Craniofacial Resection of Advanced Juvenile Nasopharyngeal Angiofibroma

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Objective: To describe the results of a craniofacial approach to resection of stage IIIB juvenile nasopharyngeal angiofibroma, performed by an integrated skull base surgical team.

Design: A retrospective case-series review was conducted with postoperative follow-up ranging from 28 to 63 months.

Setting: Operations were performed at a tertiary medical center.

Patients: A referred sample of 5 male patients, ranging in age from 10 to 23 years (mean, 15 years).

Interventions: All patients underwent resection of nasopharyngeal angiofibromas with intracranial extension. The procedure involved an infratemporal fossa approach via zygomatic osteotomy and subtemporal craniectomy. Anterior exposure was gained through a standard facial translocation. Dissection of the cavernous carotid artery was required in 3 patients.

Main Outcome Measures: Intraoperative and postoperative morbidity.

Results: The average operating time was 12 hours 47 minutes. Estimated blood loss ranged from 700 to 1750 mL (mean, 1120 mL), with 2 patients requiring intraoperative transfusion. Patients were hospitalized for a mean duration of 5.6 days. Long-term morbidity includes facial dysesthesia, nasal crusting, and malodorous nasal discharge. No patients sustained stroke, oculomotor dysfunction, vision loss, or auditory impairment. At most recent follow-up, which ranges from 28 to 63 months, tumor recurrence has been confirmed in 1 patient.

Conclusions: A combined craniofacial approach is appropriate for juvenile nasopharyngeal angiofibroma that extends intracranially. Complete tumor removal with acceptable morbidity can be expected.

Neurosurgery. The patients were white males, ranging in age from 10 to 23 years (mean, 15 years) at the time of surgery. All cases represented initial tumor presentations, with the exception of 1 patient referred with a recurrent tumor 23 months after his initial surgery. Each patient presented with evidence of intracranial extension as determined by imaging studies. Initial patient evaluation involved a comprehensive head and neck history and physical examination, including fiberoptic examination of the nasopharynx. Radiological staging was obtained by computed tomography and magnetic resonance imaging. Studies were reviewed by a neuroradiologist specializing in head and neck imaging. Angiography was performed to identify primary feeding vessels to the tumor and to selectively embolize them prior to surgery. All embolizations took place within 24 hours of tumor resection.

Surgery was initiated with an infratemporal fossa approach via zygomatic osteotomy and subtemporal craniectomy (Figure 1). After complete exposure and dissection of the tumor from the sphenoid sinus, the superior orbital fissure, the middle cranial fossa, and the lateral cavernous sinus, anterior exposure was gained through a standard facial translocation (Figure 2). The additional exposure permitted tumor mobilization from the nasopharynx, the paranasal sinuses, the pterygopalatine fossa, the infratemporal fossa, and the cavernous sinus.

Figure 1. Hemicoronal and Weber-Fergusson incisions.

Figure 2. Bone cuts associated with a standard facial translocation.

After induction of anesthesia, a subarachnoid lumbar drainage catheter was inserted to facilitate intraoperative brain relaxation. The patient was placed supine with his head extended and rotated 60° away from the intracranial tumor bulk. A hemicoronal skin incision was initiated in the preauricular region and extended behind the hairline. A scalp flap was subsequently elevated to expose the underlying zygomatic bone and arch. The flap, which extended from the zygomaticomaxillary suture to the external auditory canal, incorporated the temporal fat pad to preserve the frontal branch of the facial nerve. A zygomatic osteotomy was performed, and the temporalis muscle was reflected inferiorly through the resulting defect. Single burr holes were drilled at the pterion and above the zygomatic root. A bone flap was turned with a craniotome and stored in a covered basin of sterile isotonic sodium chloride solution. Perimeter holes were then placed to allow the dura to be secured with suture.

Under an operating microscope, the dura was dissected from the floor of the middle cranial fossa. The floor was then entered with a high-speed pneumatic drill to sequentially expose the following structures: (1) the superior orbital fissure; (2) the foramen rotundum with the second division of the trigeminal nerve (V2); (3) the foramen ovale with the third division of the trigeminal nerve (V3) and the accessory meningeal artery; (4) the foramen spinosum with the middle meningeal artery and vein; (5) the greater superficial petrosal nerve; and (6) the carotid canal. The middle meningeal vessels and the trigeminal nerve divisions were selectively divided to provide access to the tumor extensions in the middle cranial fossa and the cavernous sinus. Additional removal of bone medial to the foramen rotundum provided a lateral approach to the tumor within the sphenoid sinus. Likewise, drilling medial to the foramen ovale permitted access to the tumor abutting the petrous carotid artery. In cases of orbital tumor extensions, an orbitozygomatic osteotomy was performed to facilitate tumor dissection from the orbital fissures.

