

Original Research Article

Trend analysis in management of juvenile nasopharyngeal angiofibroma: our institutional experience

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Received: 01 August 2019

Revised: 30 October 2019

Accepted: 05 November 2019

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ABSTRACT

Background: Juvenile nasopharyngeal angiofibroma is a vascular malformation rather than true neoplasm. It accounts for 0.5% of all head and neck tumors and its general incidence is 1:150,000. It is uncommon, benign, extremely vascular tumor that arises from tissues in the sphenopalatine foramen, the pterygoid base causing Early spread submucosally towards nasopharynx. Surgical excision is mainstay of treatment.

Methods: This is a retrospective observational study carried out from March 2002 to March 2019, 130 patients underwent surgical resection of juvenile nasopharyngeal angiofibroma at Civil Hospital, Ahmedabad. Patients were divided in two groups with Group-A having patients from 2002-2010 and Group B having patients from 2011-2019.

Results: A total 130 cases of juvenile nasopharyngeal angiofibroma with a mean age of 16.25 years (range 9-33) were treated by surgical excision from March 2002 to March 2019. In Group A patients open approach was used most often with endoscopic approach used only till stage II-A. In Group B patients the preferred surgical approach was trans-nasal endoscopic approach cases up to stage IV-A and open approach including the midfacial degloving approach and infratemporal approach was used cases with extensive intracranial, lateral infratemporal fossa, orbit, optic nerve, cavernous sinus involvement.

Conclusions: Our study shows that over the years there has been shift in surgical approaches used in treating angiofibroma with the advent of newer endoscopic method and addition of various technology such as coblation, navigation has made the procedure for a highly vascular tumor simpler.

Keywords: Endoscopic method, Juvenile nasopharyngeal angiofibroma, Trend analysis

INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) consists of proliferating, irregular vascular channels with fibrous stroma. Incidence is less than 0.5% of all head and neck tumors.¹ It is seen exclusively in adolescent male, but also reported in children, elderly, males, pregnant females. Tumor blood vessels lack smooth muscle and elastic fibers leading to sustained bleeding. Plump cells in stroma forms collagen which makes some tumors very hard to firm others soft.²

Androgen receptors are present in 75% of the tumors both in stromal and vascular elements.³ VEGF, vessel density and expression and localization of VEGF correlates with proliferative marker Ki67, but it does not correlate with degree of aggressiveness.⁴ Two main presentations are nasal obstruction and recurrent severe epistaxis, facial swelling, visual disturbance are also seen. Bony erosions of pterygoid, clivus, sphenoid sinus floor is seen. JNAs spread along natural foramina, fissures. Surgical resection is accepted as treatment modality for JNA.⁵ Indeed, refinements in cameras, instrumentation,

and navigation systems allowed trans-nasal endoscopic removal with significant improvement in morbidity and mortality and better cosmetic and functional results.⁶

Aim of our study was to observe the trend of approaches used in management of angiofibroma over the years in a tertiary care centre of Gujarat, India. We used trans-nasal endoscopic method in different stages of angiofibroma with use of recent technologies like coblation, microdebrider, navigation in addition to for tumor excision.

METHODS

This is a retrospective observational study of 130 cases of juvenile angiofibroma presented at Civil Hospital Ahmedabad from March 2002 to March 2019. We collected the data from our hospital record system and follow up examinations. Data such as clinical findings at presentation, demographic profile, endoscopic findings, radiological investigations, ophthalmological findings, laboratory investigations were collected both pre and post operatively.

Patients were divided in two groups, Group-A contained patients from 2002 to 2010 and group B contained patients from 2011 to 2019. Most of these patients underwent surgical removal of angiofibroma except few inoperable cases which underwent for radiotherapy hence excluded from the study. We used modified FISCH classification in the staging of tumor. Embolization was not done in our patients. Post-op scan was done after 48 hours. Residual disease was defined as tumor seen in immediate post-op scan; recurrent disease was defined as tumor seen in scan after a negative post-op scan. Follow up was taken every month for first 6 months then every 6 months for next 2 years and then every yearly. Parameters such as Blood loss, hospital stay, recurrence, complications intra and post operatively were used to measure the efficacy of the approach used.

