ENDOSCOPIC EXCISION OF JUVENILE NASOPHARYNGEAL ANGIOFIBROMA - A COMPREHENSIVE APPROACH

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ABSTRACT : *Objective: Endoscopic excision of Juvenile Nasopharyngeal Angiofibroma (JNA) was carried out with the objective of minimizing blood loss and attempting a complete excision of the tumor under direct vision with the help of Hopkins telescopes.*
Study Design: A prospective 4 year study of 23 cases of JNA treated by endoscopic excision is presented. Of these, 18 were treated by endoscopic excision alone. The remaining 5 were treated with a two staged approach either by mid-facial degloving followed by endoscopy or by 2 endoscopic procedures.
Results: The tumor was excised completely in 17 out of the total 18 cases that were treated exclusively by endoscopy. One case has shown a recurrence. The 5 cases treated by the staged approach represented very large tumours or tumours with intra-cranial extensions. In 1 of these cases, inoperable tumor remnant engulfing the internal carotid artery was treated by radiotherapy post-operatively.
Conclusion: With successful excision of JNA in all but one case, we could reasonably conclude that endoscopic excision of JNA could become a safer and a more precise alternative to open surgery provided it is practiced judiciously by surgeons who have considerable experience in endoscopic surgery and the necessary backup to convert to open surgery should the need arise.

Key Words: *Juvenile Nasopharyngeal Angiofibroma, Endoscopic excision,*

INTRODUCTION

JNA is a histologically benign yet locally aggressive vascular head and neck tumor. Though it is not a very common neoplasm, accounting for only 0.05% of head and neck tumors¹, it is nevertheless considered the most common benign neoplasm of the nasopharynx. The highest incidence is found in India and Egypt. It commonly affects young pubescent males.

JNA usually originates from the region of the sphenopalatine foramen just posterior to the middle turbinate². Although benign, the tumor causes pressure erosion of the bones. It can spread laterally into the pterygopalatine and infra-temporal fossae. Its medial and anterior spread involves the nasopharynx, nasal cavity and maxillary sinus. Superior extension of the tumor can involve the sphenoid sinus, the apex of the orbit and superior orbital fissure³. It can erode intracranially either medial or lateral to the cavernous sinus and internal carotid artery. Intracranial extension is the most commonly used criterion to divide patients into prognostic and therapeutic groups.

The preferred treatment for extracranial disease is surgical excision and radiation is reserved for unresectable tumors

probably those which have spread intracranially medial to the cavernous sinus⁴. Many surgical approaches have been used for resecting JNA depending on the tumor extent and spread as evidenced on CT scan. The transpalatine approach, lateral rhinotomy, mid facial degloving, Weber Fergusson, transmandibular, transantral, tranzygomatic infratemporal approach or a combination of these have been used in an attempt to ensure complete tumor removal⁵. We present our experience with a relatively new surgical

Fig 1 : Axial pre operative CT scan showing an enhancing mass in the right nasal cavity, nasopharynx, pterygopalatine fossa & infratemporal fossa with widening of the pterygomaxillary fissure.

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Fig. II : External carotid artery embolisation, pre-embolisation plate showing tumor blush.

Fig. III : Post embolisation plate showing disappearance of tumor blush.

technique that involves endoscopic excision of JNA using Hopkins telescopes.

MATERIAL AND METHODS

23 cases of JNA were operated endoscopically from August 1999 to October 2003. 14 of these were newly diagnosed cases while 9 were referred as cases of recurrence following previous surgery done elsewhere. All patients presented with common symptoms of epistaxis and nasal block. One patient had frank meningitis and presented with headache and vomiting.

The investigation findings and time interval between embolisation and surgery of individual patients are recorded in table no. I. All patients underwent contrast enhanced CT scan of the paranasal sinuses to determine the exact tumour extent (Fig.I). The selection criteria of the patients were based on the tumor extension evident on CT scan as well as on diagnostic nasal endoscopy. Patients with tumors present in the nasal cavity, nasopharynx and extending into the sphenoid sinus,

Fig. IV : Schematic diagram showing tumor extensions marked by the shaded area. The posterior half of the medial and posterior walls of the maxillary sinus are removed to release the bottlenecking of the dumb-bell shaped tumor.

