10.5005/jp-journals-10003-1099
ORIGINAL ARTICLE

Tailoring the Optimal Surgical Strategy for the Endoscopic Removal of Juvenile Nasopharyngeal Angiofibromas

Vicky S Khattar, Bachi T Hathiram

ABSTRACT

Being centered around the sphenopalatine foramen, juvenile nasopharyngeal angiofibromas are notorious for growing toward the infratemporal fossa via the pterygomaxillary fissure. Since, this tumor grows from 'within' the sinonasal space, it would be best to surgically follow it from the same path that it took to grow. With the development of endoscopic techniques and instrumentation, endoscopic approaches to the infratemporal fossa have been universally accepted. Nevertheless, since the tumor does not follow a definitive pattern, each surgery needs to be tailored. In the authors' experience, a preoperative radiological assessment can prove to be a relatively valuable guide to estimate and plan the surgical trajectory that need be taken for this particular tumor.

Keywords: Nasopharyngeal angiofibromas, Infratemporal fossa, Surgical approach, Radiological appearance.

How to cite this article: Khattar VS, Hathiram BT. Tailoring the Optimal Surgical Strategy for the Endoscopic Removal of Juvenile Nasopharyngeal Angiofibromas. Int J Otorhinolaryngol Clin 2012;4(3):136-143.

Source of support: Nil

Conflict of interest: None declared

INTRODUCTION

Juvenile nasopharyngeal angiofibromas are slow-growing, histologically benign yet locally aggressive, nonencapsulated vascular hamartomas, commonly occurring in adolescent males. They usually spread in an ameboid fashion, with 'pseudopods' that can even at times be found in bone.

The tumor is pseudoencapsulated. As it grows (and especially for virgin/unoperated tumors) the tumor follows the path of least resistance, expanding into cavities, foramina, canals, and widening them as it progresses. It eventually remodels, attenuates and finally may even erode bone. It may be understood that since the tumor grows from within the nose it should be removed also from within. This forms the rationale for the endoscopic management of this tumor.[1]

They are postulated to arise posterosuperior to the sphenopalatine foramen in the region of the pterygoid canal (Hackman et al 2009).[2] From the pterygopalatine fossa, these tumors usually follow a path of least resistance whether medially or laterally; the former would be toward the nasal cavity and nasopharynx, while the latter toward the infratemporal fossa. The tumor may sometimes also grow superiorly toward the inferior orbital fissure, and onwards through the orbit and intracranially.

Surgical access to the pterygopalatine and infratemporal fossae has been a topic of much discussion, with various approaches having been proposed as well as practiced. Presented here is a review of the various approaches employed, with their trajectories discussed.

SURGICAL ACCESS TO THE INFRATEMPORAL FOSSA

The infratemporal fossa is situated precariously between important anatomical structures at the base of the skull making it difficult to access. However, there have been various approaches described in literature, most of them being open approaches. The endonasal endoscopic approach is most definitely the preferred one but, one has to keep in mind that this approach is not practical in each and every case.

In the authors' experience, among the external approaches, the maxillary swing/osteoplastic maxillotomy has proven to be the most versatile in reaching the deepest recesses of this space with reasonably good preservation of function and cosmesis. Although no endoscopic approach can match the wide end-on exposure of the infratemporal fossa achieved by the osteoplastic maxillectomy approach, some of the endoscopic approaches described in this article are most definitely capable of giving comparable results when judiciously used. Endoscopy of course carries with it the great advantage of excellent cosmetic results in addition to reduced morbidity.

The endoscopic approach should be tailored to suit the pathology. It may range from the minimalistic to the radical; wherein removal of the medial and posterior walls of the maxilla forms an integral part of all the procedures mentioned below and the choice of approach is further dictated by the trajectory and angulation required to be used to gain complete access.

Prior to discussing the various approaches that may be employed, it is prudent to admit, that the basic principles of this tumor excision should not be compromised at any stage:

a. Complete exposure of the tumor and its surroundings prior to excision
b. Subperiosteal dissection
c. Drilling away of any bony attachments/potential sites of bony penetration by tumor such as basisphenoid

d. En-bloc removal as far as possible
e. The protrusion of the infratemporal fat into the surgical field to be taken as the end point of surgery.[3]

With the exception of '(d)' (above), which may be possible in certain cases, almost all of these tumors respond well to endoscopic excision if the above are followed.

