Content available at: https://www.ipinnovative.com/open-access-journals

IP Indian Journal of Anatomy and Surgery of Head, Neck and Brain

Journal homepage: https://www.ijashnb.org/

Case Report

Olfactory neuroblastoma: A case report

Sunil Kathuria[1],*, Chikku Sunny[1]

[1] *Dept. of ENT, Batra Hospital and Medical Research Centre, New Delhi, India*

ARTICLE INFO

Article history:
Received 23-02-2022
Accepted 08-03-2022
Available online 02-04-2022

Keywords:
Sinonasal Tract
Olfactory Neuroblastoma
Nasal Cavity

ABSTRACT

Olfactory neuroblastoma (ON) is a rare malignant neuroectodermal tumor of the nasal cavity. The malignancy accounts for <3% of tumours originating in the nasal cavity. Through the nasal cavity, ON may infiltrate the sinuses, the orbit and the cranium. The tumour is characterized by a pattern of slow growth and local recurrences. Olfactory neuroblastoma is a neoplasm that can histologically mimic many tumours within the sinonasal tract. Treatment options are surgical excision combined with a radiotherapy and/or chemotherapy combination treatment.

The present study reports the case of a 45 year-old male patient with a mass in the nasal cavity who was treated by combined surgical excision and radiotherapy. The literature for ON and the treatment of the tumour are also discussed.

This is an Open Access (OA) journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprint@ipinnovative.com

1. Introduction

Olfactory neuroblastoma (ON) is an uncommon malignant nasal tumour which is originated from neuro ectoderm. It comprises approximately 2% of all sinonasal tract tumours. Its incidence is 0.4 per million in population.[1,2] Previously called esthesioneuroblastoma, olfactory placode tumour, esthesioneurocytoma, esthesioneuroepithelioma, and esthesioneuroma, these terms highlight the sensory (olfactory) and primitive neuroectodermal origins, although the use of these older terms is discouraged. Although rare, but cases primarily arising from other parts of nasal cavity, paranasal sinuses and frontal lobe of brain have also been reported

The most common symptoms of ON are unilateral nasal obstruction (70%), and epistaxis (50%). Other symptoms include headaches, pain, excessive lacrimation, rhinorrhea, anosmia and changes in vision. Even though ON originates from olfactory epithelium, it rarely causes anosmia (5%).[3,4] Metastasis to distant as well as cervical lymph nodes occurs in 10 to 30% of the cases.[5]

The management of ON requires bicranial-facial surgical approach, trephination procedure, which is technically challenging and achieving good results are difficult. Treatment modalities for ON are en bloc resection, extra cranial resection or surgery combined with radiotherapy and/or chemotherapy.

The present study reports the case of a patient with a mass in the nasal cavity who was treated by combined surgical excision and radiotherapy.

2. Case Report

A 45-year-old male patient presented to Batra Hospital and Medical Research Centre, New Delhi with chief complaints of unilateral (left sided) nasal bleeding, nasal obstruction with blurring of vision since 1 month. His blood profile for biochemistry and haematology was within normal limits.

Endoscopy showed the presence of a mass with blood clots within the nasal cavity. Upon Computed tomography

* *Corresponding author*.
E-mail address: kathurias1@rediffmail.co (S. Kathuria).

https://doi.org/10.18231/j.ijashnb.2022.006
2581-5210/© 2022 Innovative Publication, All rights reserved.

scan (CT) of paranasal sinuses, a mucopolypoidal and soft tissue density was noted in bilateral frontal, left ethmoid, left maxillary and bilateral sphenoid sinuses with widening of left osteomeatal complex with extension in anterior cranial fossa and left orbit -?? Fungal or Malignant etiology.(Figure 1 andFigure 2)

The patient underwent transnasal endoscopic removal of the mass from the nasal cavities and paranasal sinuses from below through a skull base surgery. Tissue was sent for biopsy. On histopathological examination (HPE), the tumor was composed of prominent small nested pattern of small dark cells with blood vessel rich fibromyxoid intervening stroma. Lobules were cellular with nuclear crowding.Tumour cells showed mild nuclear pleomorphism with round to oval nuclei, darkly clumped chromatin and scanty ill defined cytoplasm, Mitoses was seen. Presence of metaplastic squamous epithelium was noted trapped within tumour substance. The histopathological report was conclusive of "poorly differentiated high grade malignant tumour with squamous differentiation."

