Endoscopic-Assisted Craniofacial Resection of Esthesioneuroblastoma: Minimizing Facial Incisions

Technical Note and Report of 3 Cases

James K. Liu, M.D., Brent O'Neill, B.S., Richard R. Orlandi, M.D., Randy L. Jensen, M.D., Ph.D., and William T. Couldwell, M.D., Ph.D.

Department of Neurosurgery (JKL, BO, RLJ) and Division of Otolaryngology – Head and Neck Surgery (RRO), University of Utah School of Medicine, Salt Lake City, Utah

Please address correspondence to:
William T. Couldwell, M.D., Ph.D.
Department of Neurosurgery
University of Utah Health Sciences Center
50 North Medical Drive
Salt Lake City, Utah 84132
Tel: (801) 581-6908
Fax: (801) 581-4385
email: william.couldwell@hsc.utah.edu

Running Title: **Endoscopic Approach for Esthesioneuroblastoma**

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Abstract

The surgical management of esthesioneuroblastomas has traditionally been craniofacial resection, which combines a bifrontal craniotomy with a transfacial approach. The latter usually involves a disfiguring facial incision, mid-facial degloving, lateral rhinotomy, and/or extensive facial osteotomies, which may be cosmetically displeasing to the patient. The advent of angled endoscopes has provided excellent magnification and illumination for surgeons to remove tumors using minimally invasive techniques.

The authors describe their experience with three cases of esthesioneuroblastoma, which were surgically removed using a transnasal endoscopic approach, without the utilization of transfacial incisions. Preoperative radiographs were reviewed and tumors were staged according to the Kadish staging system. One patient had a recurrent esthesioneuroblastoma (Kadish stage B) which was removed entirely through a transnasal endoscopic approach. Two patients had intracranial extension (Kadish stage C), which were resected with a combined approach, endoscopically from below and a bifrontal craniotomy from above to remove intracranial disease. All patients underwent reconstruction of the anterior skull base.

Esthesioneuroblastomas confined to the nasal and paranasal cavities (Kadish stage A and B), were readily accessible through the transnasal endoscopic approach. If there was significant intracranial disease (Kadish stage C), adding a bifrontal craniotomy provided excellent exposure for complete resection of involved tumor. All patients underwent complete tumor resection with negative margins. None had a cerebrospinal fluid (CSF) leak.

The endoscopic-assisted craniofacial approach for the surgical management of esthesioneuroblastomas provides excellent exposure and adequate visualization as well as the cosmetic benefit of avoiding an external facial incision.

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Introduction

Esthesioneuroblastomas (also known was olfactory neuroblastomas) are rare, malignant neuroectodermal tumors of the upper nasal cavity that often involves the anterior skull base. Multimodality treatments, including surgery, radiation therapy, chemotherapy, or a combination, are the most frequently recommended treatments. The surgical procedure of choice has traditionally been a craniofacial resection, which combines a bifrontal craniotomy with a transfacial approach that usually includes a disfiguring facial incision, mid-facial degloving, lateral rhinotomy, and/or extensive facial osteotomies, which may not produce a good cosmetic result. The authors describe three cases of esthesioneuroblastoma which were surgically treated with an endoscopic-assisted craniofacial approach that did not require any external facial incisions. One patient was treated solely through a transnasal endoscopic approach. The other two patients underwent a combined approach, endoscopically from below and a bifrontal craniotomy from above.

Case Reports

Case 1

This 74-year-old male who had a previous craniofacial resection for an esthesioneuroblastoma presented with nasal obstruction secondary to a nasopharyngeal mass. An MRI demonstrated a recurrent esthesioneuroblastoma (Kadish stage A) involving the nasopharynx extending up to the anterior skull base (Fig.1). Using an endoscopic transnasal approach, the nasopharyngeal mass was removed completely from

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below, without using a facial incision. The anterior skull base defect was repaired with a fascia lata graft. There was no CSF leak postoperatively and the remainder of the hospital course was unremarkable. The patient has been free of recurrence for 30 months.

