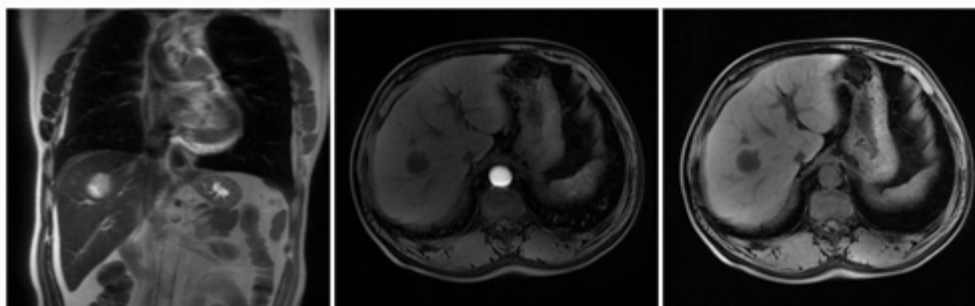
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### Case Presentation

A 58-year-old man complained of the gradual onset of an occlusive feeling in the nose, pus or bloodstain in the nasal discharge and hearing loss for approximately 1 year. Nasopharyngoscopy revealed a mass that was located in the right wall of the nasopharynx, and neuroendocrine carcinoma was suspected based on the tissue biopsy which was performed in the local hospital. Light microscopy after hematoxylin and eosin staining showed that the tumor was poorly differentiated neuroendocrine carcinoma (Figure 1A). The patient was referred to our hospital for treatment. Repeated biopsy was not performed because of the definite diagnosis in the local hospital. For imaging studies, CT, MRI were performed. The tumor invaded Sella turcica, cavernous sinus, clivus and spread to bilateral cervical lymph nodes without distant metastasis (Figure 1B). Therefore, the tumor was classified as cT3N3M0 according to the eighth edition of the Union for International Cancer Control TNM Classification [1]. The patient received a cycle chemotherapy consisted of etoposide (400 mg) plus cisplatin (120 mg), closely followed by Intensity-Modulated Radiation Therapy (IMRT) with a dose of 6600c Gy in 33 fractions over 7 weeks for the primary tumor and positive lymph nodes, whereas conventional fractionated RT was performed with a dose of 5940c Gy in 33 fractions over 7 weeks for the upper neck region and a dose of 5445c Gy in 33 fractions over 7 weeks for the lower neck region. Meanwhile, 3 cycles of concurrent chemotherapy was performed, which consisted of etoposide (400 mg) plus cisplatin (120 mg). According to the Response Evaluation Criteria in Solid Tumors (version 1.1), CT and MRI for imaging studies showed that complete remission was achieved after the completion of the CRT. Prophylactic cranial irradiation was not performed because of the upper limit of brainstem radiation in the further treatment. Isolated



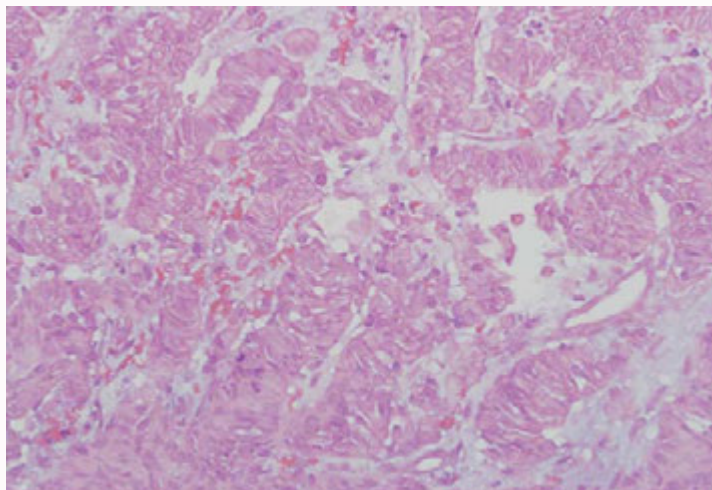
**Figure 1:** The cancer tissue was in a cord-like and adenoid arrangement, and the mitoses were easy to see (hematoxylin and eosin stain, original magnification, x400) (Figure 1A). The tumor was located in the right wall of the nasopharynx, invaded the Sella turcica, cavernous sinus, clivus and spread to bilateral cervical lymph nodes without distant metastasis. Complete response was achieved after the completion of concurrent chemoradiotherapy (Figure 1B).



