



Surgical planning in pediatric skull base surgery



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Treatment of pediatric skull base disorders is complex and requires individual planning. Compared to adult patients, the pathologies are more congenital and benign. Advanced surgical planning has been shown to improve outcomes and involves a variety of different software platforms to enable sophisticated review of image data sets, including virtual endoscopy and the details of surgical approach options. There are greater than 50 open and endoscopic surgical approaches described. Endoscopic approaches in patients under age 4 are extremely challenging due to the underdeveloped sinuses and overall smaller size of the craniofacial skeleton and nasal cavity. A multidisciplinary surgical team should develop and review surgical plan options with the patient and family.
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Introduction

Skull base surgery has unique challenges that require multidisciplinary input for optimal surgical planning and outcomes. The selection of the surgical approach is critical and it is a major determinant not only of surgical success, but also of the incurred morbidity secondary to collateral tissue damage. The term minimally invasive is often used, but a more appropriate term is minimally disruptive. The degree of “invasivity” is determined by the type of lesion being treated. Whether an open or endo-

scopic approach is chosen, the surgical task (ie complete excision or incisional biopsy) needs to be completed. The goal is to carry out the surgery through an approach that is carefully selected to minimize damage to adjacent tissue and minimize morbidity and risks to the patient. As such, surgical planning is performed in order to maximize access to the lesion for desired surgical effect while minimizing collateral tissue injury. This is all conducted within small spaces bounded by critical structures such as the internal carotid artery, optic nerve, and cavernous sinus. The challenges seen in treatment of adult skull base pathology are exacerbated in the pediatric population, further supporting the need for advanced planning.¹ Any lifelong morbidity incurred in the pediatric patient has additional challenges through adolescence.

The pathology seen in pediatric population favors more congenital and benign pathology compared to adult pa-

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Table 1 List of pediatric skull base pathology.

Juvenile nasopharyngeal angiofibroma (JNA)
CSF leak, meningoencephalocele
Pituitary adenoma
Osteoma
Inverted papilloma
Fibrous dysplasia
Craniopharyngioma
Neuroblastoma
Rhabdomyosarcoma
Chondrosarcoma

tients. A list of the common diagnoses seen in pediatric patients is listed in [Table 1](#).

A multidisciplinary surgical team should develop and review the surgical plan, ideally within a model that incorporates virtual endoscopy and specifics of the surgical pathway.² Computer models for skull base surgery have been developed, but they have barriers to use (eg, transferring DICOM data to third party software) if not integrated with the electronic medical system or intraoperative navigation system.³ When virtual surgical planning is utilized, there is evidence for improved functional outcomes of the orbit and skull base.⁴

Endoscopic skull base surgical approaches in patients under age 4 are extremely challenging due to the underdeveloped sinuses and overall smaller size of the craniofacial skeleton and nasal cavity. Precise and accurate surgical planning is especially important in this population.

Computer models and 3-dimensional (3D) printing for development of surgical pathway

Image sets have been the foundation to surgical planning for decades. There have been technologic advances in acquisition of computed tomography and magnetic resonance imaging data, and in the postprocessing of this data. Many surgical navigation systems now include software to develop a surgical plan. The features vary from segmenting structures and lesions to virtual endoscopy with the ability to make multiple measurements and volumetric calculations. Open source DICOM viewers such as Slicer and OsiriX enable researchers to make advances without building a new platform for each application.⁵

There are several ways to use a virtual model once it is created. Often, a significantly improved understanding of the surgical approach may be obtained from viewing the model virtually. 3D printing has become a popular way to produce a physical object to permit dry lab testing and to facilitate communication between surgeons, as well as with the patient and family.