Case 2

This 58-year-old female presented with recurrent epistaxis accompanied by frequent sinus infections and anosmia secondary to an intranasal mass extending into the left maxillary sinus and left ethmoid sinus detected on CT. A biopsy of the mass revealed esthesioneuroblastoma. She received seven cycles of cisplatin and etoposide combination chemotherapy with 60 Gy adjuvant radiation therapy following the third cycle. Although the tumor shrank after receiving these treatments, there was persistent tumor radiographically with intracranial extension (Kadish stage C). There was no evidence of metastasis.

The patient underwent a combined endoscopic-assisted craniofacial approach. A bifrontal craniotomy was performed initially to remove tumor that had extended through the anterior skull base (Fig. 2). The tumor was noted to be entirely extradural. A small portion of tumor that was adherent to the dura was removed leaving a dural defect. Upon inspection, there was no evidence of intradural invasion. The remainder of the tumor was removed from below endoscopically through the nasal cavity. Tumor was found involving the sphenoid sinus, ethmoid sinus, nasal septum, cribriform plate, and lamina papyracea. By working simultaneously intracranially from above and endoscopically from below, the tumor was removed en bloc. The durotomy was repaired with a

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temporalis fascia graft and reinforced with a vascularized pericranial flap, which was placed over the floor of the anterior cranial fossa. The flap was placed over the planum sphenoidale with transnasal assistance from the endoscopic surgeon, leading to improved ease and accuracy of placement. Pathological examination revealed esthesioneuroblastoma with negative margins, suggesting a complete resection. Postoperatively, the patient was neurologically intact. There was no evidence of a CSF leak and the remainder of her hospital course was uneventful. She remains free of recurrence after 8 months.

Case 3

This 36-year-old male presented with a six month history of nasal fullness, epistaxis and headache. A CT scan revealed a mass in the anterior nasal cavity. An endoscopic nasal biopsy was preformed, establishing the diagnosis of esthesioneuroblastoma. An MRI (Fig. 3) of the head and neck demonstrated extension of tumor through the cribriform plate, the medial orbital walls bilaterally, and the wall of the right maxillary sinus (Kadish stage C). Neither dura nor brain enhanced on the scan, and lymph nodes in the neck showed no evidence of metastatic disease. The patient underwent preoperative chemotherapy and achieved a dramatic response, decreasing tumor size by approximately 80%.

The patient underwent an endoscopic-assisted craniofacial resection. A bifrontal craniotomy was first performed. The tumor was purely extradural. The dura was easily mobilized from the anterior skull base without evidence of tumor adhesion. Tumor

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resection proceeded from below through a transnasal endoscopic approach as the frontal lobes were protected through the craniotomy exposure. The tumor was attached primarily to the cribriform plate. En bloc removal was obtained. Biopsies from the medial orbital walls, ethmoid, and maxillary sinuses were negative for malignancy. The anterior skull base defect was repaired with a vascularized pericranial flap. The patient awoke with no neurological deficits and the postoperative course was uncomplicated. There was no CSF leak postoperatively. He remains disease free at 17 months postoperatively.

Discussion

In 1924, Berger and Luc[1] described a nasal mass arising in the superior nasal cavity from the olfactory epithelium that exhibited cellular rosettes and fibrils. They termed it *l'estheosio neuroepitheliome olfactif*. Since then, this aggressive malignant tumor has presented a surgical challenge to skull base surgeons. Esthesioneuroblastomas can grow in all directions involving the nasal cavity, paranasal sinuses, cribriform plate, intracranial cavity, or orbit. They can be locally aggressive by invading brain parenchyma and seeding CSF. They can also metastasize via lymphatic and hematogenous routes.[2]

Treatment requires a multidisciplinary approach including surgery, radiation therapy, and chemotherapy.[3-6] The surgical procedure of choice has been a craniofacial resection for *en bloc* removal of tumor.[7-14] In the experience of Levine et al., craniofacial resection has improved disease-free survival from 37.5 to 82% when compared to extracranial excision.[4,12,15] The transfacial portion of the procedure usually involves a

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Weber-Fergusson incision that may leave a cosmetically disfiguring scar. Proponents of traditional craniofacial approaches are quick to justify that facial incisions allow for maximal exposure and visualization facilitating complete resection.[16]

In the last decade, there has been an increased application for endoscopic approaches in minimally invasive skull base surgery.[16-28] Surgical resection of nasal, paranasal, and skull base neoplasms are being performed with endoscopic approaches.