Depending upon the status of the preoperative radiological assessment, any/a combination of the below may be considered:

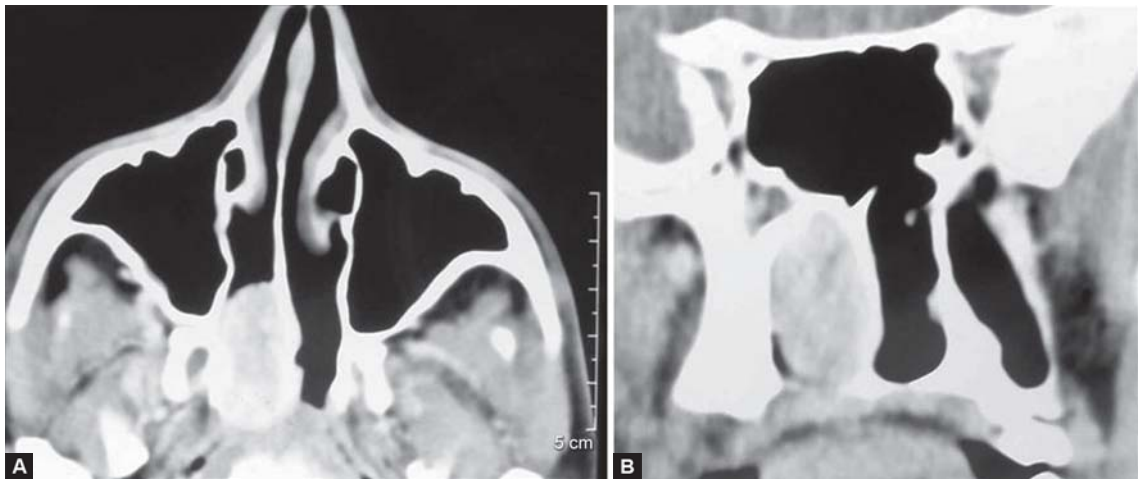
1. A Sphenoethmoidectomy and a Wide Maxillotomy

This is an approach that can be used for medially placed tumor; the ethmoidectomy is simply performed to release the middle turbinate from its lateral bony septae, following which it can be partially amputated. The ethmoidectomy also ensures that postoperatively, a loss of the middle turbinate as a landmark will not interfere with subsequent sinus problems (if they do occur at all). The maxillotomy will ensure an exposure of the posterior maxillary wall that may then have to be removed (at least partially) even in the most medially placed tumor (*vide infra*) (Figs 1A and B).

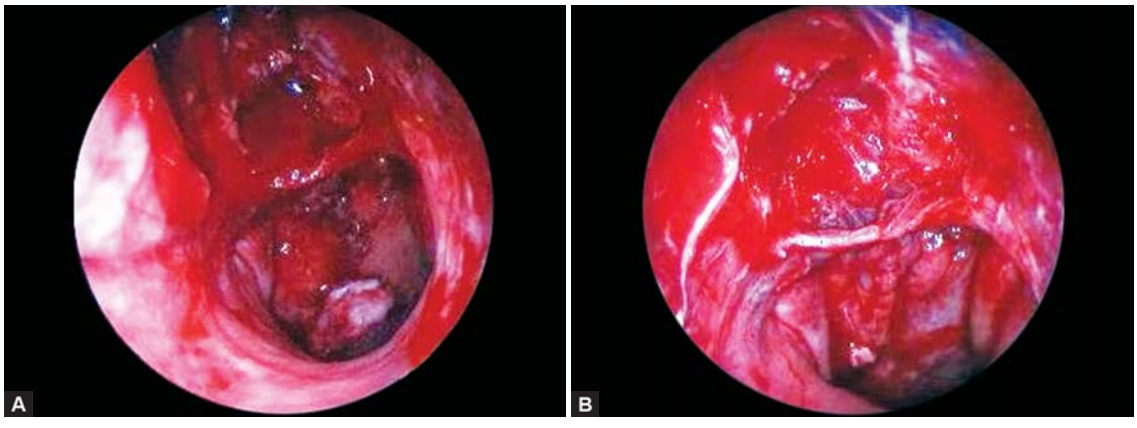
Certain Points to be Considered

Surgical trajectory: Lack of a definitive lateral extension of the tumor precludes much lateral dissection. A relatively straight trajectory may be used. Removal of a part of the middle turbinate may be considered to gain access (Figs 2A to 3).

Teaching point: On reassessing the preoperative computed tomographic (CT) scan of the patient, one can notice a subtle widening of the pterygopalatine fossa on the right side, which was evident on serial cuts. In an overzealous attempt to remove a 'relatively small' tumor, one must not forget that achieving a circumferential access to the tumor periphery is a must before attempting removal. Relatively straight surgical trajectories must not trick the surgeon to forget drilling adjacent bone (Fig. 4).