After the superior aspect of the angiofibroma had been completely mobilized, the transfacial approach was initiated through a Weber-Fergusson incision that extended through the underlying bone. The resulting cheek flap was turned to expose the piriiform aperture, the inferior orbital rim, and the inferior aspect of the anterior maxillary wall. The orbital contents were then reflected posteriorly to expose the lacrimal sac. The lacrimal sac was divided, the orbital contents were retracted, and the soft tissues lining the inner aspect of the piriiform aperture were elevated. Multiple bone cuts were then made to allow the anterior maxilla and overlying cheek flap to be laterally reflected as a vascularized osteoplastic flap (Figure 3). Specifically, cuts were made (1) from the superior piriiform aperture to the inferomedial orbit; (2) across the inferior aspect of the maxillary buttress; (3) across the anterior wall of the maxilla; and (4) through the lateral orbit. The bone of the posterior maxillary wall was removed, and the internal maxillary artery was clamped and divided. The pterygoid plates were then removed with a high-speed pneumatic drill from either the transfacial or the cranial approach. Working concomitantly from above and below the skull base, the tumor was dissected and removed en bloc. Although the presence of a tumor pseudocapsule largely
obviated the need for frozen sections, they were used in isolated cases in which a plane of dissection could not be clearly identified. Specifically, multiple frozen sections were obtained from the eustachian tube region in 2 patients and from the pterygoid musculature in 1 patient. Successive sections were sent when necessary to ensure that all operative site margins were free of tumor.

Upon achieving complete tumor removal and hemostasis, the temporalis muscle was split. One half of the muscle was folded under the temporal lobe and placed into the sphenoid sinus. The craniotomy flap was then restored to its anatomic position and secured with a plating system (Synthes 1.3; Synthes, Paoli, Pa). The wound was passively drained, and the scalp flap was closed in 2 layers.

Prior to closing the facial wound, an internal dacryocystorhinostomy was performed. Specifically, the divided lacrimal sac was incised vertically and marsupialized with 2 sutures. The facial bone flap was then replaced and secured with miniplates. The entire nasal cavity was packed with gauze impregnated with antibiotic ointment. A Foley catheter served as a buttress against which the packing was placed. The skin incision was closed in layers.

RESULTS

The most common presenting symptom, reported by all 5 patients, was unilateral nasal obstruction. Three of 5 patients also developed severe recurring epistaxis or eustachian tube dysfunction. Two of 5 patients experienced sinus pressure, rhinitis, and/or anosmia. Symptom duration prior to presentation ranged from 1 to 9 months. Physical examination revealed a unilateral, fleshy, red mass obscuring the nasal cavity in 4 of 5 patients. Tumor extension into the nasopharynx was demonstrated via fiberoptic nasopharyngoscopy in 3 of these patients, with 1 individual exhibiting posterior displacement of the soft palate. Facial deformity in the cheek area was also noted in 2 of the 5 patients. One of these individuals exhibited marked proptosis.

Radiological evidence of tumor extension and sites of intracranial involvement for each patient are presented in Table 1 and representative imaging studies are shown in Figure 4 and Figure 5A-C. Skull base invasion and/or intracranial extension were demonstrated via computed tomography or magnetic resonance imaging in all 5 cases. All tumors extended into the cavernous sinus, and 3 tumors abutted the internal carotid artery. Invasion of the vidian canal was demonstrated in 4 patients. Orbital involvement with tumor extension through the inferior and/or superior orbital fissures was noted in 3 of 5 patients.

Angiography showed that the majority of tumors were supplied by branches from both the internal and external carotid arteries. Branches of the external carotid artery included the internal maxillary and the accessory meningeal arteries, while those of the internal carotid artery included the mandibulovidian (Figure 5D) and the cavernous arteries. Preoperative embolization of external carotid artery branches was accomplished without complications in all patients. Thirty-minute balloon-occlusion tests of the internal carotid artery ipsilateral to the primary tumor were also conducted in 3 patients. This procedure was performed under fluoroscopic guidance following systemic heparinization. No patients ex-

Figure 3. Reflection of a vascularized anterior maxillary osteoplastic flap.