This is demonstrated in the complex geometry of the optimal surgical pathway to the lateral cavernous sinus in this example case (*See Figure 1*).⁶ It is designed to optimize access to the target location, while minimizing orbital retraction and the size of the craniotomy in the greater wing of the sphenoid. In [Figure 1](#) below, note that the shape of the pathway in (B) is reduced in volume significantly as the final shape (C) is designed through an iterative process. This takes advantage of the endoscopic

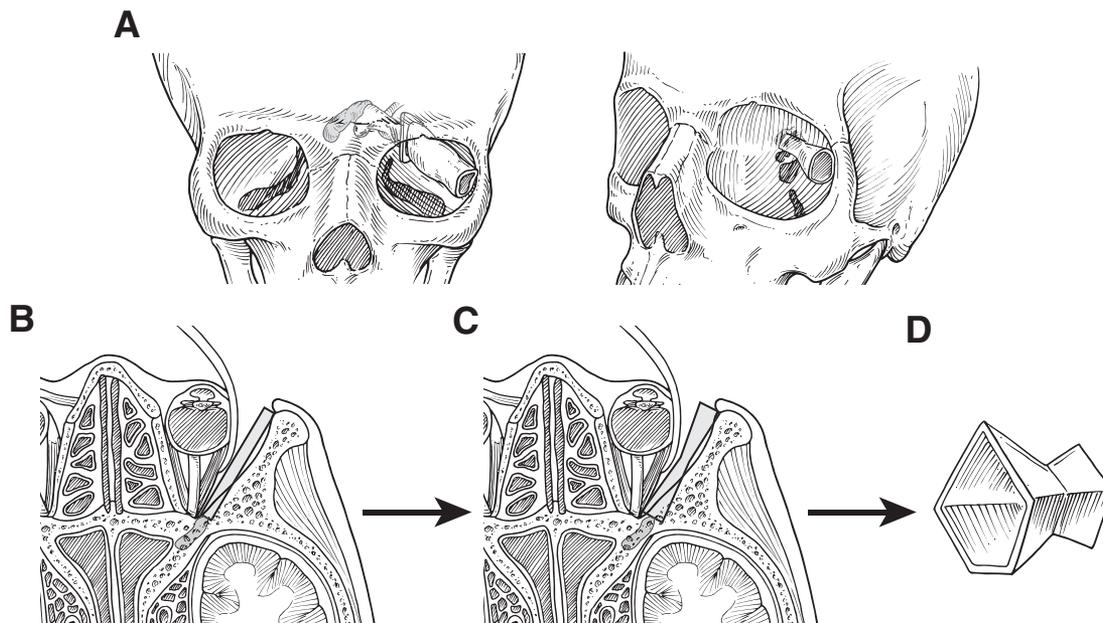


Figure 1 The dimensions of the surgical pathway are carefully designed. The iterative process begins with a lateral transorbital corridor approaching the target location to the lateral cavernous sinus with a craniotomy through the greater wing of the sphenoid (A, B) It is refined, and the volume is reduced creating the biconical shape, which maintains instrument range of motion (C) The entrance to the surgical pathway is bounded by the orbital rim and the retracted orbital contents. As the pathway dimensions are refined, note that the size of the craniotomy in the greater wing of the sphenoid is reduced without compromising surgical access. A physical model can be produced (D) to perform dry lab testing and to aid in communication between surgeons and patients and families.

instrument range of motion in a biconical shape. The ability to perform surgical tasks at the target for both (B) and (C) is nearly identical; the same operation can be performed yet the amount of tissue disruption is significantly reduced in the smaller volume (C) corridor. Further, if indicated, a physical model can be created to perform dry lab testing (D).

