Endoscopic Laser-Assisted Excision of Juvenile Nasopharyngeal Angiofibromas

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Background: Juvenile nasopharyngeal angiofibromas (JNAs) are highly vascular tumors that originate in the nasopharynx of young males. The primary treatment is surgical excision. Traditional surgical approaches are associated with significant morbidity and facial deformity. We introduce and outline the clinical advantages of an endoscopic surgical approach to JNAs using the Nd:YAG laser with image-guided surgery.

Design: Case series.

Setting: Tertiary care medical center.

Patients and Methods: Our study included 5 male patients (age range, 8-21 years) with extensive JNAs. Their tumors were large and ranged from Fisch stage IIA to IIIA. Embolization of tumor-feeding vessels was performed before surgery. The tumors were photocoagulated via a transnasal endoscopic approach using a Nd:YAG laser. Devascularized, lased tumor was removed with a microdebrider. Image-guided navigation systems were used to assist skull base tumor removal, and sublabial and buccal incisions were used as needed to gain lateral endoscopic tumor access. Endoscopic tumor margins were obtained for frozen section.

Results: All patients achieved symptomatic remission, with no complications. No blood transfusions were necessary. The patients were ready for discharge 1 to 2 days after surgery. Postoperative and magnetic resonance imaging scans showed 2 skull base recurrences, which were removed endoscopically. Follow-up ranged between 2 and 3 years.

Conclusions: Traditional external surgical approaches to large JNAs may result in significant morbidity. Laser-assisted image-guided endoscopic excision of JNAs is a safe and effective minimally invasive surgical treatment. Its distinct advantages include (1) diminished blood loss, (2) superior cosmesis without observed altered facial growth, (3) direct access of skull base with minimal morbidity, and (4) ease of endoscopic follow-up.


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Juvenile nasopharyngeal angiofibromas (JNAs) are aggressive, poorly encapsulated, and highly vascular but histologically benign tumors affecting young males. The etiology of JNAs is not well understood. Although JNAs are slow growing as a rule, they are associated with significant morbidity caused by hemorrhage and extensive local invasion. Standard presenting symptoms include unilateral nasal dyspnea and epistaxis. Facial edema, proptosis, hearing loss, and neurologic changes occasionally follow due to local spread of the tumor. Treatment options include surgery, radiotherapy, hormonal manipulation, chemotherapy, and cryosurgery.

Reports of JNAs date back to antiquity. Surgical excision remains the preferred standard treatment. Hippocrates attributed the oldest recorded surgical treatment which involved a “longitudinal splitting of the ridge of the nose.” More than 55 types of JNA surgical procedures have been reported in the literature. Today, common surgical approaches include lateral rhinotomy, transpalatal, midfacial degloving techniques with medial maxillectomy and/or Le Fort I osteotomy, facial translocation, and craniofacial resection. Unfortunately, these open surgical approaches may lead to diminished development of the pediatric facial skeleton and are associated with potentially great blood loss.

With the advent of minimally invasive endoscopic techniques, lasers, and enhanced operative imaging modalities, new surgical methods are available to minimize skeletal deformity and blood loss. There have been recent isolated case reports of the transnasal endoscopic approach for the treatment of small JNAs of the nasopharynx. We have extended these applications to describe an endoscopic laser-assisted, image-guided surgical technique to safely excise large extracranial JNAs with diminished blood loss and facial deformity.

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Our study is a retrospective review of the clinical charts of 5 patients who presented with JNAs from January 1999 to July 2000 at Walter Reed Army Medical Center, Washington, DC. All patients underwent endoscopic laser-assisted image-guided excision of large JNAs. We also successfully treated 1 male patient with a sinonasal glomangioma and 1 female patient with a vascular sinonasal hamartoma with this operative technique but did not include them in this review because the histopathologic diagnosis was not JNA.