There are few reports in the literature describing endoscopic approaches for esthesioneuroblastoma.[16,18] Casiano et al.,[16] reported 5 patients with Kadish stage A and B esthesioneuroblastomas which were resected and immediately reconstructed through a transnasal endoscopic approach. One patient had a CSF leak and one had an orbital hematoma. Two patients developed regional metastasis to the orbit and cervical lymph nodes requiring transorbital excision and a modified neck dissection. Walch et al.,[18] described 3 patients (2 with Kadish stage B, 1 with Kadish stage C) who underwent transnasal endoscopic resection followed by stereotactic radiosurgery. None of these patients had intracranial extension. The patient with the Kadish stage C tumor had destruction of the lamina papyracea. All three remained disease free in a 39 to 71 month follow-up period. In the present report, we describe three patients with esthesioneuroblastoma who underwent an endoscopic-assisted craniofacial resection without the utilization of transfacial incisions.

Our initial preoperative radiological assessment includes a CT and/or MRI, which allows for staging of the esthesioneuroblastomas according to the Kadish staging system (Table 1).[29] Esthesioneuroblastomas are classified into three stages depending on location of the tumor: stage A if the tumor is limited by the nasal cavity; stage B if the tumor

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extends into the paranasal sinuses; stage C if the tumor extends beyond the paranasal sinuses (intraorbital and/or intracranial extension). We use the Kadish staging system to preoperatively plan our surgical approach. Kadish stage A and B tumors are readily accessible through the transnasal endoscopic approach. Kadish stage C tumors involving the lamina papyracea without intracranial extension can still be accessed endoscopically. In treating Kadish stage C tumors with intracranial extension, adding a bifrontal craniotomy allows adequate exposure and visualization of intracranial pathology. Sometimes, it may be difficult to determine radiographically if tumor has invaded the dura until the time of surgery. If tumor is adherent or has invaded the dura and/or parenchyma, the involved elements must be resected and repaired with autologous fascia lata, temporalis fascia, and/or vascularized pericranium.[30] In these situations, the bifrontal approach is very useful for intracranial tumor ablation as well as for skull base reconstruction.

Successful tumor ablation must be followed by proper reconstruction of the skull base defect.[31-34] It is paramount to create an anatomic and functional seal between the intracranial and extracranial contents, preferably using vascularized tissue for dural coverage. Failure to do so can result in potentially life-threatening infectious complications.[35-40] A watertight closure of dura and a vascularized cover is necessary to enhance primary healing of the intracranial wound, especially when the dural closure is tenuous and/or has been repaired with a graft. The use of well-vascularized tissue for dead-space obliteration and skull base coverage is especially important in the setting of planned administration of perioperative radiation therapy. If possible, attempts should be made to restore function and optimize the cosmetic result. All three patients in this study

underwent successful repair with autologous fascia lata supplemented by vascularized pericranial flap. None had sequelae of CSF leaks.

Our approach to esthesioneuroblastomas combines the cosmetic benefit of avoiding transfacial incisions and the curative benefit of gross total tumor resection. The advent of angled endoscopes has provided excellent illumination and magnification as well as the ability to extend visualization into anatomic recesses.[16] For patients with intracranial extension, adding a bifrontal craniotomy to the endoscopic procedure provides excellent protection of intracranial structures, safe resection of involved tumor, and adequate exposure for skull base reconstruction.