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*SS indicates sphenoid sinus; ES, ethmoid sinus; CCS, carotid cavernous sinus; IF, infratemporal fossa; MCF, middle cranial fossa; PPF, pterygopalatine fossa; OF, orbital fissure; IMA, internal maxillary artery; U, unilateral; MS, maxillary sinus; VC, vidian canal; ET, eustachian tube; AMA, accessory meningeal artery; CB, cavernous branch; MVB, mandibulovidian branch; ICA, internal carotid artery; B, bilateral; and ACF, anterior cranial fossa.
to 63 months, has revealed minimal postoperative complications. The most common complication is nasal crusting, found in 4 patients. Serous otitis media was present in 3 patients at the initial postoperative evaluation, but has resolved completely in all cases. Two patients developed facial anesthesia due to sectioning of the second division of the trigeminal nerve. One patient has loss of sensation in the second and third division territories, the other experiences a deficit only in the second division territory. Additional isolated complications included transient facial lymphedema, intermittent eye irritation, chronic sinusitis, and a persistent malodorous nasal discharge.

Recurrent disease has been confirmed in 1 patient who presented 19 months after resection with complaints of epiphora and intermittent swelling in the medial canthal region. Magnetic resonance imaging revealed a cystic lesion that was centered in the pterygopalatine fossa and extended into the right orbit, the right sphenoid sinus, and the medial wall of the right middle cranial fossa. Initial management included cyst aspiration, which failed to yield cerebrospinal or other fluid contents, and marsupialization through an endoscopic approach. Four months after this procedure, follow-up magnetic resonance imaging showed replacement of the cyst with enhancing solid tumor. Given the distribution of disease in the cavernous sinus and along the second division of the trigeminal nerve, the tumor was deemed unresectable, and the patient was referred for radiation treatment. Representative radiographic images are shown in Figure 6A and B. Follow-up of a second patient has also revealed an area suspicious for recurrence near the petrous carotid artery (Figure 7). This area has been monitored at 3-month intervals, with no evidence of change over the last 18 months. The stability of the lesion is most consistent with postoperative scarring. However, the region will continue to undergo observation for evidence of recurrent disease.

The epidemiological profile, clinical presentation, and diagnostic evaluation of this sample are consistent with previously published reports. Although isolated cases of JNA have been observed in adult and female populations, the tumor typically occurs in pubescent boys. The classic presentation includes nasal obstruction, recurrent epistaxis, and the presence of a smooth, lobulated, red-gray mass in the posterior nasal cavity or nasopharynx. Physical examination may reveal facial deformity, proptosis, palate expansion, serous otitis, and visual or auditory impairment. Our patients were initially evaluated 1 to 9 months after the onset of these characteristic signs and symptoms. Their delay in presentation can be attributed to a tendency to associate the indolent symptoms of JNA with more common disease entities, such as rhinitis, sinusitis, and antrochoanal nasal polyps.

Characteristic routes of JNA invasion are anterior infiltration of the nasal cavity, anterolateral erosion of the posterior maxillary sinus wall, and/or anterosuperior destruction of the ethmoid air cells. Lateral growth leads to invasion of the pterygopalatine fossa, erosion of the pterygoid plates, and potential extension into the infra-

Figure 4. A 10-year-old boy with juvenile nasopharyngeal angiofibroma. A, Axial T2-weighted (repetition time [TR], 2500 milliseconds; echo time [TE], 90 milliseconds) magnetic resonance image with fat suppression. There is extension of the patient’s juvenile angiofibroma into the posterior ethmoid air cells and sphenoid sinus (arrows). In addition, neoplasm has extended from the pterygopalatine fossa to the foramen rotundum (arrowheads), the inferior orbital fissure, and the masticator space (white arrow) via the pterygomaxillary fissure. Neoplasm abuts the precavernous right internal carotid artery (curved arrow). B, Enhanced fat-suppressed coronal T1-weighted (TR, 600 milliseconds; TE, 17 milliseconds) magnetic resonance image. This again shows neoplasm extending to the sphenoid sinus (arrows) and foramen rotundum (arrowhead), with frank invasion of the pterygoid bone and vidian canal (curved arrow).

Figure 6. A and B. Follow-up of a second patient has also revealed an area suspicious for recurrence near the petrous carotid artery (Figure 7). This area has been monitored at 3-month intervals, with no evidence of change over the last 18 months. The stability of the lesion is most consistent with postoperative scarring. However, the region will continue to undergo observation for evidence of recurrent disease.

