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Technical Note

# Expanded endonasal endoscopic approach for resection of a juvenile nasopharyngeal angiofibroma with skull base involvement

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ABSTRACT

Juvenile nasopharyngeal angiofibromas (JNAs) are rare vascular tumors which arise in the nasopharynx of adolescent males. Patients with these tumors can be cured by surgery, which is the treatment of choice in the majority of patients. Traditional surgical techniques for patients with JNAs have been via open surgical approaches. Since 2000, however, the surgical management of JNAs has changed due to advances in endoscopic procedures and such approaches are standard for early stage lesions which are limited to the nasal cavity, nasopharynx and the paranasal sinuses. The role and limitations of endoscopic approaches for JNAs with skull base and intracranial involvement are being defined. In this report, we describe a patient with a JNA with skull base involvement who underwent an expanded endonasal endoscopic approach for a complete resection. Additionally, we review the literature of endoscopic approaches to JNAs with skull base involvement.



## 1. Introduction

Juvenile nasopharyngeal angiofibromas (JNAs) are relatively rare, benign, vascular neoplasms which occur in adolescent males and are characterized by slow but locally aggressive growth. These tumors typically present with unilateral symptoms including nasal obstruction and epistaxis. Other associated signs and symptoms can include facial swelling and proptosis. Cranial nerve compression may result in diplopia, sensory changes on the face and visual loss.[1,2]

Although there is some debate regarding the specific cell of origin, JNAs typically originate from the superior margin of the sphenopalatine foramen.[1] From its origin, the tumor can grow into the nasal cavity and nasopharynx, paranasal sinuses, infratemporal fossa, orbit via the inferior orbital fissure and intracranial compartment. Intracranial extension can occur either through the middle cranial fossa anterior to foramen lacerum and lateral to the cavernous sinus and carotid artery or through the sella medial to the carotid artery and lateral to the pituitary gland.[1,3,4] There have been numerous staging systems described for JNAs based on the anatomic extent of the tumor.[3–5]

Patients with JNAs can be cured with complete excision and therefore surgical resection is the treatment of choice for this tumor.[6,7] Traditional surgical approaches for these lesions include transpalatal, transfacial (through a lateral rhinotomy or midfacial degloving) and infratemporal skull base approaches as well as combined surgical approaches.[8–14] More recently, endonasal endoscopic surgical approaches have become integral in the management of these tumors and are considered the treatment of choice for lesions limited to the nasal cavity and nasopharynx.[15–20] The utility and limitations of endoscopic techniques for more advanced lesions with skull base and/or intracranial involvement is less defined. In this report, we describe a patient with a large JNA with skull base involvement who underwent an expanded endonasal endoscopic approach for complete resection.

## 2. Case report

A 20-year-old man with no significant past medical history was referred to our institution for a nasopharyngeal mass discovered after an evaluation for intermittent, recurrent right sided epistaxis. A maxillofacial CT scan demonstrated a mass centered in the right posterior nasopharynx, with involvement and erosion of the sphenoid bone in the region of the base of the ptyergoids and clivus. There was erosion and widening of the right vidian canal. High resolution MRI of the skull base with isotropic T1-weighted and T2-weighted sequences and post-gadolinium T1-weighted sequences demonstrated the mass involving the right posterior nasal cavity and right nasopharynx. There was involvement of the sphenopalatine foramen with minimal extension into the medial portion of the pterygopalatine fossa. There was also involvement of the right

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side of the clivus and sphenoid bone. Within the sphenoid bone on the right, there was extension to the cerebral surface of the greater wing of the sphenoid with potential involvement of the dura in this region, corresponding to a Radkowski stage IIIA tumor (Fig. 1).[5]

Initial endoscopic examination demonstrated a large hyperemic mass occupying the entire right nasopharynx. Due to the vascularity of the mass, the patient was taken to the operating room for an endoscopic biopsy under general anesthesia. The pathology of this biopsy specimen returned as a JNA.

The patient underwent a preoperative angiogram which demonstrated vascular supply to the mass from both terminal and proximal branches of the right internal maxillary artery as well as the vidian artery (Fig. 2). The right internal maxillary artery branches supplying the tumor were embolized with Embospheres (BioSphere Medical, Inc., Rockland, MA, USA) and coil embolization; the vidian artery was not embolized. The patient was awakened from anesthesia, examined neurologically and then taken to the operating room for an expanded endonasal endoscopic approach and tumor resection.

Surgery was performed with neuronavigation using both the preoperative high resolution CT and MRI scans. The nasal cavity was injected with lidocaine and epinephrine; given the size and location of the tumor, the right sphenopalatine artery was injected via a trans-palatal injection. A right middle turbinectomy and uncinectomy were performed for endoscopic access to the tumor. A large maxillary antrostomy was then performed in conjunction with a total ethmoidectomy and sphenoidotomy. A left-sided nasoseptal "rescue flap"[21] preserving the vascular pedicle (left posterior nasal artery) was elevated for possible reconstruction. The inferior cut in the flap was not made to allow easy tacking to the remnant septum if the flap was not required for reconstruction of the surgical defect. A posterior septectomy was performed allowing binaral access.