Figs 1A and B: Contrast-enhanced CT scan showing an angiofibromas in the right nasal cavity, occupying the choana and nasopharynx



Figs 2A and B: Intraoperative pictures of the same tumor during and after removal showing preservation of the inferior turbinate. The lower end of the middle and superior turbinates required a little trimming to gain access around this tumor

Fig. 3: Three-month postoperative endoscopic view of the patient showing the beginnings of a recurrence near the region of the sphenopalatine foramen. This was handled successfully with a bipolar cautery and drilling of the adjacent bone

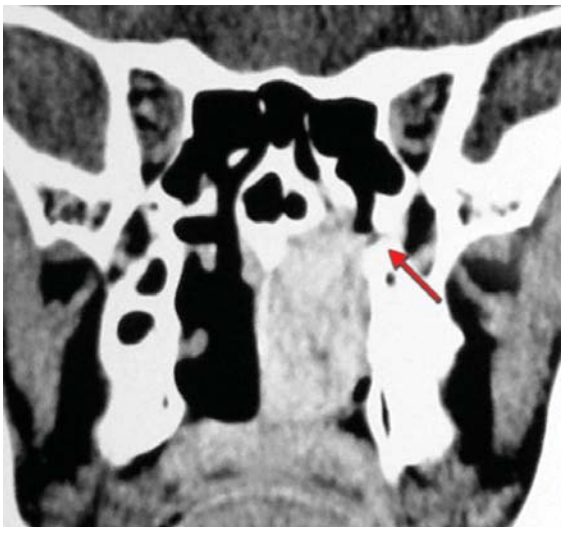


Fig. 4: Another case of a left-sided JNA seen on a contrast-enhanced coronal CT scan showing the minimal lateral extent toward the pterygopalatine fossa that needs to be dealt with

Thus, even seemingly medially placed tumors need lateral exposure.

2. A Partial Inferior Turbinectomy with a Sphenoethmoidectomy and a Wide Maxillotomy

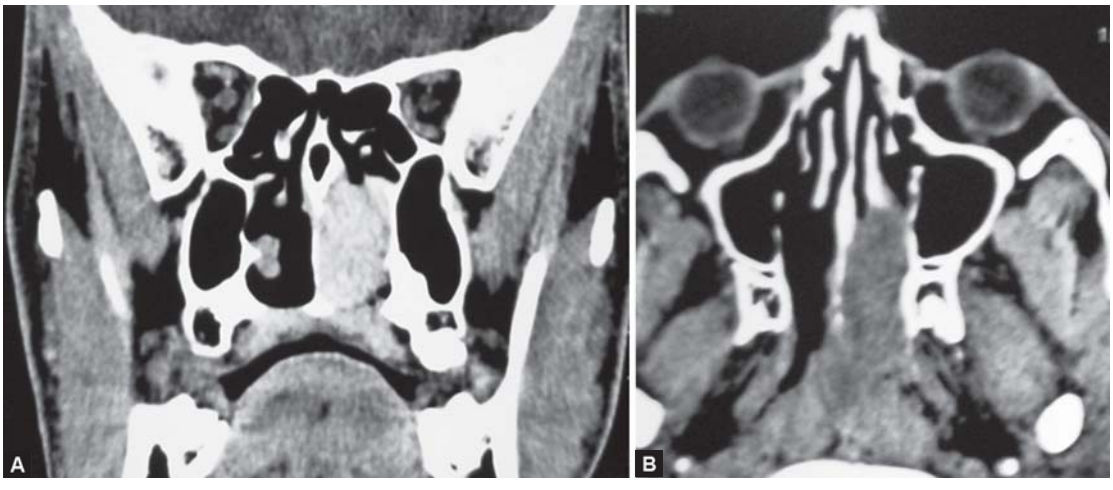
Tumors similar to those seen above, can be tackled by this technique, provided the tumor has attachments to the inferior turbinate, or has extended anteriorly into the nasal fossa, between the nasal septum and inferior turbinate (Figs 5A and B).

3. A Complete Inferior Turbinectomy with a Sphenoethmoidectomy and a Wide Maxillotomy

Sometimes, this anterior extension can reach right up to the anterior nares, thus necessitating the removal of the entire inferior turbinate for gaining access (Fig. 6).

This is the pre-embolization image. Considering some amount of shrinkage after the embolization, the tumor still remains difficult to resect due to sheer lack of working space. In such cases, removal of the inferior turbinate gives just the amount of a 'head start' to begin the exposure. Being essentially confined to the nasal cavity without much lateral extension, the rest of the removal is relatively straight forward.