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temporal fossa via the pterygomaxillary fissure. From the infratemporal fossa, JNA may invade the inferior orbital fissure and erode the greater wing of the sphenoid bone. The tumor may also exhibit intracranial extension. Specifically, JNA may enter the middle cranial fossa by destroying the superior bony region triangulated by the foramen rotundum, foramen ovale, and foramen lacerum. Further extension from the middle cranial fossa tends to respect the dural barrier, such that the tumor remains lateral to the cavernous sinus. However, particularly aggressive angiofibromas may invade the cavernous sinus and threaten multiple cranial nerves (III, IV, V [divisions 1 and 2], and VI), the internal carotid artery, the hypophysis, the optic chiasm, and the lateral wall of the sphenoid sinus. A second pattern of intracranial destruction may arise in cases of posterosuperior extension from the tumor origin. In these situations, JNA spreads into the nasopharynx, erodes through the inferior wall of the

Figure 5. A 13-year-old boy with juvenile nasopharyngeal angiofibroma with skull base extension and invasion of the left cavernous sinus. A, Axial unenhanced T1-weighted (repetition time [TR], 600 milliseconds; echo time [TE], 17 milliseconds) magnetic resonance image at the level of the skull base/clivus. Abnormal tissue is seen in the region of the left vidian canal (white arrow), and neoplasm is seen along the horizontal portion of the left petrous internal carotid artery (arrowheads). B, Axial enhanced fat-suppressed T1-weighted (TR, 600 milliseconds; TE, 17 milliseconds) magnetic resonance image obtained at the same level as panel A. Enhancing tumor is seen in the skull base at the left vidian canal (black arrow), and medial to and abutting the left petrous internal carotid artery (arrowheads). C, Axial T-weighted (TR, 2500 milliseconds; TE, 85 milliseconds) magnetic resonance image obtained at the level of the cavernous sinus demonstrates hypointense tumor in the left cavernous sinus (arrows). Again, tumor abuts the left internal carotid artery (curved arrow). D, Lateral cerebral angiogram obtained following selective catheterization of the proximal left internal carotid artery. This image demonstrates supply to the neoplasm from the mandibulovidian artery (arrows).
sphenoid sinus, and enters the cavernous sinus from an inferomedial approach.

Angiofibromas with significant intracranial extension have historically been associated with increased rates of uncontrollable hemorrhage, neurological deficits, subtotal resection, and recurrence. A recent analysis suggests that tumors that infiltrate the infratemporal fossa, the sphenoid sinus, the base of the pterygoids, the cavernous sinus, the foramen lacerum, and the anterior fossa are most vulnerable to incomplete excision and recurrence. All tumors in our series extended into at least 3 of these regions.

Since 1981, several classification systems have been proposed to exploit the predictive value of site extension. We used the most recently revised system, introduced by Radkowski et al. in 1996 (Table 2). This system reflects the incremental rise in tumor recurrence observed at progressively higher levels of skull base/intracranial involvement. Based on this approach, all tumors in our series were classified as stage IIIB angiofibromas.

Our preoperative assessment of patients was facilitated by the development of modern imaging techniques. Since 1974, head computed tomographic scans have been used to identify characteristic patterns of bone erosion by JNA. Axial and coronal sections permit visualization of septal deviation, orbital fissure expansion, hard palate displacement, and erosion of the medial pterygoid plate, the maxillary sinus, and the basisphenoid. Computed tomographic studies may also reveal anterior bowing of the posterior wall of the maxillary antrum. Otherwise known as the Holman-Miller sign, this finding is classically—though not exclusively—associated with JNA.
For more than a decade, the diagnostic contributions of computed tomography have been supplemented by the use of magnetic resonance imaging. This technique had a dramatic impact on the evaluation of intracranial extension in our series. Magnetic resonance imaging offers improved soft tissue resolution combined with multiplanar capabilities. These features enhance the evaluation of intracranial landmarks and facilitate the differentiation of tumor from the surrounding mucosa, brain tissue, and paranasal sinus contents. Moreover, they allow a detailed assessment of spatial relationships between the tumor and vital intracranial structures. This was especially critical in our assessment of tumor extension into the cavernous sinus.

The vascular nature of JNA has made embolization an important component of preoperative management. Although this procedure is not associated with reductions in intraoperative hemorrhage in all reports, it has been shown to limit blood loss in patients with the level of advanced disease observed in this series. Early experience has underscored the importance of performing embolization in the immediate preoperative period (1-2 days prior to surgery), as premature vessel occlusion can promote collateralization and limit the efficacy of the procedure. We also performed temporary balloon-occlusion tests of the internal carotid artery in select cases of cavernous sinus involvement. This additional step allowed us to assess the potential impact of vessel ligation during resection.