The research was approved by the local Ethical committee. Prior consent was taken from the patients and for patients who were minors consent was taken from their parents.

Inclusion criteria

- Patients having histologically proven JNA.
- Patients who underwent surgical excision of tumor for the treatment.

Exclusion criteria

- Patients with inoperable tumor.
- Patients in who did not give consent.
- Patients in whom proper follow up was not achieved.
- Patients with bleeding disorder.

Pre-op evaluation

A thorough pre-operative assessment is must including detailed clinical history, endoscopic assessment, hematological profile, co-morbidities and p/h/o any previous surgery. Figure 1 shows characteristic appearance of JNA as seen on endoscopic assessment.

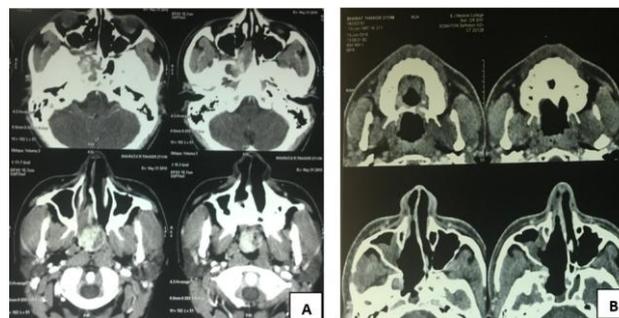


Figure 1 (A and B): Pre-operative and post-operative stage I JNA respectively.

Radiological investigations

CECT of paranasal sinus is the investigation of choice for staging of juvenila nasopharyngeal angiofibroma as well as deciding the surgical approach. “Holman-Miller sign”, “Hondousa sign”, “Ram-Haran sign” are characteristic of JNA.⁷ CECT used is Helical with 0.625 mm thickness, UV contrast is injected at 3 to 3.5 ml/s and pre and post contrast images are made in 2 different window settings. CT angiography, 3 DCT volume rendering DSA are other important investigations. MRI brain with PNS helps when the tumor involves skull base. It shows salt-pepper appearance and on contrast it chows intense enhancement.⁷ Post-op CECT is done after removal of packs and is repeated every 6 months. Computer tomography angiography, although not essential, is often useful in defining the feeding vessels. We have used this investigation in patients with stage IV (FISCH) JNA.⁷

Pre-operative embolization

We don't prefer to use pre-operative embolization in our patients. In some centers preoperative embolization is a prerequisite for endoscopic surgery in all cases of JNA.¹² This idea is questionable. It seems that preoperative embolization in small tumors has no role in reducing intraoperative bleeding.¹³ Lloyd believes that preoperative embolization increases the risk of leaving residue during surgery.¹⁴ This idea has not been proved in many other similar studies. We used vessel loops around ECA for reducing intra tumoral blood flow intraoperatively.

Radiation therapy

We preferred radiation therapy in inoperable tumor. Some surgeons recommend this type of treatment in advanced tumors or in recurrent cases that have no chance of total tumor resection with acceptable morbidity.¹⁰

Surgical management

Positioning and preparation

After the routine set-up of sinus surgery. The head of the bed is elevated 20-30 degrees to aid in hemostasis. Multiple studies have showed decreased central venous pressure at 5-25 degree of reverse trendelenburg position. 2 wide bore intravenous lines are secured in case of significant blood loss and need of blood transfusion.