Bone		Tumor	
MS - Maxillary Sinus		NP - Nasopharynx	
S - Septum		PPF - Pterygopalatine fossa	
		NC - Nasal cavity	
		IT - Infratemporal	

..... Maxillary sinus wall removed

Fig. V : Photograph of specimen indicating different parts of the tumor.

pterygopalatine fossa and the infra-temporal fossa (Fisch Stage I, II, IIIa) were selected for the endoscopic approach alone. Large tumours with or without intra-cranial extensions (Fisch Stage IIIb, IV) were selected for the staged approach. Bilateral carotid angiography with embolisation of the feeder vessels using gelfoam or polyvinyl alcohol was done prior to surgery in all the patients (Fig II, Fig III).

All patients were operated within 24-72 hrs of

Fig. VI : Pre-operative axial CT scan showing extensive tumour involving both the nasal cavities, posterior choanae, nasopharynx, left pterygopalatine fossa, infratemporal fossa, bilateral ethmoid and sphenoid sinuses, left cavernous sinus and surrounding the left internal carotid artery.

Fig. VII : Post-operative axial CT scan after mid-facial degloving where the bulk of the tumour has been removed leaving behind tumour in the sphenoid sinus and cavernous sinus.

Fig. VIII : Post-operative axial CT scan following staged endoscopic procedure showing a small residual tumor in the left cavernous sinus.

Fig. IX : Pre-operative axial CT scan showing an enhancing mass in the right nasal cavity and nasopharynx.

Fig. X: Follow-up post-operative axial CT scan of the same patient for comparison showing a clear nasal cavity and nasopharynx.

embolisation. None of the patients faced any complications of angiography and embolisation.

The basic operative technique was similar in all patients with minor individual variations. The patient was placed in supine position with head elevation of 30 degrees. Hypotensive general anaesthesia was used with the systolic blood pressure being maintained between 80-90 mm Hg. The eyes were not covered by drapes. Both nasal cavities were packed with cotton pledgets soaked in saline-adrenaline solution in a concentration of 1:100,000. After good decongestion of the nose, the tumor extension in both the nasal cavities and nasopharynx was examined carefully using 4 mm 0° and 30° telescope transnasally and a 70° telescope transorally. Well-lubricated nasal catheters were gently passed through the nasal cavities under endoscopic guidance to elevate the soft palate.

The middle turbinate was infiltrated with saline adrenaline (1: 200,000 concentration) and excised after cauterizing and dissecting away the tumour attached to it. This was

Table I: Investigation findings

Case No.	Age (yrs) Sex	CT Scan findings													Stage Fisch classification	Interval between embolisation & surgery
		RNC	LNC	PC	NP	PPF	ITF	MS	SS	ES	IOF	TF	IC	CS		
1. SK	14/M	✓		✓	✓				✓						II	48 hours
2. AB	18/M		✓	✓	✓				✓						II	24 hours
3. SA	15/M	✓	✓	✓	✓	✓		✓							IIIa	60 hours
4. RS	13/M	✓		✓	✓	✓									IIIa	48 hours
5. AS	20/M	✓		✓				✓							II	65 hours
6. RG	20/M	✓		✓	✓	✓	✓		✓	✓					IIIa	48 hours
7. MP	14/M	✓		✓	✓				✓						II	40 hours
8. MG	17/M		✓	✓		✓		✓		✓	✓	✓	✓		IIIb	72 hours
9. DY	16/M		✓	✓	✓	✓	✓		✓	✓	✓				IIIa	48 hours
10. SA	25/M		✓	✓	✓				✓	✓					II	72 hours
11. MD	15/M	✓		✓	✓	✓		✓	✓	✓					IIIa	72 hours
12. AJ	11/M		✓	✓	✓	✓		✓							IIIa	72 hours
13. MK	15/M			✓	✓			✓		✓					II	36 hours
14. GN	14/M	✓	✓	✓	✓	✓	✓	✓	✓	✓			✓	✓	IVb	48 hours
15. N	17/M			✓	✓	✓			✓						IIIa	16 hours
16. AV	15/M		✓	✓	✓				✓						II	72 hours
17. KL	25/M		✓	✓	✓	✓			✓						IIIa	72 hours
18. SJ	M/20		✓	✓	✓	✓	✓		✓						IIIa	48 hours
19. RT	M/23		✓	✓	✓	✓			✓		✓				IIIa	72 hours
20. SS	M/19		✓	✓	✓	✓	✓		✓		✓		✓		IIIb	48 hours
21. A	M/12	✓		✓				✓							II	48 hours
22. AD	M/17	✓		✓	✓	✓	✓		✓						IIIa	48 hours
23. DP	M/20		✓	✓	✓	✓	✓	✓	✓						IIIa	48 hours

