



Esthesioneuroblastoma: Management of the N0 neck

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Abstract

Keywords

- ▶ esthesioneuroblastoma
- ▶ salvage treatment
- ▶ sinonasal malignancy
- ▶ olfactory neuroblastoma

Esthesioneuroblastoma is a rare sinonasal malignancy that arises from the olfactory epithelium. The overall incidence of lymph node metastases is 25%. However, neck disease can present in a delayed fashion. As such, management of the clinically negative neck is controversial, with some advocating for elective neck treatment and others recommending observation with salvage treatment if necessary. At this time, no prospective head-to-head comparisons of elective versus salvage treatment have been performed.

Background

Esthesioneuroblastoma (ENB) is a rare sinonasal malignancy that arises from the olfactory epithelium.¹ The incidence of cervical lymph node metastases in ENB at the time of initial diagnosis is between 5 and 12%, but most regional lymph node disease is delayed, arising 6 months to 20 years after primary treatment, with an overall incidence of 25%.^{1–3} The propensity for delayed neck disease in ENB poses a dilemma regarding whether there is a role for elective neck treatment in the clinically negative (N0) neck.^{1,4}

Case Report

A 59-year-old man presented with right-sided nasal obstruction, headaches, and a 4-cm right ethmoid mass involving the nasopharynx and anterior skull base with erosion of the right cribriform. Biopsy revealed a Hyams III ENB with lymphovascular invasion. Positron emission tomography (PET) found no evidence of cervical lymph node disease or distant metastasis (Kadish stage C). He underwent a combined endoscopic and open craniofacial resection and reconstruction with a pericranial flap and fat graft. He received adjuvant

radiation to the primary site (66 Gy) and elective bilateral neck irradiation (56 Gy). He is currently 12 months post-treatment with no evidence of disease recurrence.

Literature Review

Delayed neck metastases in ENB constitutes a significant predictor of poor overall survival (OS).⁵ Risk factors associated with cervical lymph node disease include high-grade Hyams pathology (Hyams III and IV), dural involvement, and positive surgical margins at the primary tumor resection site.^{5–7}

Even for those at high risk of developing neck disease, the challenge in predicting where potential nodal disease may arise gives rise to significant controversy surrounding the appropriate laterality of any elective neck treatment. ENB primary site laterality has been shown to be poorly predictive of neck disease laterality, as metastases in all levels of the neck, contralateral, and bilateral are well documented.^{8,9} For these reasons, elective neck dissection is not ideal in this population. Additionally, elective neck dissection poses significant morbidity risk without a demonstrated benefit in OS.¹⁰

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Elective neck irradiation (ENI) may improve regional disease control in high-risk patients. A meta-analysis of 489 patients reported 88% lower odds for regional nodal relapse with ENI compared to observation, but no difference in OS.⁴

Rather than performing ENI, some have suggested observation with salvage treatment if neck disease occurs. A systematic review of 45 patients with delayed neck metastases found that 68.8% failed salvage therapy, defined as disease recurrence within 1 year.¹¹ When stratified by salvage treatment modality, a combination surgery and radiotherapy increased the rate of successful neck salvage over either surgery or radiation alone.¹¹

In July 2019, the American Rhinologic Society published an International Consensus Statement recommending consideration of ENI in patients with high Hyams grade (III/IV) and Kadish stage C involving more than the dura for prevention of regional recurrence.¹⁰ Long-term surveillance (over 10–20 years) of the neck was also recommended.

Conclusion

Cervical lymph node disease in ENB can occur decades following primary treatment and is associated with decreased OS. The benefit of elective treatment of the NO neck is not well established, although ENI has been associated with improved regional control in some small studies. Salvage treatment has also been proposed, although no prospective head-to-head comparisons of elective versus salvage treatment have been performed.

Conflict of Interest

None declared.

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