Surgical steps

Nasal cavity is decongested with 1: 100000 adrenaline solutions and xylometazoline solution for 15 minutes. This reduces the mucosal bleeding intraoperatively and creates adequate space for instrumentation. Trans-nasal infiltration of nasal mucosa with 2% lignocaine and 1:100000 adrenaline is done around the tumor. Warm saline irrigation is done to facilitate vasoconstriction. Anatomical endoscopic landmarks are identified. Optimum tumor exposure and visualization is achieved by several surgical maneuvers. (posterior septectomy, medial maxillectomy). When approaching the tumor, establishing the location of origin is imperative. Careful blunt dissection around the visible tumor may identify tissue planes that delineate the tumor from surrounding normal structures.

Endoscopic removal of modified fisch stage I of JNA

After wide antrostomy and ethmoidectomy, thin bone between ethmoid and sphenoid sinus is removed to visualize tumor.

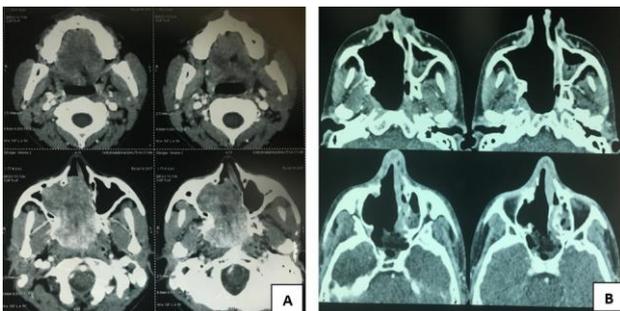


Figure 2 (A and B): Pre-operative and post-operative stage II JNA respectively.

Endoscopic management of Fisch stage II JNA uncinectomy, wide antrostomy followed by middle and inferior turbinectomy is done. Ethmoid and sphenoid sinus opened, posterior septectomy is done, periosteum of pterygopalatine fossa is incised, sphenopalatine artery is cauterized. Tumor is released from its attachments to pterygopalatine fossa, sphenoid sinus and ethmoid sinus, tumor is freed from nasopharynx and delivered.

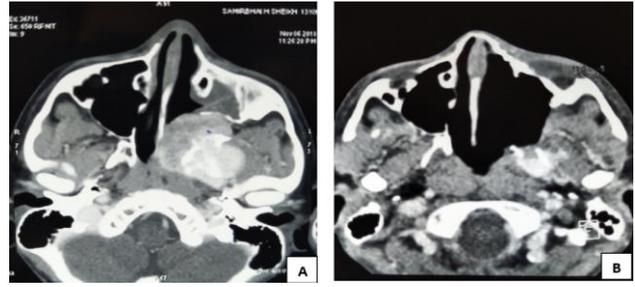


Figure 3 (A and B): Pre-operative and post-operative stage IIIA JNA respectively.

Management of stage IIIA JNA

Modified denker's approach-Incision of anterior edge of pyriform aperture; exposure and drilling/gauging of anterolateral wall of maxillary sinus, posterior wall of maxillary sinus removed; infratemporal fossa is exposed; internal maxillary artery is clipped; tumor is mobilized from its attachments in infratemporal and cheek; it is resected from pterygoid wedge and delivered

Endoscopic management of stage III-B JNA

Resection from pterygoid wedge remain same as stage III-A after which vidian canal identified, anterolateral wall of sphenoid is removed, quadrangular space exposed, internal carotid artery medialized, tumor lateral to ICA removed.

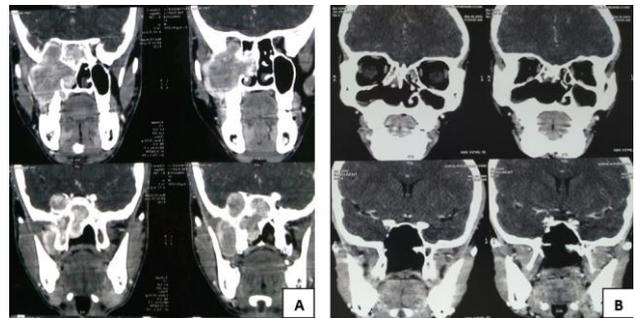


Figure 4 (A and B): Pre and post-operative stage IIIB JNA respectively.

