Minimally Invasive Approach for Resection of Parameningeal Rhabdomyosarcoma

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Abstract

Background About one-third of rhabdomyosarcomas arise in the head and neck, with parameningeal primaries accounting for half of these. Principles of management involve chemotherapy, radiation, or both, in addition to surgical biopsy, debulking, and complete or near-complete resection. In the head and neck, diagnostic biopsies have historically been performed without attempt at resection due to proximity to critical structures and cosmetic considerations.

Methods Retrospective chart review of three cases of rhabdomyosarcoma at the cranial base managed through minimally invasive endoscopic surgical resection and adjuvant therapy.

Results Three patients were identified as having undergone endoscopic surgical debulking or margin-negative resection of a rhabdomyosarcoma of the cranial base. Two of three patients had complete resection based on intraoperative margin control. All three patients underwent adjuvant therapy within 1 month of diagnosis. Follow-up time ranged from 5 months to 3 years with all patients disease-free at last follow-up.

Conclusion Skull base surgeons should routinely be involved in multidisciplinary treatment planning for parameningeal rhabdomyosarcomas, as surgical options have evolved to allow for potential endoscopic resection with low morbidity and no or minimal delay in additional treatment options.

Keywords

- rhabdomyosarcoma
- parameningeal
- endoscopic
- minimally invasive

Introduction

Rhabdomyosarcoma is the most common sarcoma of childhood and the third most common extracranial solid tumor after neuroblastoma and Wilms tumor. It accounts for 4.5% of all childhood cancer.¹ About one-third of rhabdomyosarcomas arise in the head and neck, with parameningeal sites accounting for half of these. Parameningeal locations include the nasal cavity, nasopharynx, paranasal sinuses, parapharyngeal space, infratemporal fossa, pterygopalatine fossa, mastoid, and middle ear.² Due to proximity of critical structures, cosmetic considerations, and risk of cranial nerve and meningeal spread, parameningeal rhabdomyosarcomas are classically biopsied without attempt at tumor resection with resultant reduced survival outcomes.^{1–5} In recent years, studies have shown endoscopic sinonasal resection can result in tumor resection with acceptable morbidity in carefully selected patients.^{6,7}

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Current principles of management for rhabdomyosarcoma in general involve multimodality treatment with chemotherapy, surgical resection tailored to the tumor site, and radiation when indicated.⁸ Studies have shown disease-free and overall survival benefit to surgical resection. The Third Intergroup Rhabdomyosarcoma Study found that patients with localized, gross residual disease after initial surgery (surgical-pathologic group III) had a 5-year survival rate of approximately 70%, compared with approximately 80% for patients with microscopic residual tumor after surgery (group II), and more than 90% 5-year survival rate for patients without residual tumor after surgery (group I).⁹ Clinical grouping is an independent predictor of outcome. In current treatment paradigms, a patient's postoperative group is used in conjunction with tumor histology, location, and extent (local, regional, and systemic) to stratify prognosis and plan adjuvant treatment.¹ The goal of this study is to describe clinical outcomes of three cases in which parameningeal rhabdomyosarcoma was primarily surgically resected with minimal surgical morbidity and good disease control.

Methods

After obtaining internal review board approval for the proposed study, patients who underwent surgical resection of rhabdomyosarcoma were identified by the otolaryngologist performing surgery. The charts of identified patients were retrospectively reviewed. Intervention performed were debulking or complete surgical resection with negative margins of the tumor. The primary outcome was effect of surgery on tumor-induced symptoms. Secondary outcomes evaluated were margin status, time from diagnosis to surgery, time from surgery to adjuvant treatment, length of follow-up, and disease status at last follow-up.

Case Review

Patient 1

A 4-month-old female with a history of pharyngeal rhabdomyofibrosarcoma discovered at an outside hospital at birth was referred to the University of Michigan (UM) due to progression of disease despite four cycles of chemotherapy consisting of vincristine, adriamycin, and decadron. She underwent tracheostomy and gastrojejunostomy tube placement 4 days after birth due to respiratory distress and inability to take nutrition by mouth. Imaging revealed progression of disease, with an enhancing mass seen within the nasopharynx. The mass extended from the clivus to the level of C6, measuring 5 × 4 cm and obstructing both the nasopharynx and the oropharynx (→Fig. 1).

Repeat operative biopsy revealed a friable nasopharyngeal tumor based at the right lateral pharynx and confirmed her pathologic diagnosis. With the diagnosis confirmed, she underwent stealth-guided endoscopic transnasal and transoral resection with frozen section margin control. After surgery, there appeared to be resolution of nasal obstruction as well as improvement in apparent discomfort. The child was evaluated by a speech-language pathologist, who felt that she had an excellent prognosis for future speech and swallowing as a result of her resection, although at last follow-up, she was still tracheostomy and gastrostomy tube dependent. She went on to complete chemotherapy consisting of doxorubicin and ifosfamide. She had no evidence of disease at last follow-up, which was 10 months after surgery.

Patient 2

A 14-year-old male previously diagnosed with seasonal allergies not responsive to treatment presented to the UM emergency department (ED) with a 3-week history of foul smelling

Fig. 1 Preoperative imaging in a 4-month-old female reveals enhancing mass within the nasopharynx extending from the skull base to the level of C6, measuring 5 × 4 cm and obstructing both the nasopharynx and the oropharynx. (A) Coronal T1 magnetic resonance imaging (MRI). (B) Sagittal T2 MRI.