Teaching point: A good option used here was to release the nasopharyngeal adhesions first. Usually this step is performed toward the end of the surgery, prior to releasing the tumor into the oropharynx and delivering it transorally. The reason for the latter is that these adhesions tend to bleed while being released, and this may obscure the surgical field during an endoscopic procedure. In this case, however, the sheer lack of space will prevent any blood from trickling anteriorly; the additional benefit of releasing the nasopharyngeal adhesions earlier during surgery



Figs 5A and B: A left-sided JNA encroaching upon the nasal fossa anteriorly and involving the posterior part of the left inferior turbinate. Removal of the posterior part of the left inferior turbinate would ensure the availability of working space, and also ensure that adequate tissue has been removed around the tumor

would be the option of giving traction with the assistant holding the tumor from below (perorally) and applying inferior traction. This, coupled with removal of the inferior turbinate (*vide supra*) would give just the required amount of space to start the endoscopic work in the anterior part of the nasal fossa (Fig. 7).

4. A Partial Medial Maxillectomy without Drilling the Frontal Process of the Maxilla

This is by far the most commonly used approach (in the authors' experience) for removal of tumors that have grown toward the infratemporal fossa laterally. The frontal process of the maxilla can be saved in such cases only, if the tumor has not crossed the anterior wall of the maxilla (in the contrast-enhanced axial CT scans), or the anterior most part of the middle turbinate (as seen during the endoscopic evaluation). This entails performing a complete spheno-ethmoidectomy, a partial middle turbinectomy, an inferior turbinectomy, and a wide medial maxillotomy (Fig. 8).

Fig. 6: A large JNA in the left nasal fossa occupying practically all of the available space

Fig. 7: *En bloc* removal of the tumor in Figure 6

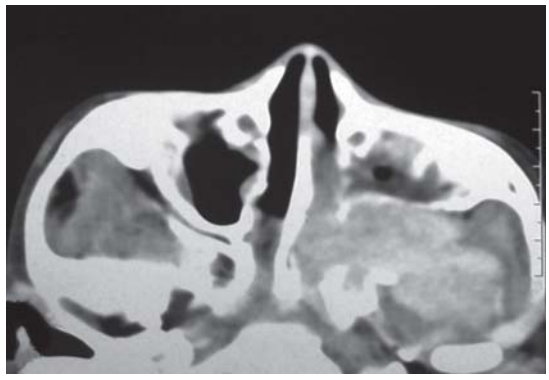


Fig. 8: A contrast-enhanced axial CT scan of a patient with a left-sided JNA with extensive lateral spread into the infratemporal fossa. The patient underwent the endoscopic procedure and a complete *en bloc* removal was possible

Teaching point: Given the benign nature of this tumor, parts of the inferior turbinate and medial maxillary wall that are not in contact with the tumor may be preserved, depending upon the case. It must however, be kept in mind that often the removal of the latter two are more for space to allow instrumentation, rather than exposing the tumor (Figs 9 and 10).

It may be argued here that with the use of angled endoscopes and instruments, it may be possible to still preserve the inferior part of the maxillary wall, and successfully remove the tumor. That may be true, but the lack of a significant postoperative benefit, coupled with the sheer ease of removal has made removal of the bone in question the method of choice for the authors; added to this the luxury of using straight, nonangled telescopes with an increased exposure, has decreased the operative time significantly in the authors' experience (Fig. 11).

This is the view with a 0-degree rigid telescope. Although the exposure for removal of this tumor is adequate with the 0-degree endoscope, the use of the angled telescopes after the procedure would allow for adequate inspection, as well as postoperative cleaning of crusts (Fig. 12).

5. A Partial Medial Maxillectomy with Drilling the Frontal Process of the Maxilla and Performing an Elective Dacryocystorhinostomy

This can prove necessary at times when the tumor has lateral as well as anterior extensions, significant enough to preclude a successful removal without removal of the frontal process of the maxilla. The removal of the frontonasal process of the maxilla would range from partial to complete (in width) and usually be partial in vertical height, as the authors have not come across a tumor in their experience that necessitated removal of the entire frontonasal process of the maxilla up to its superior articulation. Hypothetically, this would be

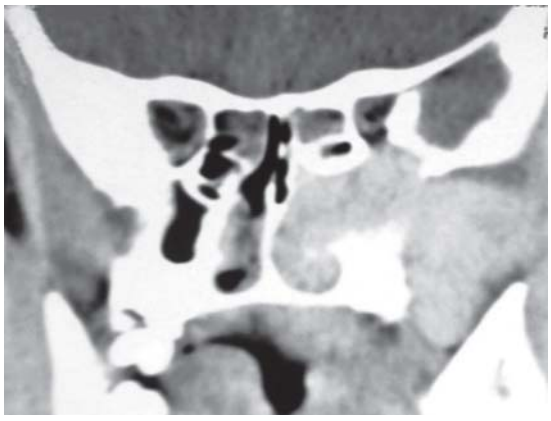


Fig. 9: The contrast-enhanced coronal CT scan of the same patient as in figure 8 showing the lateral extent of the tumor. It seems logical from this image, that preservation of the inferior part of the medial maxillary wall would add no benefit in the postoperative period, but would certainly impede the exposure and working space if this entire lateral extent of this tumor has to be dealt with