Midfacial degloving approach

Midfacial degloving approach: It is a bilateral extended trans nasal maxillary approach. There is no visible scarring, adequate exposure of nasal complex, nasopharynx and middle third of the face is obtained. Sublabial incision is performed from one maxillary tuberosity to the other. Intercartilagenous incisions are given to separate soft tissue from the upper lateral cartilages. Incisions along the pyriform aperture connects the circumferential septal vestibular incisions to the sublabial incision. This allows total mid facial degloving up to the roof of the nose and infra orbital foramen

Transcranial approach combined with Dieffenbach's modification of Weber-Ferguson incision

Incision is made starting from the upper lip from philtrum, to gingivobuccal gutter, externally it extends in nasolabial and superiorly in nasofacial groove. Once the medial canthal region is approached the incision is extended inferior to lower eyelid-which is called Dieffenbach's modification of Weber-Ferguson incision Bicornal provides exposure to upper and middle 1/3rd of facial skeleton and anterior cranium. Incision-from upper attachment of helix of one side to opposite side, can be extended preauricular region to expose zygomatic arch. Flap is raised deep to the sub-aponeurotic areolar tissue. Periosteum is incised 3 cm above the supraorbital rim nasoethmoid nasofrontal, frontozygomatic region can thus be exposed. An incision in superficial layer of temporal fascia is made 2 cm lateral and 3 cm above the margin of the brow to preserve branch of facial nerve, the incision is connected with supraorbital incision.

Outcome variables and approach used were correlated using the spearman correlation, with significance level of 0.05.

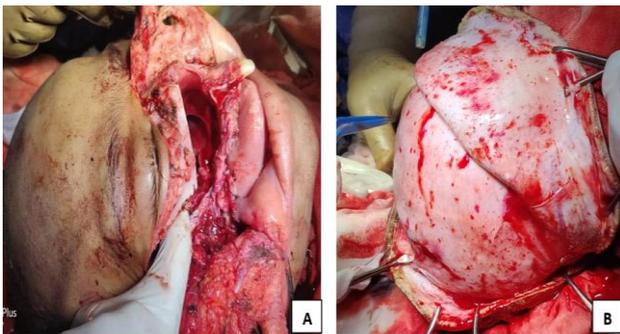


Figure 5 (A and B): Subtotal maxillectomy in the same patient.

RESULTS

Different therapeutic strategies have been proposed in the management of JNA during the recent years: radiotherapy, hormone therapy, cryotherapy and electrocoagulation; nevertheless, in most cases, surgery is the treatment of choice.^{10,11}

In our study all patients were male, mean age of 16.25 years (9-33). JNA at our institute presented as C/o nasal bleeding (91%) associated with or without nasal obstruction. Facial swelling and decreased vision were also seen as associated findings in 15% and 10% patients respectively (Figure 6).

In the Group A we had 56 and Group B we had 74 patients. Group A had 25 (44.64%) patients who were detected in stage III whereas Group B had more patients detected in stage II (43.24%). Detection in early stages was due to early clinical detection by nasal endoscopy

and radiological investigations. In Group A various open approaches such as trans-palatine, lateral rhinotomy, midfacial degloving were used at our institute. 56 cases in Group A of juvenile, open approach was used in 71% which included stage I-IV and endoscopic approach was used in 29% which included stage I-II. 74 cases of Group B were operated from 2011-2019 out of which open approach was used in 14% which mainly comprised of stage III-B and stage IV and endoscopic approach alone or in combination with open approach (for improving lateral access) was used in 86% of patients up to stage IV-A (Table 1). Transcranial approach was used in some patients which involved zygomatic and temporal region and with intracranial involvement. Comparison of approaches used in surgical management over the years is shown in the line diagram (Figure 7) which shows massive shift in management of angiofibroma at our institute. Complications such as massive intraoperative hemorrhage was seen in 3 patients with stage III-B and 1 stage IV-A, which led to intraoperative hypotension and further complication leading to death of the patient. Numbness over face, facial scarring, facial deformity post maxillectomy, trismus was more in open approach. orbital complication was more in endoscopic method. We had 3 cases of septal perforation. Rate of compliance was more in patients treated with endoscopic method and time of hospital stay was also less in the same group.