Approach selection and pathway design

There are greater than 50 open and endoscopic surgical approaches to the skull base described in the literature.⁷ For endoscopic surgical approaches, each is described typically by the entrance portal, whether that be the nose (transnasal), the anterior maxillary wall (transmaxillary), or through the orbit (transorbital). In order to optimize the surgical plan, it is best to specifically design the dimensions of the corridor through which the endoscopic surgical intervention occurs. By the nature of endoscopes and endoscopic instrumentation, this is a biconical shape in many instances, as described in Figure 1. Endoscopic approaches are often augmented using multiple portals in order to obtain the best visualization while permitting instruments to approach from a different angle.^{8,9}

Virtual endoscopy and other methods of visualization can be helpful to determine how many surgical pathways are required based on individual anatomy. The goals of the planning process are to determine which surgical pathways are required to perform the operation with the least morbidity. In many cases, the favorable approaches are those that do not involve retraction of the brain. An “open” approach such as a keyhole supraorbital craniotomy can be performed with the assistance of an endoscope to augment visualization. As such, the distinction between the traditionally “open” surgical approaches and “endoscopic” is becoming less important, and the emphasis is shifting to the ability to perform the necessary surgical task while minimizing risk and morbidity to the patient.

Two example cases will be reviewed below to illustrate the surgical planning process.

Case example #1

A 13-year-old male presented with persistent nasal congestion, epistaxis, and visual changes of recent onset. Computed tomography and magnetic resonance imaging demonstrated involvement of lateral cavernous sinus, middle cranial fossa of a vascular and infratemporal fossa mass consistent with juvenile nasopharyngeal angiofibroma (JNA), classified as Radowski stage IIIb (See Figure 2) and UPMC Stage V.¹⁰

Endoscopic evaluation revealed a vascular mass filling the nasal cavity and displacing the septum to the contralateral side (See Figure 3).

Navigation software (iNtellect, Stryker Corporation, Kalamazoo, MI) was used to segment the tumor for 3D visualization, including adjacent anatomy. Surgical vector

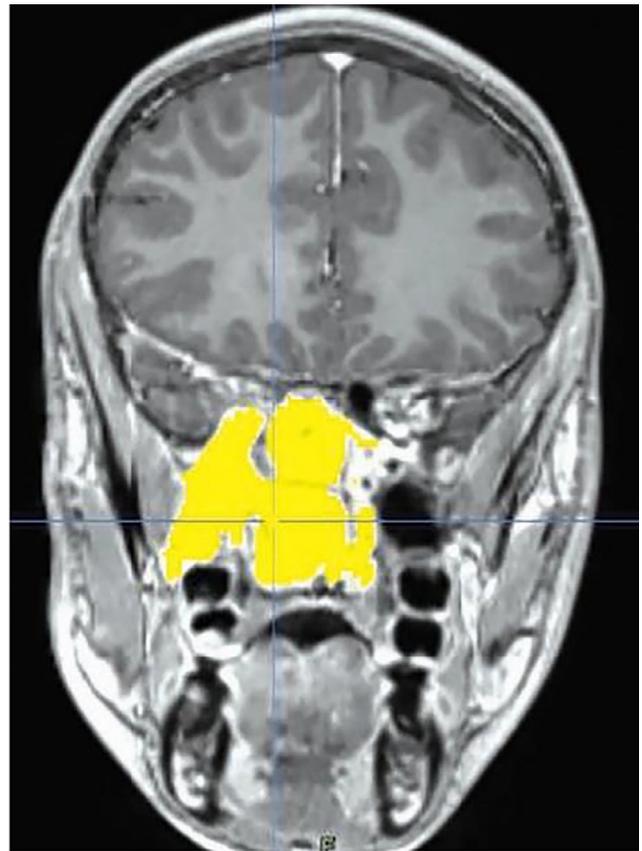


Figure 2 MRI visualization of middle cranial involvement of juvenile nasopharyngeal angiofibroma (JNA). Tumor segmented in yellow. (Color version of figure is available online.)