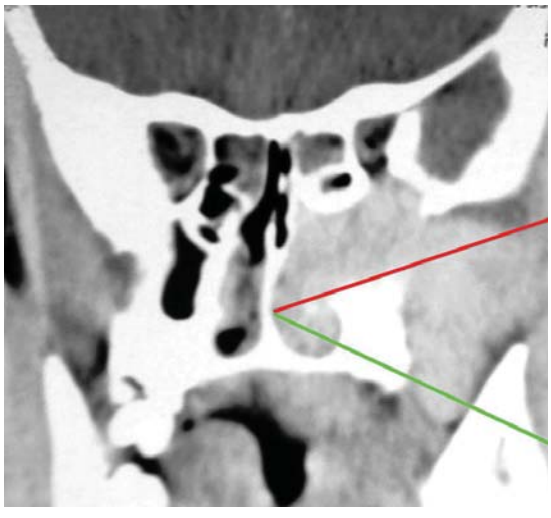


Fig. 10: The difficult trajectory used here (red line) without removal of the inferior part of the medial maxillary wall, and the relatively easier trajectory (green line) that may be possible with removal of the latter

Fig. 11: Endoscopic intraoperative image of the same patient (in Figs 9 and 10) showing a majority of the lateral extent of the tumor being seen after sheer removal of the medial maxillary wall

Fig. 12: The tumor (Figs 9 to 11) removed *en masse*

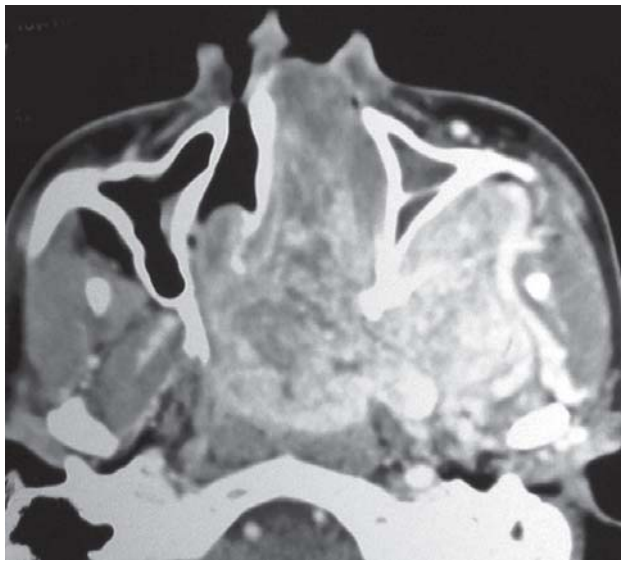


Fig. 13: A large tumor seen on the contrast-enhanced axial CT scan occupying the entire left nasal cavity, with extensive infratemporal fossa extension, and reaching the anterior nares and also crossing over to the other side

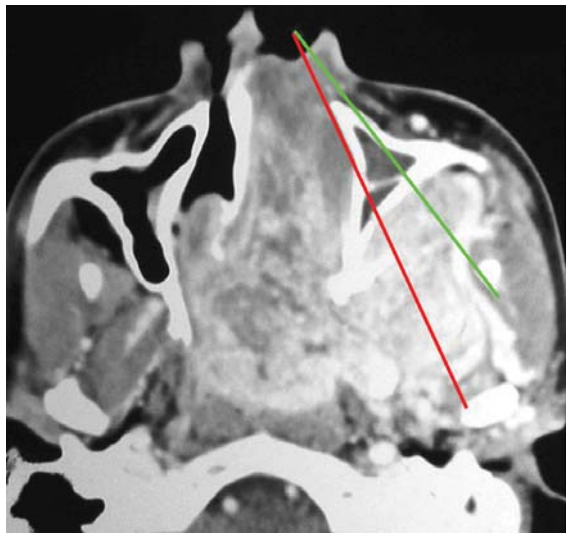


Fig. 14: The surgical trajectory without removal of the frontonasal process of the maxilla (red line) and after removal (green line) of the same. The added benefit for instrumentation is obvious

