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Case Report

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A rare case of olfactory neuroblastoma

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ABSTRACT

Olfactory neuroblastoma, also known as esthesioneuroblastoma, is a rare tumor of the nasal cavity with an incidence of approximately 0.4 per million individuals. The most common symptom is nasal obstruction, although patients may also present with epistaxis and nasal discharge. Treatment options primarily include surgical resection and radiotherapy, with a limited role for chemotherapy. There is currently no universally accepted treatment protocol for this condition. This report describes a case of olfactory neuroblastoma staged as Kadish stage C, in which the patient underwent chemoradiotherapy. Follow-up imaging revealed no evidence of disease recurrence. This case suggests that chemotherapy may play a role in the treatment regimen for advanced-stage olfactory neuroblastoma, potentially improving therapeutic outcomes.

Keywords: Olfactory neuroblastoma, chemotherapy, radiotherapy

INTRODUCTION

Olfactory neuroblastoma (ONB), also referred to as esthesioneuroblastoma (ENB), is a rare tumor originating from the olfactory neuroepithelium in the nasal cavity.[1] Its incidence is approximately 0.4 per million and it accounts for about 2% of tumors in the nasal cavity and paranasal sinuses.[2] The tumor is most commonly diagnosed in the fifth and sixth decades of life.[2,3] Although it is more frequent in males, the gender distribution is nearly equal.[2,3] The most common symptom of ONB is nasal obstruction.[2] Other frequent symptoms include epistaxis and nasal discharge.[3] Additionally, symptoms such as anosmia, excessive lacrimation, headache, and ear pain may occur due to local tumor invasion.[2,3] Paraneoplastic syndromes such as hypercalcemia and inappropriate antidiuretic hormone (ADH) syndrome are extremely rare.[3] The rarity of the tumor and its symptoms overlapping with those of upper respiratory tract infections often lead to delayed diagnosis.[1] Consequently, delayed diagnosis is common.[2-4]

Treatment for olfactory neuroblastoma generally involves surgical resection and radiotherapy (RT), with chemotherapy (CT) playing a limited role.[5] There is no established universally accepted treatment protocol.[4] It is suggested that chemotherapy may become part of the treatment regimen in advanced stages.[6] This report presents a case of a patient with Kadish stage C ONB and details the treatment plan.

CASE

A 40-year-old male patient presented to the otolaryngology clinic with persistent nasal obstruction and headaches lasting approximately 6 months. Despite receiving prolonged medical treatment for what was initially suspected to be an upper respiratory tract infection, there was no improvement in his symptoms.

Paranasal sinus computed tomography revealed soft tissue densities in the right nasal cavity, causing air loss in the ethmoid cells with indistinct borders from the middle and inferior turbinates, and extending from the accessory ostium of the right maxillary sinus into the nasal cavity, partially obstructing the nasal passage and extending to the choana (**Figure**).

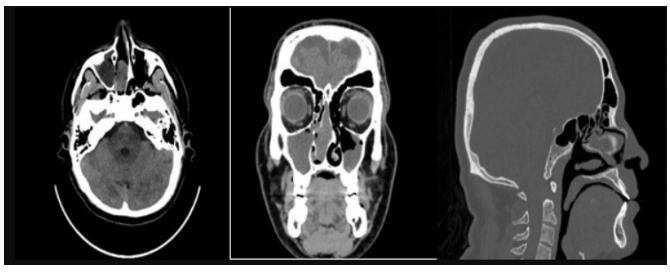

Figure. Paranasal sinus computed tomography of the patient

The brain tomography of the patient, in whom no pathological findings were detected, revealed that the punch biopsy taken from the mass was reported as ONB. Additionally, there were no PIK3CA or p53 mutations. The tumor, which invaded the posterior and anterior ethmoid cells and extended to the frontal recess, was excised via curettage through the skull base. Following surgery, the Kadish stage C patient was evaluated in the head and neck tumor board. The treatment plan included 33 fractions of radiotherapy totaling 66 gray, with concomitant systemic chemotherapy (weekly cisplatin at 40 mg/m2). During the treatment, the patient experienced minor symptoms such as dryness of the oral and nasal mucosa due to RT, and nausea

and vomiting from chemotherapy. However, these side effects did not interrupt the treatment, which continued as planned. Imaging three months after treatment showed no evidence of disease recurrence.

