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Case Report

# Olfactory neuroblastoma: A rare case report

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ABSTRACT

Olfactory neuroblastoma (ONB) is an uncommon malignant tumor of the sinonasal tract that arises from the olfactory neuroepithelium and exhibits neuroblastic differentiation. Here, we present the case of a 48-year-old woman who complained of epistaxis for 4 months, nasal obstruction, and headaches for 6 months. Upon examination, a mass was identified in the left nasal cavity and confirmed by radiological imaging. Surgical excision was performed, and the tissue specimen was sent for histopathological analysis, which confirmed the diagnosis of olfactory neuroblastoma. Immunohistochemical markers revealed focal positivity for chromogranin, synaptophysin, AE1/AE3, CK7, with a high Ki-67 index (60-70%), and negativity for S-100.



## 1. Introduction

Olfactory neuroblastoma (ONB) is a malignant neuroectodermal tumor that exhibits neuroblastic differentiation. It typically originates from the olfactory mucosa located in the superior nasal cavity.[1] It comprises approximately 2-3% of all sinonasal tumors and demonstrates a bimodal peak in incidence during the 2nd and 6th decades of life, although it can occur across a wide age range (2–90 years) with no apparent gender preference. ONB can histologically resemble tumors found in the sino-nasal tract and small round blue cell tumors.[2] Immunohistochemistry (IHC) typically shows diffuse positivity for synaptophysin, chromogranin, CD 56, and variable results for S-100.

## 2. Case Report

48-year-old female presented with complaints of epistaxis since 4 months, nasal blockage and headache since 6 months. On examination, a mass involving left side of nasal cavity was present and confirmed radiologically. Excision was done and tissue specimen sent for histopathological examination.

Grossly, multiple grey white to red brown bony and soft tissue portions received, largest measuring 5 x 1.5 x 1.5 cm. Cut surface appeared grey-tan. Representative sections were taken. Tissue processing was done with routine paraffin embedding technique. Slides were stained with H&E.

### 2.1. Microscopic examination

Sections reveal tumor to be composed of nested and diffuse growth pattern with prominent vascularity. The round to polygonal cells show markedly pleomorphic nuclei with frequent mitoses. Necrosis is present. At places Homer Wright pseudorosettes, Flexner Wintersteiner rosettes and ganglion like cells are seen.

### 2.2. Immunohistochemistry report

Chromogranin (LK2H10) - Focal positive

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Synaptophysin (SP11) - Focal Positive
AE1 (AE1/AE3) - Focal Positive
CK7 (OV-TL12/30) Focal Positive
TTF1 (8G7G3/1) - Negative
Ki-67 (MIB1) 60-70%
S-100 (4C4.9) Negative
P40 (ZR8) Negative
CK5/6 (D5/16 B4) - Negative
INI (MRQ-27) - Retained

Overall findings were reported as olfactory neuroblastoma.

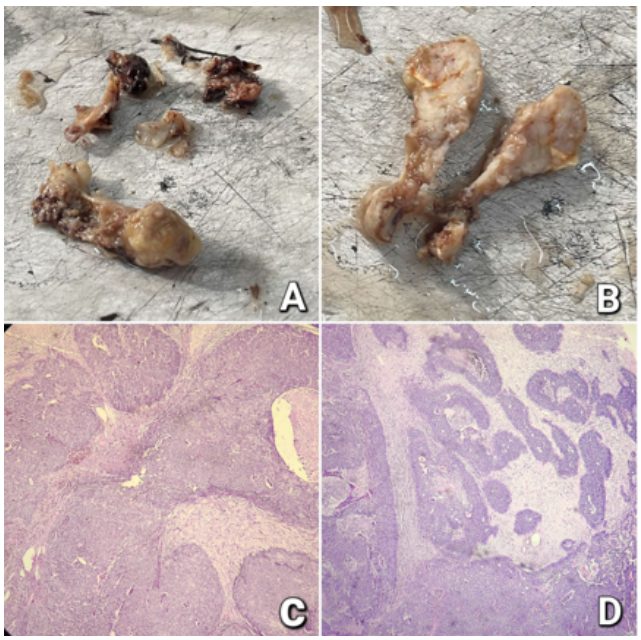


**Figure 1: A:** Gross picture of nasal tissue; **B:** Cut section of nasal tissue; **C,D:** Nested and diffuse growth pattern with prominent vascularity

## 3. Discussion

Olfactory neuroblastoma, a rare malignant tumor of the sinonasal tract, originates from neuroepithelial elements in the olfactory mucosa. These cells are typically found in the upper nasal cavity, including the septum, superior nasal concha, roof of the nose, and ethmoid sinus cribriform plate.[3] The tumor presents a broad age range (2–90 years), with a median age around 50 years. Grossly, it appears as a vascular polypoid mass, typically soft, and located in the nasal fossa roof. Although primarily originating there, it can rarely arise in the nasopharynx, maxillary sinus, or ethmoid sinus. Known for its local aggressiveness, it metastasizes hematogenously and via lymphatic routes.[4] The exact cause remains unknown.