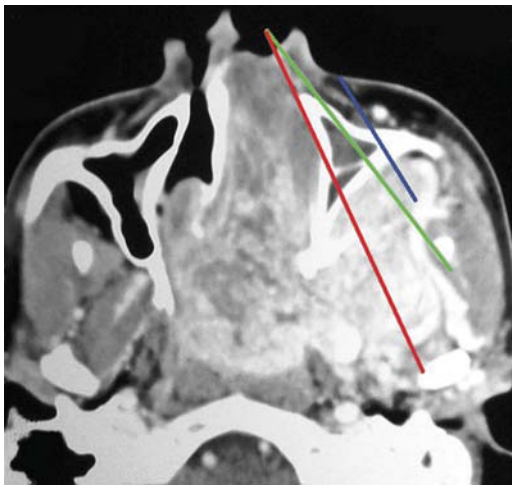


Fig. 15: The benefit obtained from this additional exposure in the blue line (*vide supra*), though minimal for the higher part of the maxillary antrum which is relatively narrower than the floor also allows for a shorter 'effort arm' for instrumentation. This decreases the effort required by the operating surgeon to manipulate the tumor, adding to the overall ease of removal and benefit of this trajectory

the natural approach used should the need arise. Only in cases where the lacrimal system has not been exposed after drilling would it be possible to get away without doing an elective dacryocystorhinostomy; in most cases, however, this would be required.

When compared with the erstwhile external approaches, this trajectory (as above) achieves a similar outcome as in Denker's approach (Figs 13 and 14).[4]

6. The above in Step 5 combined with a Canine Puncture

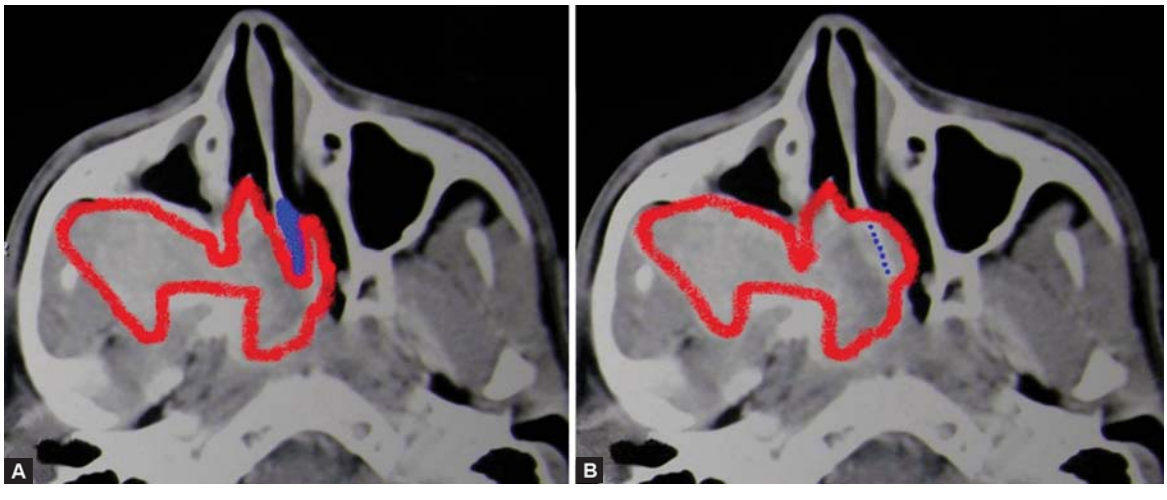
As above, the approach allows for optimum visualization and instrumentation for the most part. However, as can be seen in Figure 14, even the improved trajectory after removal of the frontonasal process of the maxilla can be occasionally suboptimal. This may be further improved (if required) when coupled with a separate port for access and visualization through a separate canine puncture, and traversing transantrally. This would provide for the maximal possible exposure possible with an endoscope.

This is similar to Sciarretta et al (2006),[5] who felt that tumors having a lateral extension toward the infratemporal fossa could be tackled by either an endoscopic radical medial maxillectomy, or a combined/transantral endoscopic approach, as these will help in achieving a better lateroposterior visualization of the lesion. In the authors' experience, this has proven to be an approach with very wide applications, provided the approach described by Sciarretta et al (2006)[5] entails a separate puncture through the canine fossa. More than the added exposure, however, it also allows for a decreased 'effort arm', thereby greatly contributing to the surgeon's comfort with lesser fatigue. Also, the authors feel that occasionally even malleable (but sturdy!) instruments can be used here, since the length of the instruments required is lesser (*vide supra* – shorter effort arm), and their tendency to bend/buckle while dissecting is less (Fig. 15).

7. A Trans-septal Single-surgeon Technique

Often these tumors grow through the choana to the opposite side of the nose, and abut against the posterior edge of the nasal septum. As a regular part of their removal, the endoscopic surgeon is often compelled to remove a part of the posterior bony septum to release any adhesions of 'bottlenecks' of the tumor and deliver it *en masse* (Figs 16A and B).