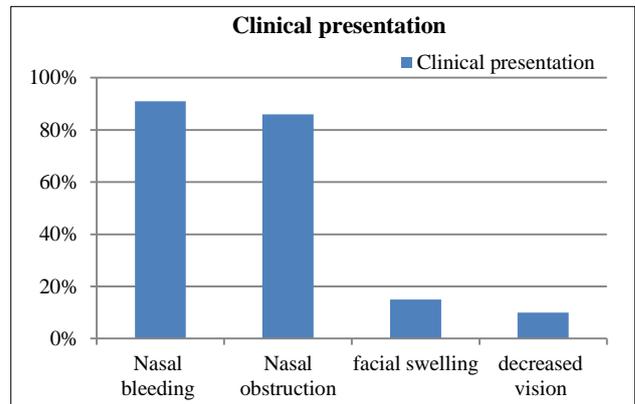


Figure 6: Clinical presentation of JNA.

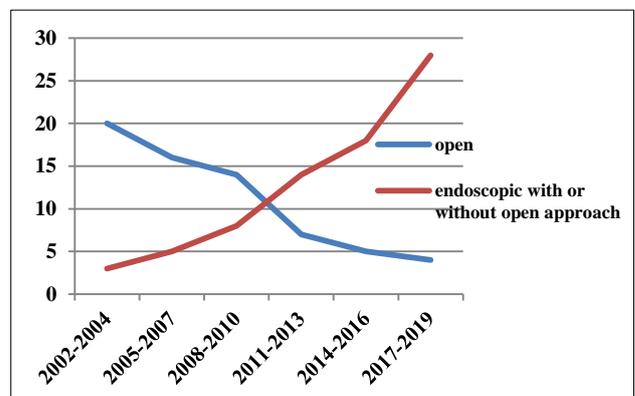


Figure 7: Tend of surgical approaches at our hospital facility.

Table 1: Comparison of variables between two groups of the study.

	Group-A (2002-2010)	Group-B (2011-2019)
Number of patients	56	74
Age distribution (in years)	9-33 years	9-33 years
Stage at presentation	Stage I -10 Stage II - 12 Stage III - 25 Stage IV - 9	Stage I -14 Stage II - 32 Stage III-18 Stage IV -10
Clinical presentation	Nasal bleeding Nasal obstruction Facial swelling Decreased vision	Nasal bleeding Nasal obstruction Facial swelling Decreased vision
Surgical approach used	Open-40 Transpalatine: Stage I: 4 Lateral rhinotomy: Stage II: 2 Stage III: 12 Mid facial degloving: Stage III: 13 Stage IV: 9 Endoscopic: 16 Stage I: 6 Stage II: 10	Open-16 Lateral rhinotomy: 6 Stage II: 2 Stage III: 4 Midfacial degloving: 8 Stage III: 3 Stage IV: 5 Transcranial: Stage IV: 2 Endoscopic alone or in combination with open approach: 58

Table 2: Comparison of variables between open and endoscopic approach.

	Open approach	Endoscopic approach alone or in combination with open approach
t	56	74
Mean procedure time	Stage I: 200 min Stage II: 230 min Stage III: 270 min Stage IV-A: 300 min	Stage I: 160 min Stage II: 180 min Stage III: 210 min Stage IV-A: 240 min
Mean blood loss intra op	Stage I: 1000 ml Stage II: 1400 ml Stage III: 2000 ml Stage IV-A: 2500 ml	Stage I: 400 ml Stage II: 550 ml Stage III: 600 ml Stage IV-A: 800 ml
Average hospital stays	5 days	3 days
Recurrence rate	17.85%	16.98%

Mean procedure time was 197 min in endoscopic method in all stages but was 250 min in open approach. This was due to better surgical field and magnification provided by endoscopic methods. Newer intervention such as coblation and microdebrider were also used to do maxillectomy, removal of surrounding tissue and hemostasis which led to faster hassleless procedure and less operative time. Mean blood loss was 1474 ml in open approach as we travel through the facial planes and limited exposure provided by open approach whereas in endoscopic method it was 587 ml. Average hospital stay for endoscopic method was 3 days while in open it ranged from 5-10 days (Table 2).