Surgical resection has been the preferred treatment for extracranial JNA since 1955. However, the management of tumors with significant intracranial extension continues to be a source of considerable controversy. Numerous alternatives to surgery include hormonal treatment, chemotherapy, and external-beam radiation therapy. Despite promising preliminary studies, hormonal treatment has received little attention, and chemotherapy has been largely abandoned due to associated toxic effects. Radiation has been advocated as both a primary treatment and as an adjunct to surgery. Support for this approach is primarily derived from a single study of 55 patients treated for JNA with moderate-dose radiation (3000 rad or 3500 rad [30 or 35 Gy]). In contrast to prior reports of inadequate tumor control, this series demonstrated an 80% initial control rate with limited long-term complications. However, the well-known risks of radiation-induced secondary malignancies (eg, sarcoma and thyroid carcinoma) and growth arrest have fueled ongoing skepticism about the use of this approach in a pediatric population.

Surgical excision of extensive intracranial JNA has historically been associated with mortality. In recent decades, advanced approaches to the skull base, improved imaging techniques, and selective arterial embolization have fostered more aggressive surgical resections. Multiple approaches to intracranial angiofibromas have been described, including an infratemporal fossa approach, a total maxillectomy, a midfacial degloving approach, and an extended transcervical approach.

Among these techniques, the infratemporal fossa approach has been reported to remove all gross tumor in 80% of patients and to have the lowest rate (6%) of recurrence. This approach provides wide lateral exposure to the internal carotid artery and the cavernous sinus. However, tumor that is medial to the abducens nerve (VI) in the cavernous sinus is inaccessible and deemed unresectable. The infratemporal fossa approach minimizes facial scarring, but creates a depression when the temporalis muscle is used to reconstruct operative defects of the middle cranial fossa floor. This depression can cause facial and orthodontic asymmetry in addition to cosmetic deficits. Finally, the infratemporal fossa approach involves subtotal petrosectomy, which necessitates sacrifice of the pneumatic middle ear cleft and eustachian tube. The procedure is therefore associated with unilateral conductive hearing loss in nearly all cases.

In contrast, a combined transfacial and infratemporal fossa approach permits access to the sphenoid sinus, cavernous sinus, anterior skull base, and nasopharynx. We performed facial translocations in combination with the lateral approach in all 5 patients. The estimated average blood loss in our patients was more than 50% lower than the mean blood loss reported for the infratemporal fossa approach (1120 mL vs 2400 mL). This reduction in intraoperative hemorrhage suggests that the combined approach may diminish the need for transfusion through improved tumor exposure and vessel control. This approach also promotes complete resection by maximizing exposure to the nasopharynx, the sphenoid sinus, the pterygopalatine fossa, the infratemporal fossa, and both the medial and lateral aspects of the cavernous sinus. Complete tumor excision was achieved in all 5 cases in this series. There has been confirmed evidence of tumor recurrence in 1 patient, and follow-up of a second patient has revealed an area suspicious for recurrence that will continue to be monitored.

Our technique has been associated with minimal postoperative complications to date. The most significant morbidity has been diminished sensation in the distribution of the second and third divisions of the trigeminal nerve. Although 2 patients report some loss of facial sensation 32 months after surgery, prior studies suggest that sensation may improve over time. Indeed, facial anesthesia associated with V2 and V3 resection through the infratemporal fossa approach has been shown to improve by 25% to 50% in the majority of affected patients after an average follow-up of 2 years. Additional complications in our series, including nasal crusting, serous otitis media, and chronic sinusitis, have completely or partially resolved with appropriate management. While the majority of complications have been treated conservatively, 1 patient required permanent pressure equalization tube placement.

Until recently, the application of craniofacial surgery in the pediatric population has been limited by concerns that facial growth would be disrupted by osteotomies. These concerns have not been substantiated. A recent review of craniofacial procedures in children aged 3.5 to 14 years for multiple intracranial pathologies reported no evidence of arrest in facial skeletal growth in patients 1 to 5 years after surgery. Likewise, none of the patients in our series have demonstrated facial asymmetry at long-term follow-up.
In summary, extensive angiofibromas that invade intracranially and extend to the cavernous sinus may be safely resected through a combined approach. The temporal craniectomy provides excellent exposure to the carotid artery, cavernous sinus, and superior orbital fissure, while the transfacial approach allows for complete removal of tumor in the nasopharynx, sphenoid sinus, and medial cavernous sinus. Successful outcomes are ultimately determined by careful patient selection, imaging that clearly delineates the anatomic extent of tumor invasion, safe preoperative embolization, and the collaboration of experienced head and neck and skull-based surgery teams.

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