



# Esthesioneuroblastoma (Olfactory Neuroblastoma): Overview and Extent of Surgical Approach and Skull Base Resection

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## Abstract

Esthesioneuroblastoma is a rare malignancy originating from the olfactory epithelium. Treatment consists of surgical resection with strong consideration for adjuvant treatment in advanced Kadish stage and high Hyams grade. In the modern era, overall outcomes for esthesioneuroblastoma are favorable compared with many other sinonasal malignancies with 5-year overall survival estimated to be 80%. When selecting the optimal surgical approach, the surgeon must consider the approach that will allow for a negative margin resection and adequate reconstruction. In appropriately selected patients, endoscopic outcomes appear at least equivalent to open approaches and unilateral endoscopic approach may be used in select olfactory preservation cases.

## Keywords

- ▶ esthesioneuroblastoma
- ▶ endoscopic
- ▶ craniofacial resection
- ▶ olfactory
- ▶ neuroblastoma

## Introduction

Esthesioneuroblastoma is a rare malignancy originating from the olfactory epithelium. Treatment consists of surgical resection with strong consideration for adjuvant treatment in advanced Kadish stage and high Hyams grade.<sup>1,2</sup> In the modern era, overall outcomes for esthesioneuroblastoma are favorable compared with many other sinonasal malignancies with 5-year overall survival estimated to be 80%.<sup>2,3</sup>

## Case Presentation

A 48-year-old male presented with left-sided nasal obstruction. Magnetic resonance imaging demonstrated invasion of the anterior left cribriform plate into the inferior aspect of the anterior cranial vault, invasion of bilateral olfactory

bulbs, and slight extension laterally over the left orbit (▶ **Fig. 1A** and **B**). Workup demonstrated no regional or distant metastatic disease. This esthesioneuroblastoma was a modified Kadish stage C and Hyams grade 3. The patient underwent surgical resection with endoscopic bilateral craniofacial resection (CFR) (▶ **Fig. 2**), and margin negative resection was achieved. Reconstruction with pericranium, fascia lata, and extended nasoseptal flap was performed. He completed adjuvant radiation to the anterior skull base and disease free 3 years posttreatment.

## Overview

Treatment of esthesioneuroblastoma is dependent on the extent of the tumor at presentation. Surgical resection is the mainstay of treatment with consideration for adjuvant radiation with or without chemotherapy.<sup>1</sup> In rare cases,