Rate of recurrence was found to be lesser in endoscopic method due to better visualization of operative field and

methods such as drilling and complete resection of tumor (Table 2).

DISCUSSION

Endoscopic endonasal management has been increasingly being used for infratemporal and upper parapharyngeal space >advanced JNA's are most commonly seen in this spaces. In this context Nicolai et al reported 17 cases of laterally extended JNA (14; stage IIIA: 3; stage: IIIB) in a series of 46 patients radically dissected through extended endoscopic approach.¹⁰ In a retrospective review, Battaglia et al, evaluated the use of endoscopic approach out of which 20 cases were of JNA, it suggested that purely endoscopic approach can be safely used for tumors extending in upper parapharyngeal space or infratemporal

fossa. The drawbacks of Denker's approach include the disruption of the medial maxillary buttress with possible loss of the lateral alar support. However, a contralateral trans-septal approach provides a new feasible door to access the laterally extended JNA, when placed too anteriorly may lead to septal perforation and/or loss of a tip support. The endoscopic approach provides a well illuminated, magnified and multiangled view of the surgical field and follows the same principle of radical dissection of tumor as the open approach. Major drawback from endoscopic approach is bleeding from ICA which is difficult to control by endoscopic method. In a meta-analysis, covering 92 studies and a total 821 patients, Khoueir et al, calculated that mean blood loss from endoscopic approach was 564.21 ml as compare to our study it was 587 ml in endoscopic method. Complication rate was 9% with endoscopic method, hospital stay ranged from 2 to 8 days and 1 patient had residual out of 36 patients.¹⁸

Hackman et al, reviewed 31 cases of JNA at the university of Pittsburgh Medical Center from 1995 to 2006. They used pre-operative embolization in all their patients 24-48 hours before surgery, although we did not use the embolization procedure in our patients. Endoscopy was used by itself or in conjunction with open approach in 87% of time in our study it was 86% and plain open approach was used in 14% in Group B patients. In which he also noted that extended endoscopic approach provides un-paralled vision to skull base and middle cranial fossa which are sites of recurrence leading to complete tumor removal and lesser rates of complications, blood loss and operative time. He also supports staged resection for extensive tumors.¹⁵ In a review article Cloutier et al reported on 72 patients operated in 10 years concluded that skull base surgery progress lead to expansion of indications for JNA removal via endoscopic approach.¹⁷

CONCLUSION

From our study we can conclude that over the timespan of 18 years there has been significant shift in usage of surgical approaches for management of juvenile nasopharyngeal angiofibroma. From 2002-2010 29% patients were treated with endoscopic approach. This has risen to 78% of present time which can be combined with open approach in case of middle fossa invasion, optic nerve encasement, internal carotid artery encasement.

Retraction and manipulation of neurovascular structures, complications such as postoperative lacrimation, facial anaesthesia, trismus, pain, facial incision, removal of facial bones and plating is prevented with endoscopic approach endoscope provides multiangle vision and magnification, approach to inaccessible sites. Time of operation, duration of hospitalization is significantly less with endoscopic approach.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: The study was approved by the Institutional Ethics Committee

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Cite this article as: Vasani HH, Joshi CP, Patel KB. Trend analysis in management of juvenile nasopharyngeal angiofibroma: our institutional experience. *Int J Otorhinolaryngol Head Neck Surg* 2020;6:94-100.