RNC- right nasal cavity, LNC- left nasal cavity, PC-Posterior choanae, NP- nasopharynx, PPF -pterygopalatine fossa, ITF – infratemporal fossa, MS- maxillary sinus, SS - sphenoid sinus, ES- Ethmoid sinus, IOF –inferior orbital fissure, TF – temporal fossa, IC – intracranial, CS- cavernous sinus.

important so as to obtain a good view of the sphenopalatine foramen especially where there was a lateral extension of the tumor. The tumor attachments to various regions, e.g. the middle turbinate, septum and the roof of the posterior choana were carefully identified cauterized with a bipolar diathermy and then dissected using a 3-mm periosteal elevator. A posterior ethmoidectomy was done and the sphenoid sinus entered. Extension into the sphenoid sinus was visualized and dissected after removing part of the anterior and inferior sphenoid sinus wall adjacent to the sphenopalatine foramen.

In cases with lateral spread of the tumor, the maxillary sinus was entered through the posterior fontanelle. The

posterior half of the medial and the posterolateral wall of the maxillary sinus, the perpendicular plate of the palatine bone and the anteroinferior rim of the sphenopalatine foramen were removed with a bone punch to expose the tumor in the pterygopalatine fossa and medial part of infratemporal fossa. These extensions were cauterized and dissected meticulously and any bleeding was controlled with adrenaline soaked patties and bipolar diathermy. This released the bottlenecking of the dumb-bell shaped tumor at the region of the sphenopalatine foramen (Fig.IV).

Dissection was then continued posteriorly to free the tumor of its attachments to the roof of the nasopharynx and posterior margin of the septum. After ensuring that the

Table II: Approach used, operative and follow-up details

Case No.	Operative Approach	Blood loss	Blood transfused	Nasal packing	Follow-up
1. SK	Endoscopic	120cc	-	Nil	4 years. No recurrence
2. AB	Endoscopic	80cc	-	Nil	2½ years. No recurrence
3. SA	Endoscopic	450 cc.	1 unit	Nil	2 yrs, 4 months. No recurrence
4. RS	Endoscopic	410cc.	1 unit	Nil	3 yr, 3 months. No recurrence
5. AS	Endoscopic	200 cc	-	AN pack since SMR was done	3 yr, 3 months.No recurrence
6. RG	Endoscopic	110 cc	-	Merocel	3 yr, .No recurrence
7. MP	Endoscopic	500 cc.	1 unit	Merocel pack, since septoplasty was done	3 yr, No recurrence
8. MG	Endoscopic Endoscopic	650cc 100cc	2 unit -	AN+PN pack AN+PN pack	1 year 1 month. No recurrence
9. DY	Approach by Weber-Fergusson incision Endoscopic	1.9 litres 800cc	2 units -	AN+PN pack AN+PN pack	2 year 7 months. No recurrence
10. SA	Endoscopic	400 cc	-	Merocel	1 year 11 months. No recurrence
11. MD	Endoscopic	350 cc	-	Merocel	1 year 11 months
12. AJ	Endoscopic	700 cc	2 units	AN pack	1 year 10 months, Recurrence present
13. MK	Mid facial degloving approach Endoscopic	2.7litres 1.6 litres	5 units 3 units	AN+PN pack AN+PN pack	2yeras 2 months. No recurrence
14.GN	Mid facial degloving approach Endoscopic	2 litres 1.6 litres	5 units 3 units	AN+PN pack AN+PN pack	2 years 6 months Residual tumor in cavernous sinus subjected to radiotherapy
15. N	Mid facial degloving approach Endoscopic	2 litres 750cc	6 units Autotransfusion	AN+PN pack AN+PN pack	1 year 8 months No recurrence
16. AV	Endoscopic	150cc	-	Merocel	1 year 3 months No recurrence
17. KL	Endoscopic	50 cc	-	Merocel	1 year 7 months No recurrence
18. SJ	Endoscopic	25 cc	-	Merocel	10 months
19. RT	Endoscopic	100 ml	-	Merocel	6 months
20.SS	Endoscopic	600 cc	3 units	AN	5 months
21. A	Endoscopic	30 cc	-	Merocel	
22. A D	Endoscopic	600 cc	2 units	AN+ PN	3 months
23. D P	Endoscopic	100 cc	-	Merocel	2 months