The tumor was then defined circumferentially. The right sphenoid sinus was opened widely to the lateral most extent and the intersinus septum removed. The anterior aspect of the clivus was removed with a high-speed drill medially and inferiorly defining the margin of the tumor. The back wall of the maxillary sinus was removed with rongeurs to gain access to the pterygopalatine fossa and the lateral aspects of the tumor. Given the size of the lesion, the anterior aspect of the tumor in the nasal cavity and nasopharynx was amputated using endoscopic scissors and debulked with a microdebrider to enable better visualization of the tumor invading the skull base and clivus. Bleeding from the vidian artery was controlled with the bipolar and suction cauteries.

The medial and inferior aspects of the clivus were found to have extensive tumor invasion and were further reduced with the drill until normal appearing bone was encountered. Using various dissectors, tumor was reflected laterally and numerous fronds of tumor were removed from bony cavities in the clivus and skull base (Fig. 3). As the tumor resection continued laterally along the skull base, the vidian artery and canal were identified. A large tumor frond within the vidian canal was identified and removed further exposing the vidian artery and nerve which were coagulated with the bipolar cautery and cut sharply (Fig. 3). The tumor resection then continued laterally along the skull base. Given the corrugated and mottled appearance of the bone as well as the very fibrous nature of the tumor, the fronds were gently pulled from the bony crevices. This resection continued along the skull base posteriorly towards foramen ovale and foramen lacerum and laterally to the lateral pterygoid plate. Inferiorly, the tumor was

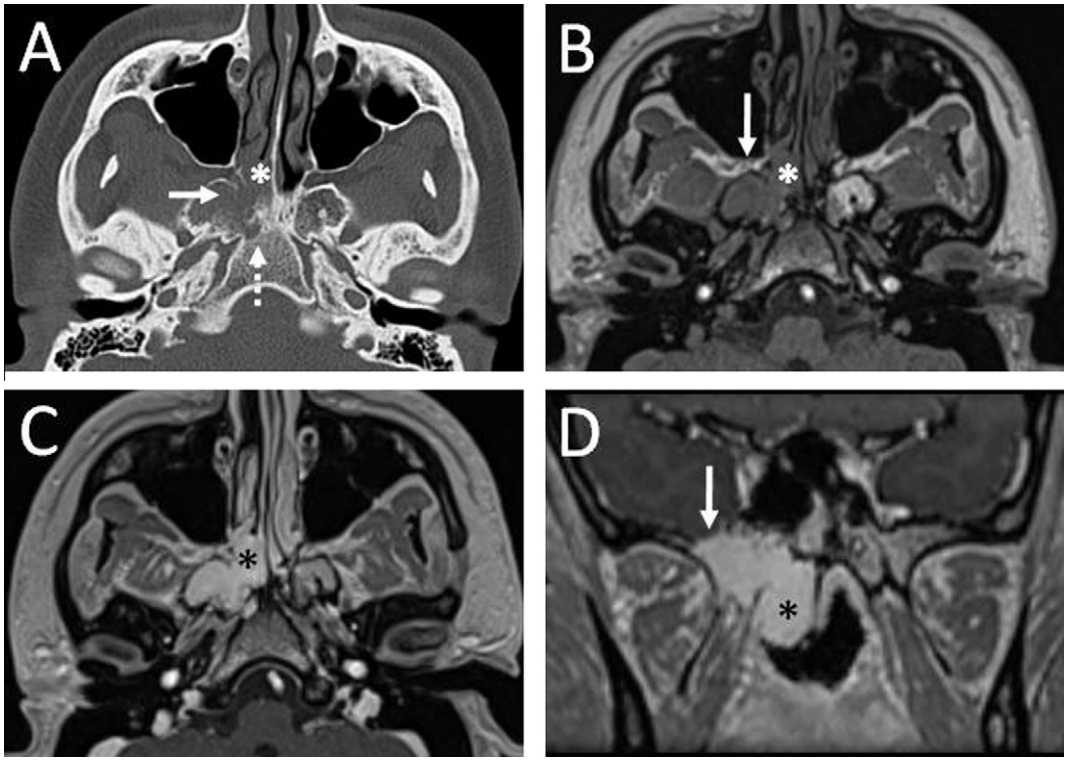


**Fig. 1.** Soft tissue mass compatible with a juvenile nasopharyngeal angiofibroma centered in the right posterior nasal cavity and nasopharynx (asterisk). (A) Axial CT scan showing an expansile lytic component of the mass within the right pterygoid (arrow) and with focal clival erosion (interrupted arrow). (B) Axial precontrast T1-weighted MRI showing preservation of fatty signal within the pterygopalatine fossa (arrow) except in the region of the sphenopalatine foramen. (C) The mass enhances intensely on the corresponding axial T1-weighted postcontrast MRI. (D) Coronal reformat of post contrast T1-weighted imaging demonstrates involvement of the floor of the right middle cranial fossa (arrow).