The standard treatment is craniofacial resection followed by postoperative adjuvant radiation therapy and/or chemotherapy. The overall 5- and 10-year survival rate for traditional craniofacial resection followed by adjuvant radiation therapy and/or chemotherapy for esthesioneuroblastoma has been approximately 80 and 50%, respectively.[10,15,41] The long-term efficacy of esthesioneuroblastoma treated endoscopically awaits longer follow-up with a larger number of patients.

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References

1. Berger L, Luc H, Richard R. L'esthesioneuro epitheliome olfactif. Bull Assoc Fr Etude Cancer 1924;13:410-420.

2. Shaari CM, Catalano PJ, Sen C, et al. Central nervous system metastases from esthesioneuroblastoma. Otolaryngol Head Neck Surg 1996;114:808-812.

3. Polin RS, Sheehan JP, Chenelle AG, et al. The role of preoperative adjuvant treatment in the management of esthesioneuroblastoma: the University of Virginia experience. Neurosurgery 1998;42:1029-1037.

4. Levine PA, McLean WC, Cantrell RW. Esthesioneuroblastoma: the University of Virginia experience 1960-1985. Laryngoscope 1986;96:742-746.

5. McElroy EA Jr, Buckner JC, Lewis JE. Chemotherapy for advanced esthesioneuroblastoma: the Mayo Clinic experience. Neurosurgery 1998;42:1023-1028.

6. Wade PM Jr, Smith RE, Johns ME. Response of esthesioneuroblastoma to chemotherapy: report of five cases and review of the literature. Cancer 1984;53:1036-1041.

7. Lund VJ, Howard DJ, Wei WI, et al. Craniofacial resection for tumors of the nasal cavity and paranasal sinuses- a 17-year experience. Head Neck 1998;20:105.

8. Bilsky MH, Kraus DH, Strong EW, et al. Extended anterior craniofacial resection for intracranial extension of malignant tumors. Am J Surg 1997;174:565-568.

9. Shah JP, Kraus DH, Bilsky MH, et al. Craniofacial resection for malignant tumors involving the anterior skull base. Arch Otolaryngol Head Neck Surg 1997;123:1312-1317.

10. Levine PA, Debo RF, Meredith SD, et al. Craniofacial resection at the University of Virginia (1976-1992): survival analysis. Head Neck 1994;16:574-577.

11. Shah JP, Galicich JH. Craniofacial resection for malignant tumors of ethmoid and anterior skull base. Arch Otolaryngol 1977;103:514-517.

12. Levine PA, Scher RL, Jane JA, et al. The craniofacial resection- eleven-year experience at the University of Virginia: problems and solutions. Otolaryngol Head Neck Surg 1989;101:665-669.

13. Bridger GP, Baldwin M. Anterior craniofacial resection for ethmoid and nasal cancer with free flap reconstruction. Arch Otolaryngol Head Neck Surg 1989;115:308-312.

14. Panje WR, Dohrmann GJ, Pitcock JK, Scher N, Weichselbaum RR, Sutton HG Jr, Vokes E, Moss J. The transfacial approach for combined anterior craniofacial tumor ablation. Arch Otolaryngol Head Neck Surg 1989;115:301-307.

15. Levine PA, Gallagher R, Cantrell RW. Esthesioneuroblastoma: reflections of a 21-year experience. Laryngoscope 1999;109:1539-1543.

16. Casiano RR, Numa WA, Falquez AM. Endoscopic resection of esthesioneuroblastoma. Am J Rhinol 2001;15:271-279.

17. Sukenik MA, Casiano RR. Endoscopic medial maxillectomy for inverted papillomas of the paranasal sinuses: value of the intraoperative endoscopic examination. Laryngoscope 2000;110:39-42.

18. Walch C, Stammberger H, Anderhuber W, Unger F, Kole W, Feichtinger K. The minimally invasive approach to olfactory neuroblastoma: combined endoscopic and stereotactic treatment. Laryngoscope 2000;110:635-640.

19. Thaler ER, Kotapka M, Lanza DC, et al. Endoscopically assisted anterior cranial skull base resection of sinonasal tumors. Am J Rhinol 1999;13:303-310.