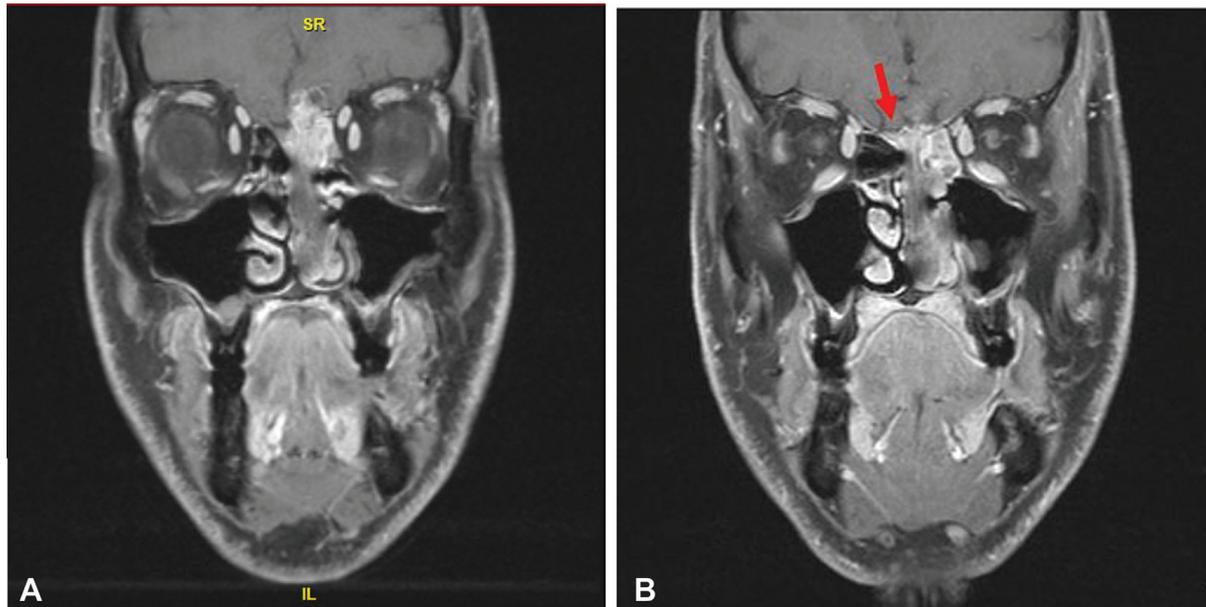
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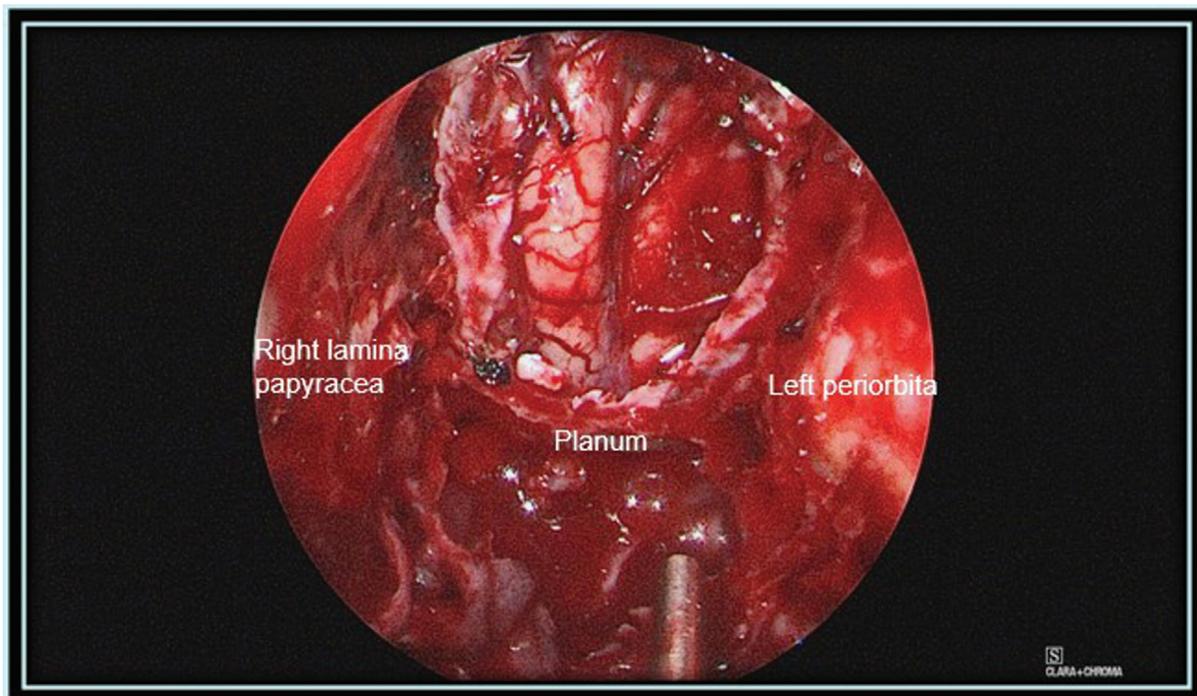
**Fig. 1** (A) Case presentation of coronal magnetic resonance imaging, T1 postgadolinium. (B) Arrow indicates contralateral olfactory tract involvement.

induction chemotherapy may be indicated prior to definitive treatment for locally unresectable tumors.<sup>4</sup>

In the modern era, open CFR refers to bifrontal craniotomy combined with assistance from an endoscopic endonasal approach for intranasal tumor resection, margin sampling, and assistance with reconstruction. An open approach is often selected where negative margins are not achievable with endoscopic approaches such as extensive intracranial involvement, orbital invasion, lateral ex-

tent of tumor past mid-plane of the orbit, extension to the optic canal and cavernous sinus, or when there is tumor high in the frontal sinus.<sup>1,5</sup> An open approach may also be selected when tumor involvement precludes intranasal reconstructive as it allows for direct dural repair with fascia lata and placement of pericranial flap for cranial base reconstruction.<sup>1</sup>

In high-volume skull base centers, many tumors can be addressed with a completely transnasal endoscopic CFR with



**Fig. 2** Endoscopic bilateral craniofacial resection.

comparable outcomes to open approaches.<sup>1,5,6</sup> Kadish stage A and B tumors can typically be managed completely endoscopically. Increasingly larger Kadish stage C tumors may be resected solely with an endoscopic approach with clear surgical margins and endoscopic reconstruction. Historical teaching includes resection of bilateral olfactory bulbs and tracts. However, modification of the traditional surgical extent for preserved olfaction may be considered using a unilateral endoscopic skull base resection.<sup>1,7–10</sup> Unilateral endoscopic resection may be considered when high-quality preoperative imaging suggests no involvement of contralateral olfactory bulb or tract along with the assistance of intraoperative frozen section margin analysis to ensure complete resection.<sup>7,8</sup>

## Conclusion

Overall, esthesioneuroblastoma has favorable survival outcomes when compared with other sinonasal malignancies.<sup>2,3</sup> When selecting the optimal surgical approach, the surgeon must consider the approach that will allow for a negative margin resection and adequate reconstruction. In appropriately selected patients, endoscopic outcomes appear at least equivalent to open approaches and unilateral endoscopic approach may be used in select olfactory preservation cases.

### Conflict of Interest

None declared.

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