**Figure 2:** The MR showed the isolated hepatic metastasis which originated from the nasopharynx.

hepatic metastasis was diagnosed 3 months after the completion of CRT, the maximum diameter of which was 2.5 cm (Figure 2). The puncture pathology showed an isolated hepatic metastasis of neuroendocrine carcinoma originated from the nasopharynx, which suggested the progression of the disease (Figure 3). The Progression-Free Survival (PFS) was 11 months. Then the patient received 2 cycles chemotherapy consisted of irinotecan plus cisplatin. Because of significant bone marrow suppression, 2 cycles irinotecan single agent chemotherapy was performed later. MRI for imaging studies showed the maximum diameter of the isolated metastasis reduced to 1 cm after the completion of 4 cycles of irinotecan for induction chemotherapy. Then ultrasound guided microwave ablation for

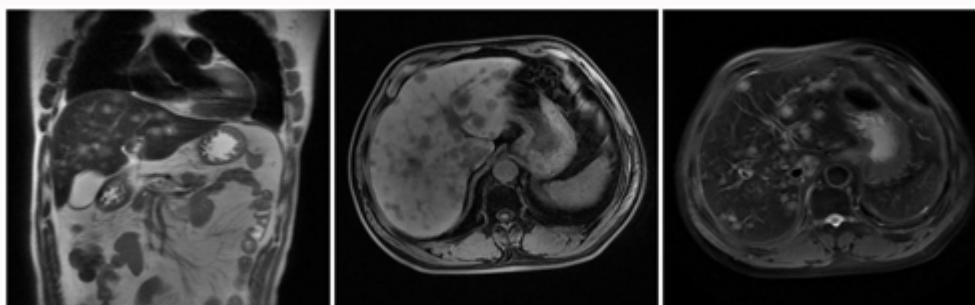
isolated hepatic metastasis after Multi-Disciplinary Team (MDT) and CR was achieved (Figure 4). Irinotecan single-agent chemotherapy was performed periodically after the operation for maintenance treatment. The MR showed multiple enlarged retroperitoneal lymph nodes after 2 months (Figure 5). Then we chose Apatinib in the third line therapy. Multiple hepatic metastasis was diagnosed 1 month after the target therapy, which suggested the resistance to targeted drugs and the progression of the disease again. Due to the poor general condition of the patient, with a PS score of 3, anti-tumor therapy was stopped and hospice care was given. This disease is a highly aggressive tumor with a very poor prognosis. The patient finally died of liver failure caused by multiple liver metastases 2 months after stopping



**Figure 3:** The cancer tissue was in a cord-like and adenoid arrangement, and the mitoses were easy to see, originated from the nasopharynx (hematoxylin and eosin stain, original magnification, x400).



**Figure 4:** The CT showed complete response was achieved after the completion of the microwave ablation.



**Figure 5:** The MR showed multiple enlarged retroperitoneal lymph nodes after 2 months of Irinotecan single-agent chemotherapy.

the anti-tumor treatment. The OS of the patient is about 18 months.

## Discussion

Neuroendocrine tumors which involve the head and neck, especially the nasopharynx, is extremely rare. The diagnosis and treatment are still challenging for modern medicine. Combined with the currently retrieved literature, extrapulmonary neuroendocrine carcinoma is similar to intrapulmonary neuroendocrine carcinoma due to its pathological and clinical manifestations, and the current treatment methods are also similarly [2]. This patient was treated with concurrent CRT in the first line therapy. The primary lesion continued to be CR after initial treatment until the death of the patient. However, because of the high degree of malignancy and a

high distant metastasis rate of this disease, the patient developed isolated hepatic metastasis later. The ultrasound guided microwave ablation for the isolated metastasis was performed after 2 cycles of chemotherapy. Imaging studies showed that complete remission was achieved after the ablation. However, the patient eventually died of liver failure caused by further multiple liver metastases. The OS of the patient is about 18 months, which is at an intermediate level among the current literature results [3-10]. According to the literature retrieved so far, nasopharyngeal neuroendocrine carcinoma has the risk of central nervous system metastasis, and it is necessary to carry out prophylactic cranial irradiation [4]. But this patient did not receive PCI because of the upper limit of brainstem radiation. However, the imaging examinations during the entire

survival period showed that the patient did not have central nervous system metastasis. Therefore, according to failure pattern, in this case, prophylactic cranial irradiation is not necessary. So, whether nasopharyngeal neuroendocrine cancer requires prophylactic cranial irradiation remains to be further studied.

Nasopharyngeal neuroendocrine carcinoma is a rare malignant tumor with the clinical manifestations of high-degree malignance, high invasion, high rate of relapse and highly distant recurrence. Due to its rarity, the treatment and prognosis are still a difficult problem in our current research, and more cases are needed to observe and study.

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