20. Cappabianca P, Alfieri A, Thermes S, Buonamassa S, de Divitiis E. Instruments for endoscopic endonasal transsphenoidal surgery. Neurosurgery 1999;45:392-396.

21. Casiano RR, Livingston J. Endoscopic Lothrop procedure: the University of Miami experience. Am J Rhinol 1998;12:335-339.

22. Voegels RL, Luxemberger W, Stammberger H. Transnasal endoscopic removal of an extensive immature teratoma in a three-month-old child. Ann Otol Rhinol Laryngol 1998;107:654-657.

23. Tseng HZ, Chao WY. Transnasal endoscopic approach for juvenile nasopharyngeal angiofibroma. Am J Otolaryngol 1997;18:151-154.

24. Yuen APW, Hung KN. Endoscopic cranionasal resection of anterior skull base tumor. Am J Otolaryngol 1997;18:433.

25. Bhattacharyya N, Shapiro NL. Endoscopic resection of a recurrent sinonasal hemangiopericytoma. Am J Otolaryngol 1997;18:341-344.

26. Jho HD, Carrau RL. Endoscopic endonasal transsphenoidal surgery: experience with 50 patients. J Neurosurg 1997;87:44-51.

27. Lanza DC, O'Brien DA, Kennedy DW. Endoscopic repair of cerebrospinal fluid fistulae and encephaloceles. Laryngoscope 1996;106:1119-1125.

28. Dodson EE, Gross CW, Swerdloff JL, et al. Transnasal endoscopic repair of cerebrospinal fluid rhinorrhea and skull base defects: a review of twenty-nine cases. Otolaryngol Head Neck Surg 1994;111:600-605.

29. Kadish S, Goodman M, Wang CC. Olfactory neuroblastoma: a clinical analysis of 17 cases. Cancer 1976;37:1571-1576.

30. Oskouian RJ Jr, Jane JA Sr, Dumont AS, Sheehan JM, Levine PA. Esthesioneuroblastoma: clinical presentation, radiological, and pathological features, treatment, review of the literature, and the University of Virginia experience. Neurosurg Focus 2002;12 (5):Article 4.

31. Liu JK, Niazi Z, Couldwell WT. Reconstruction of the skull base after tumor resection: an overview of methods. Neurosurg Focus 2002;12 (5):Article 9.

32. Couldwell WT, Stillerman CB, Dougherty W. Reconstruction of the skull base and cranium adjacent to sinuses with porous polyethylene. Skull Base Surg 1997;7:57-63.

33. Couldwell WT. Surgery of the anterior skull base. Otolaryngol Clin North Am 1993;26:673-693.

34. Johns ME, Winn HR, McLean WC, et al. Pericranial flap for the closure defects of craniofacial resection. Laryngoscope 1981;91:952-959.

35. Jones NF, Schramm VL, Sekhar LN. Reconstruction of the cranial base following tumour resection. Br J Plast Surg 1987;40:155-162.

36. Thomson JG, Restifo RJ. Microsurgery for cranial base tumors. Clin Plast Surg 1995;22:563-572.

37. Spinelli HM, Persing JA, Walser B. Reconstruction of the cranial base. Clin Plast Surg 1995;22:555-561.

38. Langstein HN, Chang DW, Robb GL. Coverage of skull base defects. Clin Plast Surg 2001;28:375-387.

39. Chang DW, Langstein HN, Gupta A, DeMonte F, Do K, Wang X, Robb G. Reconstructive management of cranial base defects after tumor ablation. Plast Reconstr Surg 2001;107:1346-1355.

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40. Stiernberg CM, Bailey BJ, Weiner RL, et al. Reconstruction of the anterior skull base following craniofacial resection. Arch Otolaryngol Head Neck Surg 1987;113:710-712.

41. Foote RL, Morita A, Ebersold MJ, et al. Esthesioneuroblastoma: the role of adjuvant radiation therapy. Int J Radat Oncol Biol Phys 1993;27:835-842.

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