On immunohistochemistry (IHC), the tumour cells were positive for synaptophysin, chromogranin and CD56 suggestive of Olfactory Neuroblastoma**.**

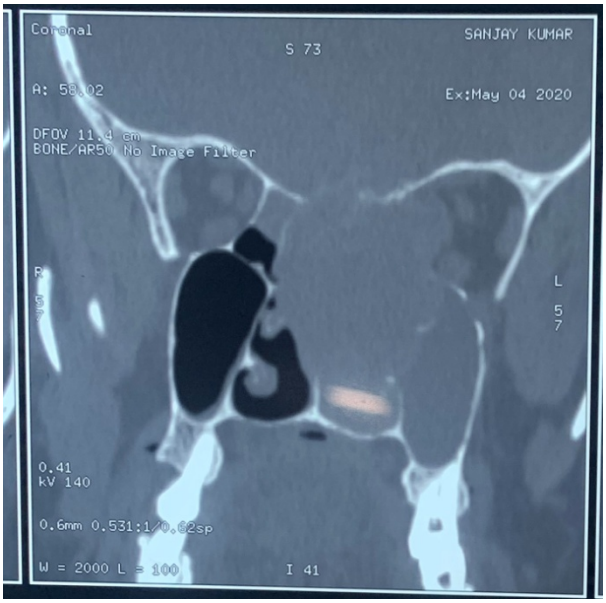

Fig. 1: Pre operative CT scan of theparanasal sinuses – coronal view showing dumbbell shaped mass in the left nasal cavity with extensions into the anterior cranial fossa, orbit and paranasal sinuses. The 'waist' of the dumbbell is at the cribriform plate.

Nasal Pack was removed on third postoperative day.Functional endoscopic sinus surgery was done after three weeks for residual tumour evaluation. Under endoscopic guidance, nasal cavity, ethmoid sinuses, sphenoid sinuses visualised and suction clearance of the crusts done.Residual tumour noted in cribriform plate

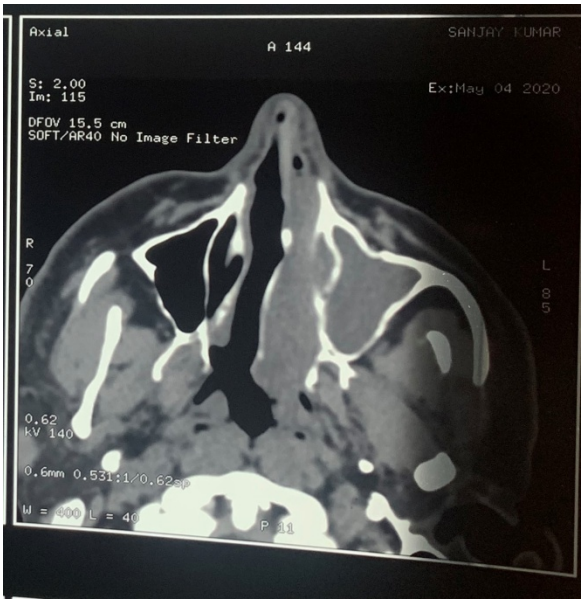

Fig. 2: Preoperative CT scan of the paranasal sinuses – axial view.

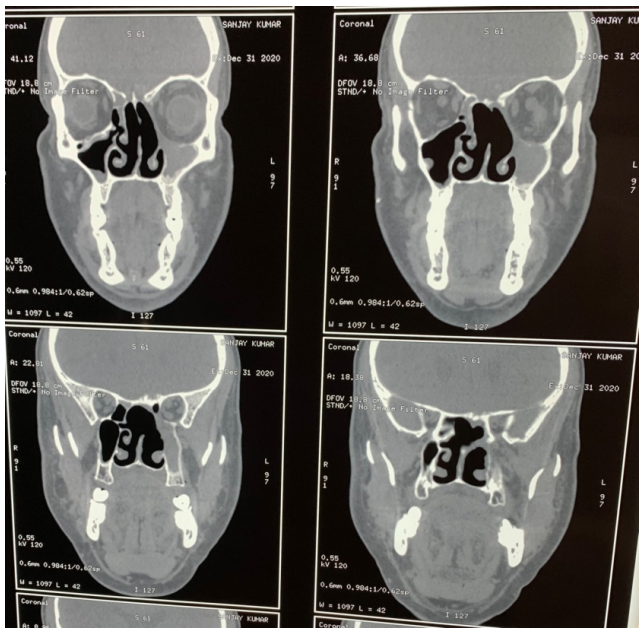

Fig. 3: CT scan ofthe paranasal sinuses repeated 6 months after the surgery showing complete resolution of mass.

from which biopsy was taken.Biopsy was positive for malignancy.

Immediate post-operative Computed Tomography scan of the paranasal sinuses showed mild soft tissue density noted in cribiform plate, inferior wall of sphenoid sinus and in the posterior choana. Patient was planned for concurrent chemoradiotherapy.Patient received 5 cycles of chemotherapy with Inj Cisplatin from 09-06-2020 to 07-07-2020.

