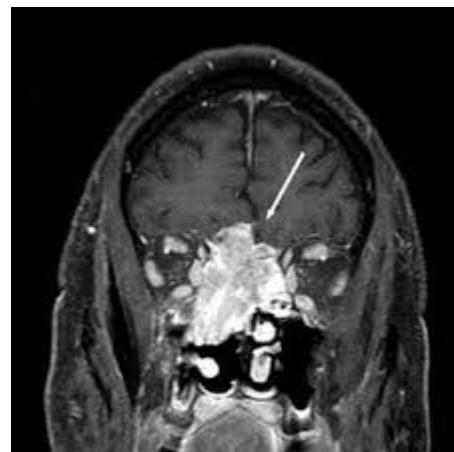


Esthesioneuroblastoma (ENB)



Esthesioneuroblastoma (ENB), originally described in 1924, AKA [olfactory neuroblastoma](#), AKA olfactory esthesioneuroblastoma, AKA [esthesioneurocytoma](#), AKA olfactory placode tumor ¹⁾.

Malignant tumor of [sinonasal](#) origin.

Named for the [stem cell](#) of the olfactory epithelium ([esthesioneuroblast](#)). A malignant tumor arising from [crest cells](#) of the nasal vault, often with intracranial invasion.

Epidemiology

A rare nasal [neoplasm](#) with an [incidence](#) of 0.4 per 1,000,000 people. ²⁾

Believed to arise from the olfactory [neural crest cells](#) in the upper nares, it is considered to be malignant. These [tumors](#) occur over a wide [age](#) range (3 to 90 years), with a bimodal peak between the second and third decade and a second peak in the sixth and seventh decades.

Etiology

[Somatostatin receptor 2](#) (SSTR2) expression has previously been documented in olfactory neuroblastoma (ONB). Here, we fully characterize SSTR2 expression in ONB and correlate staining results with clinicopathologic parameters including Hyams grade. We also assess SSTR2 immunohistochemistry expression in various histologic mimics of ONB to assess its diagnostic functionality. 78 ONBs (51 primary biopsies/excisions and 27 recurrences/metastases) from 58 patients were stained for SSTR2. H-scores based on intensity (0-3 +) and percentage of tumor cells staining were assigned to all cases. 51 histologic mimics were stained and scored in an identical fashion. 77/78 (99%) ONB cases demonstrated SSTR2 staining (mean H-score: 189, range: 0-290). There were no significant differences in staining between primary tumors and recurrences/metastases (mean H-score: 185 vs 198). Primary low-grade ONB had somewhat stronger staining than high-grade tumors (mean H-score: 200 vs 174). SSTR2 expression had no prognostic value when considering disease-free or disease-specific survival. SSTR2 staining is significantly higher in ONB than its histologic mimics (mean H-score: 189 vs 12.9, $p < 0.001$) suggesting a potential use of the marker in

diagnosis of ONB. In conclusion, SSTR2 is consistently expressed in ONB suggesting a role for somatostatin-analog based imaging and therapy in this disease. More generally, SSTR2 may be another marker of neuroendocrine differentiation in ONB ³⁾

Classification

[Esthesioneuroblastoma classification.](#)

Clinical features

These tumors typically present with unilateral nasal obstruction and [epistaxis](#).

Diagnosis

Diagnosis is confirmed on [biopsy](#).

MRI: [isointense](#) with the brain on T1-weighted imaging and intermediate to high signal intensity on T2- weighted imaging and enhance heterogeneously with gadolinium. Signal characteristics may mimic meningioma. For higher stage lesions, the [cribriform plate](#) may be eroded; better seen on thin-cut CT. The most important factor determining resectability is an intracranial extension. Magnetic resonance aids in the distinction between extradural tumors, dural invasion, or parenchymal brain invasion. None of these are specific to this tumor.

Endoscopic biopsy is typically performed in the otolaryngology office prior to surgery. A clinical oncology exam should be performed, and if there is suspicion for metastatic disease a PET scan, which is sensitive to metastatic disease, should be ordered.

Differential diagnosis

Includes SNUC, nasal melanoma, nasal squamous cell carcinoma, and meningioma.

Treatment

see [Esthesioneuroblastoma treatment](#).

Outcome

[Esthesioneuroblastoma outcome.](#)

Case series

[Esthesioneuroblastoma case series.](#)

Case reports

[Esthesioneuroblastoma case reports.](#)

References

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²⁾

Theilgaard SA, Buchwald C, Ingeholm P, et al. Esthesioneuroblastoma: a Danish demographic study of 40 patients registered between 1978 and 2000. Acta Otolaryngol. 2003; 123:433-439

³⁾

Cracolici V, Wang EW, Gardner PA, Snyderman C, Gargano SM, Chiosea S, Singhi AD, Seethala RR. SSTR2 Expression in Olfactory Neuroblastoma: Clinical and Therapeutic Implications. Head Neck Pathol. 2021 Apr 30. doi: 10.1007/s12105-021-01329-1. Epub ahead of print. PMID: 33929681.

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