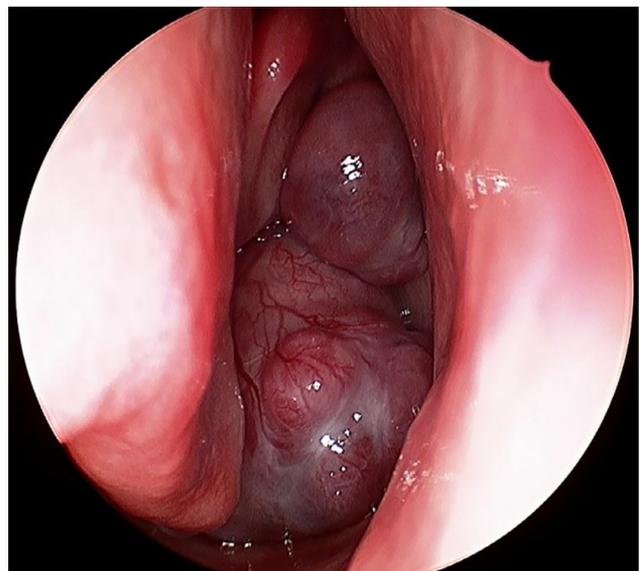


Figure 3 Endoscopic view of JNA.

analysis was then performed for transnasal, medial transorbital, inferior transorbital, and transmaxillary and/or infratemporal fossa entry portals (See Figure 4). The goals in this analysis were to determine pathway adequacy for instrumentation, angle of approach to target, risk of

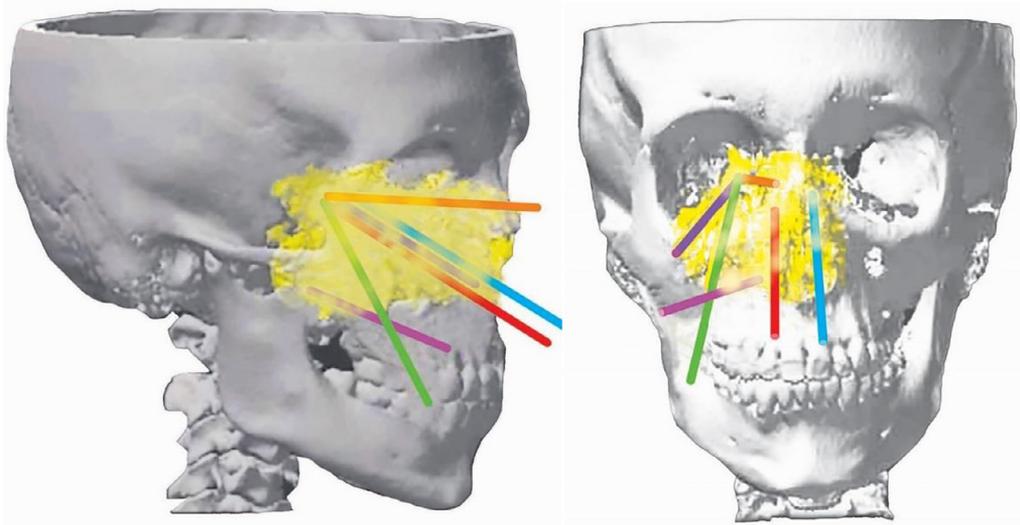


Figure 4 3D visualization of CT/MRI fusion, tumor segmentation (yellow) and potential pathways. Transnasal (blue and red), transmaxillary (pink and green), medial transorbital (orange), and inferior transorbital (purple). Left demonstrates lateral view and right demonstrates straight view. (Color version of figure is available online.)

Table 2 Measurements of potential portals and relative angles to the right transnasal portal.

Surgical approach corridor	Distance to middle cranial fossa	Angle relative to right transnasal portal
Transmaxillary /infratemporal	53 mm	22
Transnasal	50 mm	n/a
Medial transorbital	43 mm	32
Inferior transorbital	34 mm	34

instrument-visualization and instrument-instrument interference, and the risk of collateral damage to critical neurovascular structures. Target points were defined at the far lateral and superior aspects of the tumor. These specific targets were used to evaluate access for vascular control. Multiportal combinations were evaluated by analyzing angles and distances calculated from (x,y,z) coordinates. Angulation to permit adequate freedom to maneuver with 2 instruments working at the target location should be greater than 15°. ¹¹