DISCUSSION

ONB, also known as ENB, is an extremely rare tumor.[1] It originates from olfactory neuroepithelial cells.[1] Despite its slow growth, the non-specific symptoms often lead to delayed diagnosis.[1,2] Although it can present between the ages of 35 and 70, the most common presentation age is in the fifth decade.[3] Nasal obstruction is the most frequent symptom, but epistaxis, headache, facial swelling, and ear pain can also be observed.[3]

Histopathological features may include mutations in PIK3CA, p53, NF1, and CDKN2A. Immunohistochemical markers such as vimentin, S-100 protein, neuron-specific enolase, and neurofilaments are used for diagnosing neuroblastoma.[7]

Kadish staging is the most commonly used clinical staging system for ONB.[8] Kadish A indicates the tumor is confined to the nasal cavity; Kadish B includes the nasal cavity plus one or more paranasal sinuses.[8] Kadish C indicates extension beyond the paranasal sinuses.[8] Kadish D involves regional lymph node metastasis or distant metastases.[8]

Treatment for ONB involves surgery, RT and CT.[3] Due to the tumor's rarity and the need for long-term monitoring of treatment outcomes, there is no established standard treatment protocol.[3] The role of CT remains undetermined, and data on the benefits of neoadjuvant CT or concurrent chemoradiotherapy are limited.[3,4] CT should be considered as part of the treatment regimen for advanced stages, such as Kadish C-D, or in cases where surgery is not feasible due to widespread intracranial metastases or orbital invasion.[9-12]

CONCLUSION

ONB is a very rare tumor with no established standard treatment protocol. Data on the benefits of CT are limited. CT should be considered as part of the treatment regimen in advanced stages or when surgery is not possible. More studies are needed due to the very low incidence and the small number of patients in the studies.

ETHICAL DECLARATIONS

Informed Consent

The patient signed and free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

No financial support has been received for this article.

Financial Disclosure

The authors declare that this study has received no financial support.

Author Contributions

All authors declare their participation in the design, execution, and analysis of the paper, and that they have approved the final version.

REFERENCES

1. Ward PD, Heth JA, Thompson BG, Marentette LJ. Esthesioneuroblastoma: results and outcomes of a single institution's experience. *Skull Base*. 2009; 19(2):133-140.
2. Batchu S, Gill AS, Karsy M. Characterizing immune infiltration in esthesioneuroblastoma subtypes through gene expression deconvolution. *World Neurosurg*. 2024;183:e928-e935.
3. Kuan EC, Nasser HB, Carey RM, et al. A population-based analysis of nodal metastases in esthesioneuroblastomas of the sinonasal tract. *Laryngoscope*. 2019;129(5):1025-1029.
4. Gabbay U, Leider-Trejo L, Marshak G, Gabbay M, Fliss DM. A case and a series of published cases of esthesioneuroblastoma (ENB) in which long-standing paraneoplastic SIADH had preceded ENB diagnosis. *Ear Nose Throat J*. 2013;92(10-11):E6.
5. Chamberlain MC. Treatment of intracranial metastatic esthesioneuroblastoma. *Cancer*. 2002;95(2):243-248.
6. Weiden PL, Yarington CT, Richardson RG. Olfactory neuroblastoma: chemotherapy and radiotherapy for extensive disease. *Arch Otolaryngol*. 1984;110(11):759-760.
7. Gay LM, Kim S, Fedorchak K, et al. Comprehensive genomic profiling of esthesioneuroblastoma reveals additional treatment options. *Oncologist*. 2017;22(7):834-842.
8. Kadish S, Goodman M, Wang C. Olfactory neuroblastoma-a clinical analysis of 17 cases. *Cancer*. 1976;37(3):1571-1576.
9. Cranmer LD, Chau B, Rockhill JK, Ferreira M, Liao JJ. Chemotherapy in esthesioneuroblastoma/olfactory neuroblastoma: an analysis of the Surveillance Epidemiology and End Results (SEER) 1973-2015 database. *Am J Clin Oncol*. 2020;43(3):203-209.
10. Eich HT, Hero B, Staar S, et al. Multimodality therapy including radiotherapy and chemotherapy improves event-free survival in stage C esthesioneuroblastoma. *Strahlenther Onkol*. 2003;179(4):233-240.
11. Fitzek MM, Thornton AF, Varvares M, et al. Neuroendocrine tumors of the sinonasal tract: results of a prospective study incorporating chemotherapy, surgery, and combined proton-photon radiotherapy. *Cancer*. 2002;94(10):2623-2634.
12. Miller KC, Marinelli JP, Van Gompel JJ, et al. Utility of adjuvant chemotherapy in patients receiving surgery and adjuvant radiotherapy for primary treatment of esthesioneuroblastoma. *Head Neck*. 2019;41(5):1335-1341.