AN- Anterior nasal, PN- Posterior nasal

tumor was free of all its attachments it was delivered transorally (Fig.V). Bleeding from the sphenopalatine vessels usually occurred at this point and was controlled with bipolar cauterization.

After removal of the main mass, the area was examined

endoscopically for any tumor remnants and if present, these were removed with an upward biting forceps. The intraoperative blood loss varied from 50 cc to 700 cc and most patients did not require any anterior or postnasal packing following the surgery. The average operative time for the surgery was four hours. The average duration of

hospital stay following surgery was for two-three days. The 5 cases selected for a staged approach had either very large tumours or intra-cranial extensions (Fig 6). Here the bulk of the tumour was removed by a mid-facial degloving approach (Fig 7). Residual tumour in sensitive areas like the cavernous sinus, orbit and those parts of the tumour which were inaccessible with the open approach were removed at a pre-planned second sitting by meticulous endoscopic resection. Inoperable remnants engulfing the internal carotid artery were subjected to radiotherapy (Fig 8). These 5 cases had a blood loss of 100cc to 2700 cc. These patients required anterior and post-nasal packing following surgery.

RESULTS

In all the 18 cases managed endoscopically, it was possible to completely excise the entire tumor under endoscopic guidance in 17 cases. We did not encounter any complications during the procedure. One patient had a small perforation at the junction of the hard and soft palate where extensive dissection of the adhesions had to be done. The second patient (GN) who had an erosion of the roof of the sphenoid sinus with exposure of the pituitary sella and cavernous sinus developed diabetes insipidus and ptosis in the immediate post-operative period. This was as a result of the tight pressure of the intra-nasal packs. The diabetes insipidus settled after removal of the nasal packs. The ptosis recovered after the staged endoscopic removal of the sphenoid sinus remnant. One of the cases managed by the staged approach had a residual intra-cavernous tumour engulfing the internal carotid artery. This was subjected to radiotherapy and is under close surveillance. One case of endoscopic excision has a tumour recurrence in the region of the inferior orbital fissure.

Our protocol for following up the patients includes an endoscopic examination at 1 month, 3 months, 6 months and 12 months post-operatively. We also do a CT scan/MRI 1 year after surgery (Fig. IX) & (Fig. X). Intraoperative findings and follow up details of individual patients are recorded in table II.

DISCUSSION

Endoscopic surgery of the nose and paranasal sinuses has gained wide acceptance over the last decade. With its evolution, better instrumentation and increasing experience in the field, it is now possible to consider endoscopic

excision of JNA as a viable alternative or a useful adjunct to open surgery.

Although a great deal of work still needs to be done in this area, our experience indicates that complete and safe endoscopic excision of JNA is not only possible but may be the preferred method in selective tumors.

Typically, conventional open surgery has been associated with severe hemorrhage. Significant bleeding may also impede accurate visualization of tumor extensions⁶, often resulting in residual tumor being left behind. This results in the quite frequently recorded recurrence rate of the tumor. Residual remnants of the tumor can also cause postoperative haemorrhage. Thus conventionally patients operated for JNA require tight anterior and posterior nasal packing for a period of 3-5 days. These packs, besides being extremely uncomfortable for the patient have to be removed as a second procedure usually under general anesthesia. In young pre-pubescent males, surgical trauma to the facial skeleton may adversely impact the mid facial growth in the future⁷. Also, the presence of an external scar on the face in certain surgical approaches is a distinct disadvantage in these young boys.