Given the extent of the tumor, monoportal resection through a transnasal endoscopic approach alone was not feasible due to the inability to visualize and perform controlled dissection at the infratemporal fossa. Distance and instrument angulation was favorable for transmaxillary, infraorbital, and medial orbit (See Table 2). The transmaxillary portal allowed access to the most superior, inferior, and lateral aspects of the tumor. An additional transorbital portal was held in reserve depending on the adherence of the tumor to orbital tissue, along with a possible pterional

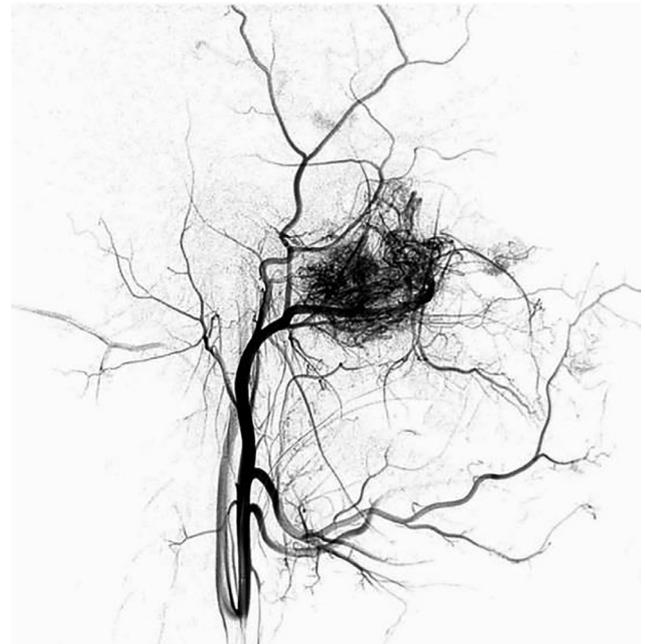


Figure 5 Preoperative angiography of JNA.

craniotomy if necessary to resect the intracranial component.

The patient underwent preoperative angiogram (See Figure 5) and embolization immediately followed by endoscopic triportal (bilateral transnasal and transmaxillary) resection of the tumor proceeded as planned without need for supplemental transorbital approach or pterional craniotomy.

After removal of the posterior maxillary wall, the portion of the tumor compressing the optic nerve was removed by following the tumor superiorly to the inferior orbital fissure and retraction of the mass inferiorly where it was

excised. The remaining portion of the tumor occupying the middle cranial fossa and cavernous sinus was resected using a 4-handed technique in which 3 instruments worked in concert under endoscopic visualization (See Figure 6). This technique at the most critical portion of the case underscores the importance of the multiportal approach where multiple instruments can work and not be subject to collision with the endoscope itself, which is entering from a different angle.

All gross disease was removed. The skull base defect was reconstructed using allogenic dermal graft (AlloDerm, LifeCell Corporation, Woodlands, TX). Total blood loss was 750mL with procedure time of 4.5 hours. The patient did have temporary square alopecia, a known complication of fluoroscopic embolization.¹² The patient remains without evidence of recurrence or persistence of juvenile nasopharyngeal angiofibroma on imaging 5 years after resection.

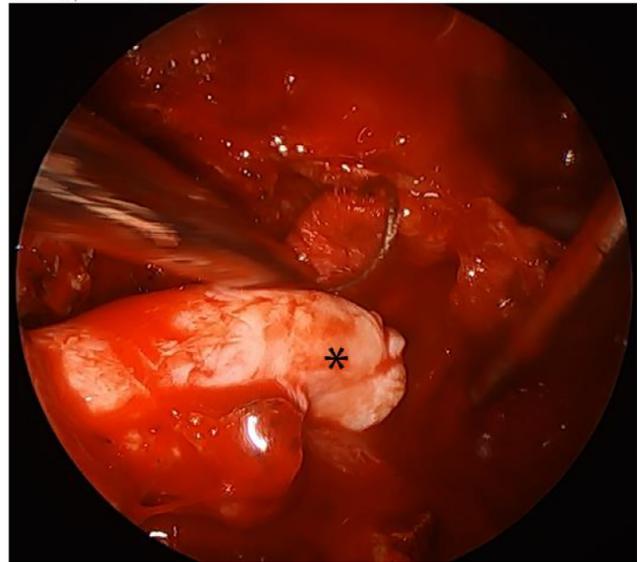


Figure 6 Tumor removal from the middle cranial fossa with pituitary ring curette. Asterisks denotes tumor.