As a continuation of the same, it could be considered an option to approach the tumor from the opposite nasal cavity (either the endoscope or the working instrument could be inserted into the opposite nasal cavity, and a 'contralateral trans-septal' trajectory could be established). While this would allow for access to the ipsilateral infratemporal fossa



Figs 16A and B: The necessity of removing the posterior bony septum (blue) in cases of tumors (red) crossing over to the opposite side via the choana

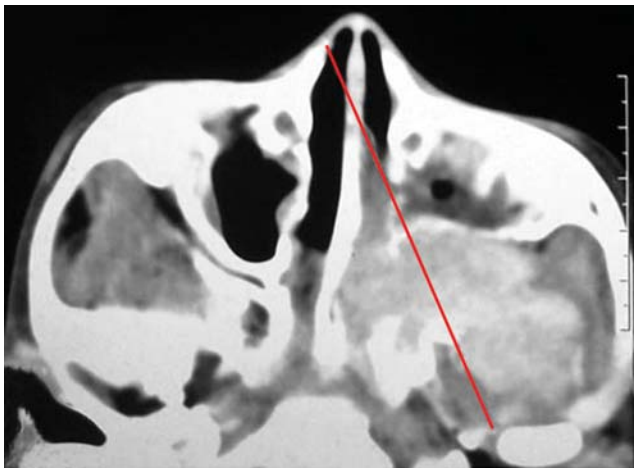


Fig. 17: The trajectory of a trans-septal approach (red). The authors consider it to be more of a 'consequence' of a mandatory posterior septectomy rather than a separate approach itself

to an extent, it would also increase the 'effort arm'. It remains best left to the surgeon to ascertain their comfort zone while employing such a technique.

Harvey et al (2009)[6] in a comparative study of 10 cadaver heads compared the ipsilateral and trans-septal access to the infratemporal fossa, and found a statistically improved surgical access, when the latter was used. They opined that the extended endoscopic maxillary surgery combined with a trans-septal option enables additional access to previously considered challenging locations. In the authors' experience, the removal of the posterior part of the septum and the subsequent improvement in access to tumors in the infratemporal fossa, is firstly more of a consequence while removing large tumors grown contralaterally due to their adhesions with the posterior bony septum, and secondly do

No.	*Surgical trajectory employed*	*Number of patients*	*Complications (recurrence residual tumor, etc.)*	*Comments*
1	A sphenoethmoidectomy and a wide maxillotomy	7	One recurrence (shown in Figs 3 and 4) occurred in the 3rd case (out of 7 mentioned here)	After the recurrence shown earlier occurred, the authors have begun drilling the adjacent bone in all cases
2	A partial inferior turbinectomy with a sphenoethmoidectomy and a wide maxillotomy	4	None	Removing the posterior part of the inferior turbinate automatically provides the extra anterior 'margin'
3	A complete inferior turbinectomy with a sphenoethmoidectomy and a wide maxillotomy	2	None	This trajectory allows for just the needed amount of space required to begin the dissection; removing the inferior turbinate allows for early division of the lateral attachments of the tumor, as well as bone drilling. Coupled with the early removal of the nasopharyngeal adhesions, allows for inferior traction, thus increasing the working space 'anteriorly'
4	A partial medial maxillectomy without drilling the frontal process of the maxilla	11	Three recurrences	Given the higher rate of recurrence in this category, it would be wise to go for the approach mentioned below (vide infra)
5	A partial medial maxillectomy with drilling the frontal process of the maxilla and performing an elective dacryocystorhinostomy	5	• One patient had persistent postoperative epiphora, and the dacryocystorhinostomy has to be revised • One recurrence	Given the fact that it would be better to err on the side of doing 'too much' rather than 'too little', this approach may be recommended whenever in doubt (vide supra)
6	The above in 5 combined with a canine puncture	3	None	Besides allowing for a shorter 'effort arm' (see text), it also allows for the second surgeon/second hand to not use the same port of access (i.e. transnasal), thus allowing for more maneuverability
7	A trans-septal single-surgeon	2	None	While this may have been described as a separate technique by others, in the authors, experience, it has been considered a consequence of the standard technique. Nevertheless, the author has used an 'extended' posterior septal removal on two occasions to gain access
8	A trans-septal 'two-surgeon, four-hands' technique	5	• One recurrence • One tumor had to be removed partially as the surgery has to be abandoned due to massive blood loss and was thus staged	This is now the preferred technique by the authors in most extensive cases

not really help in gaining much access to the posterolateral portion of the tumor (Fig. 17).

8. A Trans-septal 'Two-surgeon, Four-hands' Technique

This changes things dramatically. The advantages of a 'two surgeon, four-hands' technique has been well established as an acceptable way to tackle skull base pathologies,[7-13] and in continuation with the same, it would be a better modification of the procedure mentioned in above step (*vide supra*), with the combined benefits of a trans-septal approach (which may be considered as a 'consequence' of surgery, as well as four hands, which are separated by the cartilaginous septum, so as to compliment each other's efforts, without coming in the way of the other).