Endoscopic excision of JNA ensures that every attachment of the tumor can be visualized, cauterized with a bipolar diathermy and dissected precisely. Since this dissection is done under direct vision in the plane between the tumor and the surrounding tissues, the tumor tissue itself is not breached in the process⁸. This ensures that the blood loss is minimal. The internal maxillary artery may tether the tumour in the infra temporal fossa. The vessel may need to be clipped or cauterized before freeing the tumour. A specially designed crocodile action insulated diathermy forceps was used for this purpose. Anatomical bony partitions such as the perpendicular plate of the palatine bone, the anterior rim of the sphenopalatine foramen and the posterolateral wall of the maxilla also can be removed under direct vision. This is especially important so as to remove the bottlenecking of the dumbbell shaped tumor that occurs at the sphenopalatine foramen⁸. Virgin tumours, not operated previously, tend to follow a definite pattern of adherence to surrounding mucosa. We consistently found adhesions to the middle turbinate, superior turbinate, posterior end of septum, roof of nasopharynx and fossa of Rosenmuller. There were no adhesions to the inferior turbinate, floor of nose or the Eustachian tube. Spread in

the sphenoid sinus is usually submucosal and the tumour takes no direct adhesions to the walls of the sphenoid sinus.

Surgery in a relatively bloodless field under direct vision also ensures complete removal of the tumor with less likelihood of remnants being left behind. Any remnants of the tumor can be dissected endoscopically and removed after the main mass has been delivered per orally. With the standard external approaches some areas still remain inaccessible to the surgeon's probing finger. One such area is the junction of the sphenoid bone and the perpendicular plate of the palatine bone. Tumour remnants in these areas can then be excised endoscopically at a second stage. Understandably we expect the rates of recurrence of JNA to decrease with this surgical technique. From the patient's point of view, endoscopic surgery is a far less morbid procedure. The patient has no packs postoperatively and is able to breathe, eat and talk comfortably within hours of surgery. The total duration of hospital stay is considerably reduced and the absence of any facial scar is a significant advantage in these adolescent patients.

Relative drawbacks that we faced were :-

- 1) Increase in the intra-operative time which on an average was four hours. However with greater experience in this technique, it has been possible to reduce the operative time.
- 2) Lack of 'elbow space' to move around large intra-nasal masses.
- 3) Severe intra-operative haemorrhage may be difficult to control endoscopically. This would entail converting the endoscopic approach to an open surgical approach.

There are certain prerequisites necessary for endoscopic surgery to be successful.

1. A good and effective preoperative embolisation is mandatory if endoscopic excision is to be attempted⁹. We feel that the ideal time to operate would be within 24-72 hours of embolisation. One case was taken up for surgery within 16 hours of embolisation. Intraoperative bleeding in this case prevented endoscopic excision and open surgery became necessary. The residual tumor in the sphenoid sinus was removed later at a second stage.
2. Before attempting endoscopic excision, all preparations for conventional open surgery if required, should be kept ready, e.g. reservations of adequate blood units,

preparation of surgical instrument trolley, informed consent etc.