REPORT OF CASES

CASE 1

A 12-year-old boy presented with a 4-month history of progressive right-sided nasal obstruction, snoring, and intermittent epistaxis. Office endoscopy revealed an erythematous, bulging mass obstructing the posterior nasal cavity, with engorged blood vessels along the surface. A magnetic resonance imaging (MRI) scan confirmed the presence of a JNA with extension from the nasopharynx to the pterygopatilatine fossa, orbital apex, sphenoid sinus, and clivus (Figure 1). Image-guided sinonasal computed tomography (CT) was performed. Bilateral carotid angiography with selective bilateral external carotid embolization was performed without complication 1 day before surgery. With the patient under general anesthesia, the nasal cavity was topically vasoconstricted, and rigid nasal endoscopy was performed. Laser safety precautions were taken. A 0.6-mm fiberoptic Nd:YAG fiber was introduced endoscopically. Surface and interstitial laser tumor photocoagulation was performed at a 4- to 10-W continuous setting (Figure 2). The majority of the tumor’s blood supply was noted at the tumor periphery. Straight and curved microdebriders (StraightShot; Medtronic Xomed Inc, Jacksonville, Fla) were used to excise the devitalized lased tumor (Figure 3). A wide middle meatal antrostomy was performed with cutting forceps. The posteromedial maxillary sinus wall was removed with a curette, and the JNA extending into the pterygomaxillary space was lased and removed. There was tumor extension was into the sphenoid sinus, eroding through the sinus floor and extending into the clivus. Computed tomography image-guided surgery (InstaTrak, General Electric Medical Systems, Waukesha, Wis) facilitated tumor removal at the skull base. Tumor against the skull base was removed with Blakesly forceps, and endoscopic frozen sections along the clivus, pterygomaxillary space, para- tubal region, and sphenoid sinus were obtained. Submucosal tumor invasion into the clivus was noted on frozen section. Portions of the anterior clivus were removed until negative margins were obtained. The total blood loss was estimated to be less than 400 mL. The extended sinonasal cavity was gently packed with nasal sponges (Merocel; Mystic, Conn), and the patient was ready for discharge within 2 days. Packing was removed without bleeding in the clinic 4 days after surgery. Postoperative MRI scans obtained 6 months and 1 year after surgery showed no evidence of recurrence or residual tumor (Figure 4). Serial endoscopy performed at intervals for 3 years after surgery showed no evidence of tumor recurrence.
CASE 2

A 14-year-old boy presented with significant nasal dyspnea and recurrent epistaxis. Physical examination demonstrated an erythematous, smooth-walled mass obstructing the left choana and extending into the nasal cavity. An MRI scan revealed a JNA with extension to the left pterygopatine fossa, infratemporal fossa, clivus, and foramen rotundum (Figure 5). Endoscopic laser-assisted image-guided surgery, as described in case 1, permitted tumor excision. The lateral extent of the tumor was removed via a sublabial incision with a Caldwell-Luc approach and endoscopic removal of the posterior wall of the maxillary sinus. The estimated blood loss was 400 mL. There were no complications. Approximately 6 months after surgery, endoscopic examination revealed a recurrent JNA at the clivus. The recurrence was removed endoscopically, and the patient has remained asymptomatic since revision surgery. Postoperative MRI scans obtained 6 months and 1 year after surgery showed no evidence of recurrence or residual tumor (Figure 6). Serial endoscopy performed at intervals for 3 years after surgery showed no evidence of tumor recurrence.

CASE 3

A 16-year-old boy presented with intermittent epistaxis and unilateral nasal dyspnea. Physical examination revealed a bulbous pink mass obstructing the right nasal cavity, with evidence of clotting along the inferior periphery. An MRI scan confirmed the diagnosis of JNA, with tumor in the nasopharynx eroding into the pterygopalatine fossa, sphenoid sinus, and clivus. Embolization of both external carotid tumor vessels was accomplished 1 day before surgery. Transnasal endoscopic laser-assisted CT image-guided surgery was used to remove the gross tumor. Intraoperative frozen section revealed that multiple areas of normal-appearing mucosa in the sphenoid sinus and clivus were positive for tumor, with submucosal spread. Negative margins were obtained with difficulty. The estimated blood loss was 200 mL. No complications were observed. On routine examination 3 months after surgery, 2 small masses near the clivus and lateral sphenoid sinus were noted endoscopically. An MRI scan revealed regrowth. Revision endoscopic surgery was used to remove the masses, and there has been no recurrence noted on MRI or endoscopy for 3 years.