The patient is in the 6^{th} month after diagnosis and a complete response has been observed after post-operative treatment.PET CT scan and Repeat Computed Tomography scan of the paranasal sinuses done which showed complete resolution of mass.(Figure 3)

3. Discussion

Olfactory neuroblastoma (ONB) is a rare malignant neuroectodermal tumor of the nasal cavity. Berger and Luc first described this uncommon neoplasm in 1924.[5] Although it can be found in all age groups, it occurs more commonly in the 3^{rd} and 6th decades of life, and is present equally in each gender.

The most common symptoms are unilateral nasal obstruction and epistaxis, while rhinorrhea and anosmia may also occasionally accompany these symptoms. Extensive lesions may cause frontal headaches and diplopia.[6] ON can spread quickly and easily into the intracranial structures via the cribriform plate. Cribriform plate and orbit involvement in ON are important prognostic factors.[7]

Studies have been conducted in order to evaluate treatment and prognosis of ON staging. Computed tomography and MRI are important in staging. Kadish et al.[8] performed the first staging of ON using clinical evaluation and neuroradiological findings. According to this staging, stage A is tumor limited to the nasal cavity, stage B is tumor limited to the nasal cavity and one or more of the paranasal sinuses, and stage C is tumor extending from the nasal cavity and paranasal sinuses. Morita et al. modified the Kadish staging to include 4 groups.[9] Group D was added to the Kadish classification, which included all patients of ON with metastasis to the cervical lymph nodes or distant sites.

Neck metastasis, at presentation, occurs in 5–8% of the patients, although the incidence of neck metastasis in patients with ON is 20–25%.[10] The most common site of neck metastasis in patients of ON is the cervical lymph nodes (Level II lymph nodes are the most frequently involved followed by level I, level III, and the retropharyngeal group of lymph nodes).[10] When neck nodes are involved in the disease process, the treatment should include a selective neck dissection followed by adjuvant radiotherapy to the neck.[10] Distant metastasis occurs in 12–25% of these patients with lung, brain, and bone being the most common sites.[11]

On imaging studies, a “dumbbell shaped” mass extending across the cribriform plate is one of the most characteristic findings of this tumour. The upper portion of the dumbbell-shaped mass is in the anterior cranial fossa whereas the lower portion is in the nasal cavity with the “waist” at the cribriform plate.On IHC studies, ONs are positive for Neuron Speific Enolase, synaptophysin, chromogranin, CD56, and neurofilament protein (NFP).

Subsequent to the staging of ON, multidisciplinary approaches have been used for treatment, the aim of the treatment should prevent local and regional recurrences, and distant metastasis. Treatment options consist of surgery or RT only, surgery and RT, surgery and chemotherapy combined with RT, or only chemotherapy.[12] ON is believed to be a radiosensitive tumour. Radiation therapy is usually given in the adjuvant setting, and the dose received is usually less than 60 Gy. Chemotherapy may have a role in patients with advanced disease.A craniofacial resection has been suggested for all patients with frontal cranial base involvement.[13] It has been reported that, in selected patients, endoscopic sinus surgery and stereotactic radiosurgery lead to good results.[14]

Walch et al obtained tumour control without any patient mortality by combining stereotactic radiosurgery and endoscopic sinus surgery in individuals with stage B and C disease according to the Kadish classification.[8] In the present patient, endoscopic sinus surgery with a wide local excision was performed post-diagnosis, and RT was subsequently applied. ON may metastasize and reoccur following its removal,[12] so post-operative therapy should be added to the treatment. Even though chemotherapy and RT treatments are routine for stage C disease, the study by Benfari et al.[14] indicated that RT should be applied to all patients, with the exception of cases with tumours limited to the cribriform plate without bony destruction.

Montava et al.[15] emphasized that the gold-standard treatment for ON is craniofacial resection and that mortality is associated with RT.

A study at the Massachusetts Eye and Ear Infirmary and Massachusetts General Hospital, Harvard medical school concluded that ON can be safely and effectively treated with craniofacial resection followed by proton beam irradiation.[16] The 5-year disease-free and overall survival rates in this study were 86.4% and 95.2%, respectively.[16] The M.D. Anderson Cancer Centre approach to treat ONB is complete surgical resection (craniofacial resection) followed by adjuvant radiotherapy.[11] Overall patient survival rates with this approach at 5 and 10 years were 89% and 81%, respectively.[11]

Five year survival data for stages A, B, and C of the Kadish classification system are 75%, 68% and 41%, respectively. Most frequent recurrence is local with recurrence rates of approximately 30%. The mean time for recurrence of ONB is 4.67 years.[11]

4. Conclusion

In conclusion, a standard treatment for ON is not yet clear as the number of ON cases is limited. However, due to the 20% risk of neck metastasis in stage B and C, treatment should include a wide surgical excision and prophylactic neck irradiation should be added to the RT region.