3. Initially we restricted this technique for tumors that were limited to the nasal cavity and paranasal sinuses with minimal extension into the pterygopalatine fossa. We later used endoscopic surgery for medium size tumours involving the nasal cavity, nasopharynx, maxillary sinus, sphenoid sinus, pterygopalatine fossa and medial portion of infra-temporal fossa (Fisch stage I, II and III a)¹⁰. Larger tumours with far lateral extensions into the infra-temporal fossa and those with intra-orbital and intra-cranial spread (Fisch stage III b and IV) were tackled later in our study as our experience grew.
4. Meticulous care and patience is required to dissect all attachments of the tumor from the underlying bone without entering the tumor tissue itself. It may be necessary to excise the posterior part of the middle turbinate, the perpendicular plate of palatine bone and the medial and posterior walls of maxillary sinus in order to visualize the complete tumor.
5. A stringent follow-up protocol where the patient is assessed both endoscopically and with a CT scan with contrast / MRI should be carried out in all patients on a long term basis. Ideally if finances permit, an MRI is the preferred option at 1 year following surgery. Since very small recurrent tumours may remain asymptomatic for a longer period of time, the patients should be followed up at 6 monthly intervals thereafter over the subsequent years. This will ensure that recurrences are diagnosed and treated much earlier, even before they become symptomatic. With increasing experience we have begun to identify prognostic indicators for predicting outcome in these difficult tumours. A poor prognosis i.e. a recurrence or incomplete removal of tumour is more likely in cases where there has been excessive bleeding, dense adhesions, involvement of the superior and inferior orbital fissures, piecemeal removal and previously operated tumors. Surgery on a recurrent tumour has a poorer prognosis as these tumours tend to have diffuse and dense adhesions, a mixed supply from the internal and the external carotid artery systems and a poor demarcation of the tumour from the surrounding tissues.

CONCLUSION

On the basis of this study, one can conclude that selected cases of JNA with tumor extensions in the nasal cavity,

nasopharynx, sphenoid sinus, pterygopalatine fossa and the medial portion of the infra-temporal fossa can be successfully managed endoscopically with prior embolisation. Distinct advantages of this procedure are complete tumour excision, minimal blood loss, avoidance of nasal packs, shorter duration of hospital stay and a reduced rate of recurrences. We would like to stress here that only surgeons who have gained considerable experience in the field of endoscopic surgery must embark upon this procedure. With the inherent propensity of the tumor to bleed as well as with vital structures like the orbit and contents of the pterygopalatine and infratemporal fossa involved in the field of dissection, it is not a recommended procedure for a novice endoscopist. In conclusion, with increasing experience, endoscopic excision of JNA could become a safer, more precise and complete alternative to open surgery in selected tumors. It would also be a useful adjunct to open surgery in very extensive tumors.

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REFERENCES

1. Batsakis J G (1979): Vasoformative tumors. In Batsakis J G; editor. Tumors of the Head and Neck, Clinical and Pathological Considerations. 2nd ed. Baltimore, London: Williams & Wilkins Co. : 296-300.
2. Neel HB III, Whicker JH, Devine KD et al (1973): Juvenile Angiofibroma: review of 120 cases. American Journal of Surgery; 126: 547-56
3. Shaheen OH (1987): Angiofibroma. In: Evans John NG, Kerr AG, Groves J. editors. Scott-Brown's Otolaryngology. 5th ed. London, Boston: Butterworth: Vol 6, 291-96.
4. McGahan RA, Durrance FY, Parke RB Jr et al (1989): The treatment of advanced juvenile nasopharyngeal angiofibromas. International Journal of Radiation Oncology and Biol. Physics; 17(5): 1067-72
5. Neel H.B.III, (1985) Juvenile Angiofibroma: In: Blitzer A, Lawson W, Friedman WH, editors. Surgery of the Paranasal Sinuses. 1st ed. Philadelphia: W. B. Saunders Co.; 247-259
6. Herman P, Lot G, Chapot R et al (1999): Long-term follow-up of Juvenile nasopharyngeal angiofibromas: analysis of recurrences, Laryngoscope 109:140-147.
7. Shenoy AM, Grover N, Janardhan N et al (2002): Juvenile Nasopharyngeal Angiofibromas: A study of recurrence pattern and role of pre-operative embolisation – 'A decade's experience', Indian journal otolaryngology and head and neck surgery 54(4), 274-279.
8. Kamel RH (1996): Transnasal endoscopic surgery in juvenile nasopharyngeal angiofibroma, Journal of Laryngology and Otology; 110: 962-8.
9. Tranbahuy, Borsik M, Herman P et al (1994): A Direct intratumoral embolisation of juvenile angiofibroma, American Journal of Otolaryngology 15 (6): 429-35
10. Fisch U (1983): The infratemporal fossa approach for nasopharyngeal tumors. Laryngoscope; 93(1): 36-44.

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