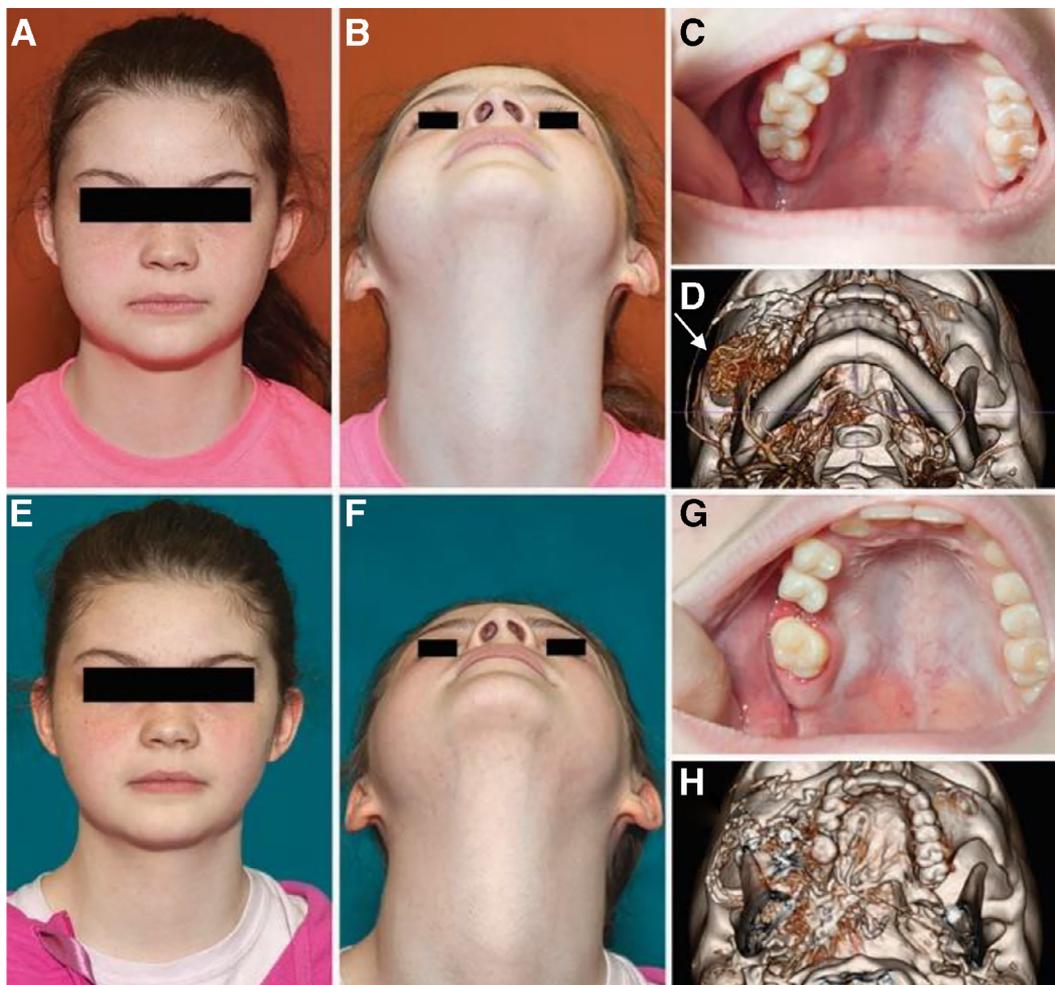


Figure 7 Clinical photographs of patient with AVM involving right maxilla and infratemporal fossa. The preoperative images (A-D) demonstrating the right sided facial swelling. The white arrow in (D) indicates the 3D reconstructed AVM mass. Postoperative images (E-H) show complete excision.