Prospective studies with a large number of patients are required in order to establish a gold-standard treatment.

5. Source of Funding

None.

6. Conflict of Interest

The author declares that there is no conflict of interest.

References

1. Berger L, Luc R, Richard D. Olfactory esthesioneuroblastoma. *Bull Assoc Fr Etude Cancer*. 1924;13:410–21.
2. Bhattacharyya N, Thornton AF, Joseph MP, Goodman ML, Amrein PC. Successful treatment of esthesioneuroblastoma and neuroendocrine carcinoma with combined chemotherapy and proton radiation. Results in 9 cases. *Arch Otolaryngol Head Neck Surg*. 1997;123(1):34–40. doi:10.1001/archotol.1997.01900010038005.
3. Rakes SM, Yeatts RP, Campbell RJ. Ophthalmic manifestations of esthesioneuroblastoma. *Ophthalmology*. 1985;92(12):1749–53. doi:10.1016/s0161-6420(85)34102-7.
4. Kutluhan A, Yilmaz N, Yakut F, Yurttaş V, Uğraş S. Treatment of olfactory neuroblastoma via subfrontal and midfacial degloving approaches: A case report. *Kulak Burun Bogaz Ihtis Derg*. 2008;18(1):56–8.
5. Berger L, Luc R. L'esthesioneuroepitheliome olfactif. *Bull Assoc Fr Etude Cancer*. 1924;13:410–21.
6. Kleihues P, Cavenee WK, editors. WHO Classification of Tumours of Pathology and Genetics Tumours of the Nervous System. Lyon, France: IARC Press; 2000. p. 150–2.
7. Pickuth D, Heywang-Kobrunner SH, Spielmann RP. Computed tomography and magnetic resonance imaging features of olfactory neuroblastoma: an analysis of 22 cases. *Clin Otolaryngol Allied Sci*. 1999;24(5):457–61. doi:10.1046/j.1365-2273.1999.00295.x.
8. Kadish S, Goodman M, Wang CC. Olfactory neuroblastoma. A clinical analysis of 17 cases. *Cancer*. 1976;37(3):1571–6. doi:10.1002/1097-0142(197603)37:3<1571::aid-cncr2820370347>3.0.co;2-l.
9. Morita A, Ebersold MJ, Olsen KD, Foote RL, Lewis JE, Quast LM, et al. Esthesioneuroblastoma: Prognosis and management. *Neurosurgery*. 1993;32(5):706–14. doi:10.1227/00006123-199305000-00002.
10. Zanation AM, Ferlito A, Rinaldo A, Gore MR, Lund VJ, Mckinney KA, et al. When, how and why to treat the neck in patients with esthesioneuroblastoma: A review. *Eur Arch Otorhinolaryngol*. 2010;267(11):1667–71. doi:10.1007/s00405-010-1360-6.
11. Diaz EM, Johnigan RH, Pero C, El-Naggar AK, Roberts DB, Barker JL, et al. Olfactory neuroblastoma: The 22-year experience at one comprehensive cancer center. *Head Neck*. 2005;27(2):138–49. doi:10.1002/hed.20127.
12. Morita A, Ebersold MJ, Olsen KD, Foote RL, Lewis JE, Quast LM, et al. Esthesioneuroblastoma: prognosis and management. *Neurosurg*. 1993;32(5):706–14. doi:10.1227/00006123-199305000-00002.
13. Howard DJ, Lund VJ, Wei WI. Craniofacial resection for tumors of the nasal cavity and paranasal sinuses: A 25-year experience. *Head Neck*. 2006;28(10):867–73. doi:10.1002/hed.20432.
14. Benfari G, Fusconi M, Ciofalo A, Gallo A, Altissimi G, T C, et al. Radiotherapy alone for local tumour control in esthesioneuroblastoma. *Acta Otorhinolaryngol Ital*. 2008;28(6):292–7.
15. Montava M, Verillaud B, Kania R, Sauvaget E, Bresson D, Mancini J, et al. Critical analysis of recurrences of esthesioneuroblastomas: can we prevent them? *Eur Arch Otorhinolaryngol*. 2014;271(12):3215–22. doi:10.1007/s00405-014-3035-1.
16. Herr MW, Sethi RK, Meier JC, Chambers KJ, Remenschneider A, Chan A, et al. An update on the massachusetts eye and ear infirmary and Massachusetts general hospital experience with craniofacial resection, proton beam radiation, and chemotherapy. *J Neurol Surg B Skull Base*. 2014;75(1):58–64. doi:10.1055/s-0033-1356493.

Author biography

Sunil Kathuria, Head of Department and Cochlear Implant Surgeon

Cite this article: Kathuria S, Sunny C. Olfactory neuroblastoma: A case report. *IP Indian J Anat Surg Head, Neck Brain* 2022;8(1):19-22.