Histologically, low-grade olfactory neuroblastoma forms submucosal nests or lobules with vascular or hyalinized fibrous stroma. Pseudorosettes (Homer-Wright rosettes)

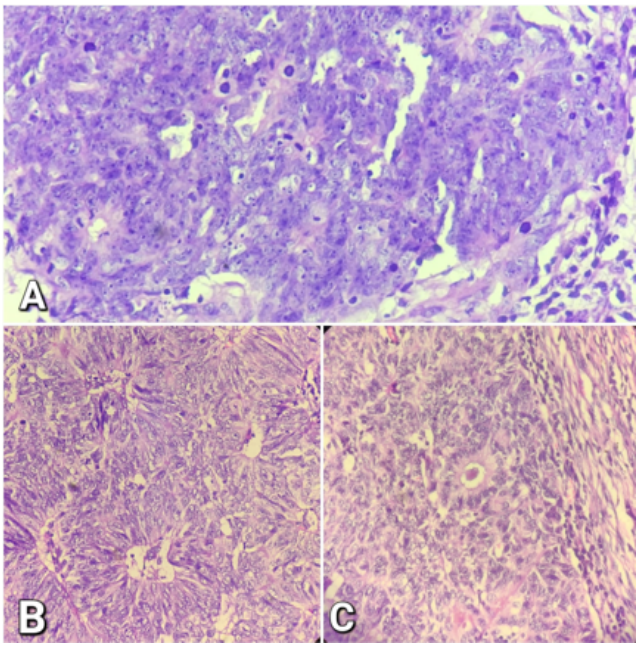


**Figure 2: A:** Round to polygonal cells show markedly pleomorphic nuclei; **B:** Homer Wright pseudorosettes; **C:** flexner winter steiner rosettes.

may appear, characterized by neoplastic cells surrounding a fibrillar neural matrix.[5] Tumor cells typically exhibit uniformity, with sparse cytoplasm, round or ovoid nuclei, punctate chromatin, and small or absent nucleoli. Higher-grade tumors may show necrosis, pleomorphism, increased mitoses, and less distinct lobular growth patterns. They can also form gland-like rings or annular formations (Flexner-Wintersteiner rosettes), though these are not solely diagnostic. Calcifications, melanin pigment, ganglion cells, and divergent differentiation may occasionally be present. Tumors that resemble olfactory neuroblastoma histologically include malignant lymphoma, plasmacytoma, embryonal/alveolar rhabdomyosarcoma, and the Ewing sarcoma/PNET tumor family.[6–8] Immunohistochemistry reveals diffuse staining for neuron-specific enolase, synaptophysin, chromogranin A, CD56 (NCAM), and beta-tubulin, along with variable S100 protein reactivity. Negative markers include CD45RB, CD99, p63, and FLI1. Ki-67 proliferation index ranges widely (2-50%), with BCL2 expression increasing with tumor grade.

Electron microscopy reveals consistent findings of neurofilaments, neurotubules, and dense-core neurosecretory cytoplasmic granules in olfactory neuroblastomas, which can aid in diagnosing challenging cases.

Olfactory neuroblastoma exhibits intricate karyotypic alterations. According to an array-based comparative genomic hybridization study, gene copy gains are more frequent than losses. The most notable changes involve

gains at 7q, 9p, 20p/q, and Xp/q, and losses at 2q, 6q, 22q, and Xp/q.[1] In one study, researchers identified a specific deletion on chromosome 11 and a gain on chromosome 1p that correlated with metastasis and poorer prognosis in olfactory neuroblastoma. Additionally, gains on 20q and 13q were noted, which may play a significant role in the progression of this tumor type, possibly involving genes with functional relevance. Expression of the OMP and RICBB genes has also been documented in olfactory neuroblastomas.[9]

Olfactory neuroblastoma is known for its tendency to locally invade neighboring structures such as the paranasal sinuses, nasopharynx, palate, orbit, base of skull, and brain. Approximately one-fifth of cases experience distant metastases, with the cervical lymph nodes and lungs being the most frequent sites.[10] The 5-year survival rate ranges from 50% to 66%. Late recurrence is a common in this condition.

## 4. Conclusion

Biopsy results reveal undifferentiated malignant tumor with morphological features consistent with high grade olfactory neuroblastoma. IHC report shows tumor was focally positive for chromogranin, synaptophysin, AE1/AE3, CK7 with high Ki-67 (60-70%) index and negative for S-100. Although clinicopathologic features may lead to diagnosis of ONB, it is necessary to differentiate ONB with other differentials due to its rare presentation.

## 5. Source of Funding

None.

## 6. Conflict of Interest

None.

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