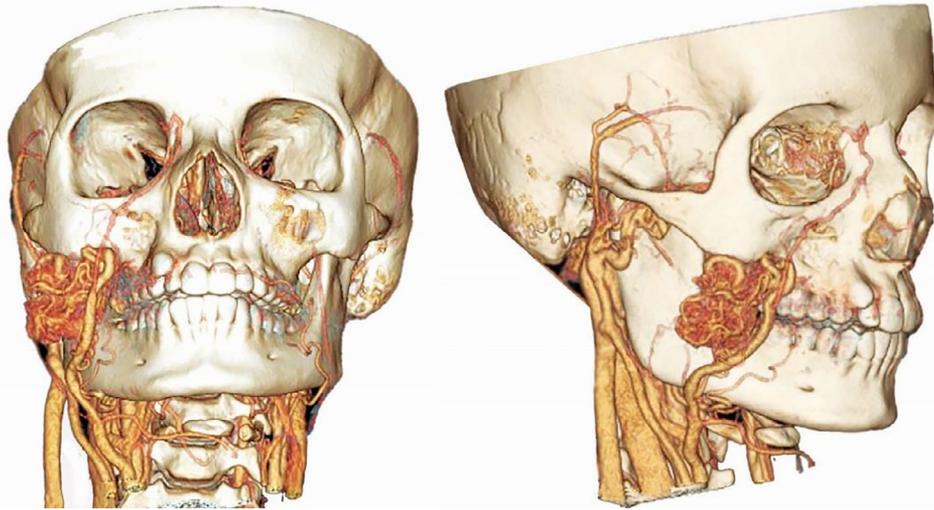


Figure 8 3D reconstruction CT angiogram of right-sided infratemporal and maxillary AVM.

Case #2

In a separate case involving an extensive arteriovenous malformation of the right maxilla into the infratemporal fossa and skull base, a similar planning technique was utilized. The patient presented with bleeding around tooth number 3 (See Figure 7).

After a series of embolizations and procedures to address the portion involving the anterior maxilla, the patient continued to have symptoms of pain, bleeding, and pressure on the right side. A triportal endoscopic approach using bilateral transnasal and ipsilateral transmaxillary endoscopic corridors was designed. This permitted adequate access to the portion of the arteriovenous malformation located at the skull base along the infratemporal fossa (See Figure 8). Excision after embolization was performed using similar endoscopic techniques as described in above case. Postoperative imaging for 2 years has demonstrated no recurrence.

Future work

The modeling and multidisciplinary preoperative planning provides the best surgical treatment while minimizing morbidity. Virtual surgical planning using available imaging data enables development of new surgical approaches in 3 dimensions, and refinement of known endoscopic pathways. As preoperative modeling software becomes more sophisticated, it is likely that an algorithm-based solution will be available to help guide the surgeon.¹³

A major goal moving forward will be to study patient outcomes based on the details of the surgical approach and surgical pathways, which will be defined by more than simply the name of the approach (ie, transnasal, transorbital, and transmaxillary). Instead, the approach will be defined by the specific geometry of the surgical corridor

utilized. There should be quantitative metrics to define the 3D shape, the volume, and the adjacent anatomy. The planned pathway dimensions determined preoperatively should be compared against the actual surgical pathway dimensions, as determined based on instrument tracking during the surgery. This knowledge will enable future study for iterative improvement as well as the design of novel surgical robotic systems, which rely upon an accurate model of instruments moving through a defined shape. Further, it will enable more robust outcomes studies once the surgical approaches are quantifiable.

Disclosures

The authors reported no proprietary or commercial interest in any product mentioned or concept discussed in this article.

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