CASE 4

An 8-year-old boy with a 6-month history of a progressively enlarging right-sided facial mass presented with prolate “audible epistaxis.” Gauze packing was used to control the hemorrhage after the patient lost approximately 600 mL of blood (hematocrit, 27%). He was immediately flown to Walter Reed Army Medical Center. Office examination revealed a huge JNA visibly distorting the cheek, zygoma, and orbit. An MRI scan revealed a large tumor involving the right infratemporal fossa and extending through the inferior orbital fissure to the middle cranial fossa. The JNA also involved the right ethmoid and sphenoid sinuses, orbital apex, and cavernous sinus (Figure 7). Arteriography showed tumor vessel feeders from both external and internal carotid systems. Embolization was performed with absorbable gelatin foam and glue 1 day before surgery. Sub-
labial and buccolabial incisions were used to gain full access to this huge tumor and to enhance endoscopic removal of the infratemporal, parapharyngeal, and pterygomaxillary portions (Figure 8). The JNA in the ethmoid and sphenoid sinuses, clivus, and cavernous sinus was removed through the transnasal endoscopic approach. Computed tomography and MRI navigation systems were used to guide extensive tumor excision and laser photocoagulation. The estimated blood loss was 600 mL. No complications were encountered, and the nasal sponges were removed 3 days after surgery. Follow-up endoscopy and MRI scans revealed that only a small tumor encasing the right cavernous carotid remained (Figure 9). The patient’s facial deformity resolved, and he was discharged 2 days after surgery, without complication. He has been asymptomatic and without tumor growth for more than 2 years.

CASE 5

A 21-year-old man presented with nasal obstruction and recurrent epistaxis 1 year after a JNA was removed via a midface degloving, Le Fort I osteotomy with palatal drop, and temporary tracheotomy. Significant tumor regrowth was noted at the skull base, nasopharynx, and pterygopalatine fossa. Computed tomography image guidance with transnasal endoscopic laser-assisted surgery permitted full tumor removal with less than 100 mL of blood loss. Serial follow-up MRI scans and office endoscopy revealed no evidence of recurrence after more than 2 years.

COMMENT

In describing JNAs in 1845, Dieffenbach wrote,

Operation on the firm, tough and fibrous polyps requires the whole skill, sureness, and fearlessness of an experienced surgeon. The sufferings of the unhappy patients certainly stimulate the surgeon to brave attack, and indeed much courage is needed for there are almost only three things to choose between: suffocation of the patient when the surgeon is making the ligature, bleeding to death when the operation consists of excision and extraction, or leaving the operation uncompleted.

Even today, the experienced surgeon must be prepared to deal with many potential complications associated with current JNA open surgical approaches, including palatal dehiscence, velopharyngeal insufficiency, facial paresthesias, hemorrhage, altered facial growth, epiphora, need for perioperative tracheotomy and/or maxillomandibular fixation, unsightly scarring, loss of dentition, meningitis, hemiplegia, blindness, and even death.7-10 Possible radiotherapy complications include radiation-induced carcinoma, cataracts, facial growth impairment, hypopituitarism, epistaxis, xerostomia, caries, and loss of intelligence.11-13 Other forms of JNA therapy, such as hormone manipulation, chemotherapy, and cryosurgery, have had only very limited success.1