Fig. 2 Preoperative imaging in a 14-year-old male shows a 3.5×9 cm mass of the left nasal cavity extending posteriorly into the nasopharynx and laterally into the maxillary sinus. (A) Coronal T1 magnetic resonance imaging (MRI). (B) Sagittal T1 MRI.

anterior rhinorrhea, nasal obstruction, and intermittent epistaxis. He had lost 20 pounds in 1 month. On examination, he was found to have a large mass filling the left nasal cavity with rightward septal deviation obstructing the contralateral nares. Imaging revealed a 3.5×9 cm mass of the left nasal cavity extending posteriorly into the nasopharynx and laterally into the maxillary sinus (**Fig. 2**).

Endoscopic biopsy performed at bedside was interpreted as alveolar rhabdomyosarcoma. Full staging evaluation revealed no metastatic disease. After tumor board discussion and presentation of therapeutic options, he underwent stealth-guided expanded endonasal approach for resection of the left sinonasal tumor with frozen section margin control (**Fig. 3**). Final pathologic margins were negative. After resection, his nasal obstruction and rhinorrhea resolved.

His appetite improved and he began gaining weight. He went on to complete adjuvant chemoradiation consisting of vincristine, dactinomycin, cyclophosphamide, and 36-Gy radiation to the primary site. It is now over 3 years since resection and he continues to have no evidence of disease.

Patient 3

A 19-year-old male presented to the UM ED with a 1-month history of persistent right-sided rhinorrhea with progressive V2 hypesthesia and trismus. Examination revealed right orbital proptosis and periorbital swelling, a pale mass filling the right nasal cavity, and V2 hypesthesia. Imaging revealed a 5×4.5 cm mass filling the right nasal cavity and right maxillary sinus, abutting the right cribriform and inferior orbit (**Fig. 4**).

Fig. 3 Intraoperative photos. (A) Left nasal cavity during endoscopic tumor resection. (B) Left nasal cavity postresection.

Fig. 4 Preoperative imaging in a 19-year-old male reveals a 5 × 4.5 cm mass filling the right nasal cavity and right maxillary sinus abutting the right skull base and inferior orbit. (A) Coronal computed tomography (CT). (B) Sagittal CT.

Endoscopic biopsy was performed in the operating room and interpreted as alveolar rhabdomyosarcoma. He underwent a stealth-guided endonasal approach for subtotal resection of the tumor involving the orbit and skull base. Gross total resection was initially the goal, though was unable to be performed due to blood loss. The operation did result in resolution of nasal obstruction and periorbital edema. Given rapid growth and requirement for preoperative embolization, chemoradiation was advised rather than an additional operation that could delay definitive management. He went on to complete chemoradiation consisting of vincristine, dactinomycin, cyclophosphamide, and 55.8-Gy radiation to the primary site. He continues to be without evidence of disease 32 months after debulking resection.

► **Table 1** summarizes the clinical presentation, days from diagnosis to surgery, days from surgery to adjuvant, and disease status at last follow-up for each patient.

Discussion

These three cases illustrate that surgical resection with meaningful benefit to the patient is possible for parameningeal rhabdomyosarcoma despite close relationships with critical structures. A minimally invasive approach was selected for these patients after physical examination and careful review of imaging. No patients suffered complications from their surgical procedures, and surgical resection did not delay adjuvant therapy with time from diagnosis to adjuvant therapy ranging from 17 to 31 days. These patients benefited from immediate symptom improvement, which was primarily resolution of nasal obstruction.

While these cases have been traditionally defined as parameningeal, the authors propose that with new techniques and improved diagnostic imaging, nasopharynx, sinonasal, and cranial base fossae rhabdomyosarcoma should not be lumped together as “parameningeal.” Rather, these cases

Table 1 Clinical summary for three cases

	Tumor stage and type	Days from diagnosis to resection	Resection type	Days from resection to adjuvant therapy	Improved symptoms control?	Tumor status
Case 1	T2aN0M0	15	16	16	Yes	NED 5 mo postresection
	Rhabdomyofibrosarcoma					
Case 2	T2bN0M0	9	8	8	Yes	NED 36 mo postresection
	Alveolar rhabdomyosarcoma					
Case 3	T2aN0M0	15	3	3	Yes	NED 32 mo postresection
	Alveolar rhabdomyosarcoma					

should be defined by precise site and should only be considered parameningeal if tumor is abutting dura, intradural, intraparenchymal, or with evidence of perineural cranial base involvement on dedicated cranial base magnetic resonance imaging. Describing the location of the lesion with improved precision will allow the entire treatment team to consider all options for therapy, thereby opening communication lines to discuss the possibility for minimally-invasive resection.

Patients who undergo surgical resection with negative margins or even microscopically positive margins may also experience a survival benefit. Surgical resection with negative margins or microscopically positive margins has been shown to improve locoregional recurrence and overall survival.^{1,7,8} Patient 1 exemplifies this well. This patient underwent four cycles of chemotherapy with progression of disease prior to referral. Despite this, surgical resection with negative margins was possible, and the patient remains disease-free 10 months after surgery. Surgical resection may also reduce adjuvant treatment toxicity by reducing the dose of radiation required. This is particularly important for children who are growing and have many decades of life ahead of them.

Further study and reporting of outcomes after surgical resection of childhood parameningeal rhabdomyosarcoma is indicated to better characterize short- and long-term benefit to patients. Routine description of the tumor location should be as precise as possible.

Conclusion

Skull base surgeons should routinely be involved in multidisciplinary treatment planning for traditionally labeled parameningeal rhabdomyosarcomas, as surgical options have evolved to allow for improved resections with low morbidity

and no delay in chemotherapeutic or radiation treatment options. We propose limiting the term “parameningeal” to those tumors abutting or involving dura or with evidence of perineural involvement at the cranial base. This will facilitate more precise reporting of tumor location and treatment outcomes.

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