It is prudent to understand that though endoscopy is here to stay the good old external methods are not fading into oblivion. A surgeon attempting radical surgery using an endoscope should keep in mind the fact that it may be essential at sometimes to convert to an open approach and hence should be conversant with these too.

CONCLUSION

In the authors' experience, planning a surgical approach has proved to be invaluable in predicting difficulties and devising techniques to overcome the same. They have had their share of recurrences (6 out of 39), and have tried to identify the critical points which may be tackled to decrease the same. Their longest follow-up has been of 57 months (with an average of 36 months) and although the sample size has been small (n = 39) it has been comparable with most series published. They (the authors) have excluded patients with intracranial involvement from their series, as the focus in this article has been more for the infratemporal fossa extensions of the tumor.

ACKNOWLEDGEMENT

A few images used in this article have been taken from the chapter "Khattar VS, Hathiram BT. Endoscopic management of juvenile nasopharyngeal angiofibroma. Atlas of Operative Otorhinolaryngology and Head and Neck Surgery (1st ed). Jaypee Brothers Medical Publishers, New Delhi, India. 2013;1:645-63." Copyrighted to Ms Jaypee Brothers Medical Publishers, New Delhi, India.

REFERENCES

1. Khattar VS, Hathiram BT. Endoscopic management of juvenile nasopharyngeal angiofibroma. Atlas of Operative Otorhinolaryngology and Head & Neck Surgery (1st ed). India, New Delhi: Jaypee Brothers Medical Publishers 2013;1:645-63.
2. Hackman T, Snyderman CH, Carrau RL, et al. Juvenile nasopharyngeal angiofibroma: The expanded endonasal approach. Am J Rhinol Allergy 2009;23(1):95-99.
3. Khattar VS, Hathiram BT, Kurup B. Overcoming challenges in the endoscopic management of sinonasal tumours. Otorhinolaryngol Clin Int J 2011;3(3):168-75.
4. Denker A. Ein neuer Weg fur die Operation der malignen Nasentumoren. Munch Med Wochenschr 1906;53:953-56.
5. Sciarretta V, Pasquini E, Farneti G, Frank G, Mazzatenta D, Calbucci F. Endoscopic sinus surgery for the treatment of vascular tumours. Am J Rhinol 2006;20(4):426-31.
6. Harvey RJ, Sheehan PO, Debnath NI, Schlosser RJ. Trans-septal approach for extended endoscopic resections of the maxilla and infratemporal fossa. Am J Rhinol Allergy 2009;23(4):426-32.
7. Briner HR, Simmen D, Jones N. Endoscopic sinus surgery: Advantages of the bimanual technique. Am J Rhinol 2005;19:269-73.
8. May M, Hoffmann DF, Sobol SM. Video endoscopic sinus surgery: A two-handed technique. Laryngoscope 1990;100:430-32.
9. Kassam AB, Gardner P, Snyderman C, Mintz A, Carrau R. Expanded endonasal approach: Fully endoscopic, completely transnasal approach to the middle third of the clivus, petrous bone, middle cranial fossa and infratemporal fossa. Neurosurg Focus 2005;19:E6.
10. Kassam A, Snyderman CH, Mintz A, Gardner P, Carrau RL. Expanded endonasal approach: The rostrocaudal axis. Part II. Posterior clinoids to the foramen magnum. Neurosurg Focus 2005;19:E4.
11. Cavallo LM, Messina A, Cappabianca P, Esposito F, de Divitiis E, Gardner P, et al. Endoscopic endonasal surgery of the midline skull base: Anatomical study and clinical considerations. Neurosurg Focus 2005;19:E2.
12. de Divitiis E, Cappabianca P, Cavallo LM. Endoscopic transsphenoidal approach: Adaptability of the procedure to different sellar lesions. Neurosurgery 2002;51:699-707.
13. Locatelli D, Levi D, Rampa F, Pezzotta S, Castelnuovo P. Endoscopic approach for treatment of relapses in cystic craniopharyngiomas. Childs Nerv Sys 2004;20:863-67.

ABOUT THE AUTHORS

Vicky S Khattar

Assistant Professor, Department of ENT and Head and Neck Surgery Topiwala National Medical College and BYL Nair Charitable Hospital Mumbai, Maharashtra, India

Correspondence Address: C-7, Swati Kalyan Complex, Panch Marg Off Yari Road, Versova, Mumbai-4000061, Maharashtra, India Phone: 91-9930977110, e-mail: vickykhattar@rediffmail.com

Bachi T Hathiram

Professor and Head, Department of ENT and Head and Neck Surgery Topiwala National Medical College and BYL Nair Charitable Hospital Mumbai, Maharashtra, India