Endoscopic laser-assisted, image-guided surgical excision of JNAs offers excellent tumor exposure through a minimally invasive approach. Distinct advantages include (1) diminished blood loss; (2) superior cosmesis, without altered facial growth; (3) direct access of skull base, with minimal morbidity; and (4) ease of endoscopic follow-up. We will address each of these advantages.
How is hemostasis improved? Surface and interstitial Nd:YAG lasers are used to photocoagulate the tumor, followed by microdebrider excision of devitalized fibrous tissue. Surface Nd:YAG laser photocoagulation is performed by touching the fiber directly to the tumor. Interstitial Nd:YAG laser photocoagulation is performed by submerging the stripped fiber into the tumor and firing the laser until the tumor blanches.14 The heater probe photocoagulative properties of the Nd:YAG laser greatly enhance tumor hemostasis. The tumor interior is then gutted using the microdebrider in a similar conceptual fashion to acoustic neuroma removal. Since JNAs do not have significant smooth muscle lining the vascular clefts, vessel vasoconstriction is minimal. Should bleeding occur, direct endoscopic packing with pledgets works well until hemostasis is controlled with laser or suction electrocautery. Preoperative embolization of tumor feeding vessels further enhances hemostasis. Preoperative autologous blood banking is recommended in cases in which tumor excision is planned on a nonurgent basis (cases 1-3). Although cell savers may be considered useful in conjunction with highly planned excision, they are not necessary. Current studies suggest that autologous blood sparing in conjunction with highly vascular JNAs,15 we do not advocate their use because of the risk of autoinoculation of tumor from transfusion. The average estimated blood loss for patients undergoing JNA resection without preoperative embolization is more than 5000 mL.16 Adding preoperative embolization decreases the blood loss to 1000 mL.16 Adding intraoperative endoscopic laser and cautery techniques further decreases the blood loss to less than 500 mL. This diminished blood loss may obviate the need for transfusion.

How is cosmesis improved? Endoscopic laser-guided excision of JNAs requires no facial incisions. Unilateral rhinophyma scars are avoided. Rhinolalia aperta and epiphora are not potential problems. Potential nasal tip deformity after midface degloving is avoided. Osteotomies, plating, tracheotomy, and maxillomandibular fixation are unnecessary. Recent studies suggest that endoscopic sinus surgery does not affect long-term facial growth in children.17 Extrapalation of facial growth data to include endoscopic JNA excision will require further study. Endoscopic lateral tumor dissection may be facilitated with sublabial and buccal incisions (case 4). Postoperative pain is minimal. Children usually tolerate unilateral nasal sponges quite well for 2 to 4 days.

How is skull base access improved? Endoscopes provide highly illuminated, magnified, panoramic views of the skull base through a keyhole pathway. Posterolateral skull base structures such as the clivus, paratubal area, foramina ovale, rotundum and lacerum, sphenoid sinus, cavernous sinus, and orbital apex are readily accessible with the endoscope through the nose. Anterolateral structures, such as the lateral aspects of the pterygoid air space and infratemporal fossa, are endoscopically accessible through adjunct sublabial Caldwell-Luc and buccal incisions. Magnetic resonance imaging and CT image-guided computer navigational systems improve understanding of difficult skull base anatomy. Computed tomography image-guided surgery with the system that we used in the present study (InstaTrak) is simple to process, does not require fiducials, and is especially helpful when contrast-enhanced soft tissue scans are used to visualize tumor. Magnetic resonance imaging navigation gives optimal soft tissue distinction in cases in which the tumor abuts the dura or cavernous sinus (case 4).

Why is follow-up easier? Endoscopic follow-up is simplified because the surgical landmarks are readily identifiable in postoperative clinical evaluations. The postoperative endoscopic view is identical to the intraoperative view. Hospitalization is minimized. Nasal sponges are removed in the clinic soon after surgery. Nasal crusting is gently debrided, and sinonasal saline irrigations are encouraged for up to 1 month. Antibiotic therapy is discontinued when nasal crusting subsides. Endoscopically guided cultures may be performed if infection is suspected. Office endoscopy using topical anesthesia is well tolerated by children and allows the surgeon to closely inspect and serially examine any areas in which tumor recurrence is suspected. Postoperative endoscopic examination complements MRI scanning in cases in which there may be recurrence. Magnetic resonance imaging is preferred over CT for follow-up because it minimizes radiation exposure in children and provides enhanced postoperative soft tissue analysis to help rule out recurrence. If a small recurrence is noted on MRI scans or endoscopically, the asymptomatic mass may be endoscopically removed with relative ease.

Tumor recurrence rates are dependent on tumor stage and are reported in up to 55%18 of all cases. Juvenile nasopharyngeal angiofibromas are known to grow rapidly and involve submucosal spread. Tumor strongly adheres to soft tissue and bone. High recurrence rates are strongly correlated with size and skull base involvement. Recurrence rates as high as 86% have been reported in a series of large JNAs.19 Recurrences commonly occur between 6 months and 1 year after surgical therapy.20 Computed tomographic scans and intraoperative frozen sections revealed skull base invasion in all 5 of our patients. Two patients had unsuspected asymptomatic skull base recurrences that were treated with revision endoscopic excision directed to the clivus and pterygoid base. Residual disease does not necessarily require surgical excision in all cases. Juvenile nasopharyngeal angiofibromas are generally extradural21 and may regress spontaneously.22,23 They may be treated expectantly in the asymptomatic patient.9 However, the laser-assisted endoscopic approach provides a minimally invasive approach for excision of recurrent tumor and may avoid the morbidity associated with radiotherapy and open surgical procedures.

There are several “pearls” of experience that will help to enhance success with endoscopic laser-assisted image-guided removal of JNAs:

1. A team approach is recommended in cases of extensive JNAs. The otolaryngologist, interventional radiologist, frozen-section pathologist, neurosurgeon, and anesthesiologist work together to optimize surgical care. The endoscopist may remove extensive tumors but must always be prepared to change to more traditional open procedures should the situation warrant.

2. Preoperative radiographic evaluation is crucial. A CT scan delineates bony margins around the tumor and is routinely obtained for image-guided surgery. An MRI scan detects soft tissue interfaces and any intracranial extension. Image-guided surgery is commonly performed using CT data, but it may also be combined with MRI scans.
in extensive tumor cases. Successful embolization is meticu-

lously performed within 24 hours of surgery. The ex-

ternal carotid tumor branches are relatively easy to embo-
lize. Embolization of internal carotid branches in larger
tumors is associated with greater risk of stroke and should
be performed only by experienced interventional radiolo-
gists. The anesthesiologist should avoid intraoperative
hypertension and keep the systolic blood pressure level
under 110 mm Hg.

3. The Nd:YAG laser is used in surface and inter-

stitial applications until the tumor blanches and begins
to bleed. A laser (or cautery) and a microdebrider are al-
ternately used to remove the bulk of the intranasal and
nasopharyngeal tumor in a systematic fashion.

5. Image-guided assistance is helpful during re-

moval of tumor in the pterygomaxillary space and infra-
temporal fossa and against the skull base. Electromag-
netic image-guided surgery systems are preferred over
infrared systems, as line-of-sight issues are not problem-
atic. Image-guided surgery is never a substitute for know-

ing surgical anatomy; it is just another tool to enhance
surgical efficiency, safety, and full tumor removal.

6. A large middle meatal antrostomy is made to al-

low ready access to the posterior wall of the maxillary
sinus. The posterior wall is commonly thinned and an-
teriorly displaced as a result of the mass effect of the tu-
omor. Removal of the posterior wall of the maxillary si-
nus permits direct access to the lateral portion of the tumor
in the pterygomaxillary space and infra-temporal fossa.
The fingerlike projections of the lateral portion of the tu-
omur are usually not tethered and can be removed intact
by teasing the tumor medially and delivering the tumor
portion down the pharynx or transnasally (case 4).

7. After tumor removal, key endoscopic frozen-

section margins should be obtained. These margins
are commonly obtained at areas of fibrous tumor infiltr-

ation at the skull base. If the margins are positive, the fi-
brous or bony areas should be removed with cutting for-
ceps or a drill, and then endoscopic frozen sections should
be repeated until the margins are negative.

8. The endoscopic surgeon should be comfortable
with standard endoscopic sinus surgery and skull base


techniques. Increased experience with endoscopic skull
base surgery allows the surgeon to resect extensive tu-

mor with confidence. A backup surgical plan should be
considered if the surgeon becomes uncomfortable with
extended endoscopic removal. An inexperienced endo-

scopic surgeon should not perform this procedure.

9. Endoscopic and traditional techniques for JNA
removal should be considered complementary and not
exclusionary. The endoscope is an excellent adjuvant tool
and should be used in conjunction with traditional tech-

niques when warranted.

10. Close follow-up care is recommended during the

first year after surgery and should include serial office
endoscopic examinations and MRI scans. If a small skull

base recurrence is seen, early endoscopic removal is di-
rect and associated with low morbidity.

There are many advantages to endoscopic laser-assisted
image-guided removal of JNAs. As special endoscopic sur-

gical technology continues to improve and becomes readily

available, we will likely move away from classic procedures
in favor of minimally invasive techniques.

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