

Original Research Article

Current trend of surgery in juvenile nasopharyngeal angiofibroma: a hospital based retro-prospective study

Sachin Jain¹, Dinesh Kumar², Shivendra P. Singh^{1*}, Ved P. Upadhyay¹,
Himani N. Singh¹, Abhishek K. Dubey¹

¹Department of ENT, Motilal Nehru Medical College, Prayagraj, Uttar Pradesh, India

²Department of ENT, Bundelkhand Medical College, Sagar, Madhya Pradesh, India

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*Correspondence:

Dr. Shivendra P. Singh,

E-mail: shivendrapratapsingh29@gmail.com

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ABSTRACT

Background: The aim of the present study was to know the current trends of surgery, to determine the surgery of choice according to stage and complications, rate of recurrence of following surgery.

Methods: Retro-prospective study was carried out from August 2004 to July 2014, in which 31 prospective and 8 retrospective cases were included. All the details of patients like general examination, ENT examination, computed tomography or magnetic resonance imaging scan finding, staging, and approach of surgery, complications, and recurrence rate were recorded.

Results: In our study all the patients were male and the mean age was 15.74 years (age ranged 11 to 22 years). Most frequent sign and symptoms were epistaxis (92.4%), nasal blockage (90%), and facial swelling (18%). Most common surgical approach was 'trans palatine approach' (51% cases) belongs to stage IIa, IIb, IIc, IIIa, IIIb and 'lateral rhinotomy approach' used in 35% cases. And by endoscopic approach (only 5.1% cases) were operated due to lack of embolization technique. Overall recurrence was about 29% (66% cases belonged to stage III). Postoperative complications were septal perforation 33%, palatal perforation 9.5%, facial disfiguration 9.5% and 9.5% hypertrophic scar.

Conclusions: Our study we concluded that, 92% of patients age group 11 to 20 years and all were males. Epistaxis and nasal obstruction were the most common symptoms. Most of the recurrent tumor belonged to stage III tumor. Most appropriate surgical approach for stage I, IIa and IIb was transpalatine approach and for stage IIc and IIIa was lateral rhinotomy. Complete excision of intracranial extension of tumor was difficult and causes recurrence of diseases.

Keywords: Transpalatine, Lateral rhinotomy, Embolization

INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is an uncommon benign vascular tumor occurring almost exclusively in pre-pubescent/pubescent males and occur in 2nd decade. JNA represents 0.05-0.5% of all head and neck tumors but is the common tumor of nasopharynx.^{1,2} Severe epistaxis accompanied by progressive nasal obstruction are the classical symptoms of juvenile

angiofibromas at the time of presentation. Other common symptoms are swelling of the cheek, hearing loss, dacrocystitis, rhinorrhea, hard and soft palate deformity, hyposmia or anosmia. Nasal endoscopy usually shows a large, lobulated mass behind the middle turbinate filling the choana with a smooth surface and clear signs of hypervascularization. Tumors are staged according to computed tomography (CT) scan finding. There are variety of staging criterias develop, which include those

developed by Radkowski, Chandler, Fisch, Andrews, Onerci and sessions.²⁻⁵ The primary treatment modality is surgical excision.⁶ Various surgical approaches are transpalatal approach, lateral rhinotomy, midfacial degloving, transfacial transmaxillary, medial maxillectomy, endoscopic approach, combined endoscopic and external approach.

Radiation therapy originally reserved for unrespectable or life-threatening tumors, radiotherapy is an alternative treatment modality in managing JNA with dose ranges between 30-46 Gy and just as effective as surgery with a 15% recurrence rate.⁷⁻⁹ Chemotherapy has been investigated as treatment option in cases of JNA that had recurred after surgery and radiotherapy. Other treatment modalities that have been reportedly implemented to address JNA include coblation, cryotherapy, electrocoagulation, gamma knife, harmonic scalpel, interstitial brachytherapy, potassium titanil phosphate-laser embolization and sclerotherapy.⁹⁻¹⁵ Recurrence is by far the most common complication encountered and is reported in up to 25% of patients regardless of the method of treatment and recurrence is more likely in patients with advanced disease and in those treated by inexperienced surgeons.

The objective of the study was to know the current trends of surgery in our institute, to determine the surgery of choice according to stage and complications related to surgery and the rate of recurrence of following surgery.

METHODS

This study was retro-prospective and carried out in Department of Otorhinolaryngology, S.R.N. Hospital, M.L.N. Medical College, Allahabad. This study included total 39 patients. Retrospective cases were from August 2004 to July 2013 which contains 31 patients and patients were called for follow up visit by letters and telephones. Prospective cases were from August 2013 to August 2014 which contains 8 patients.

Exclusion criteria

Cases who did not respond to the letter or phone calls and did not come for follow up, patients who were not willing to give their consent.

Inclusion criteria

Retrospective study

All the patients diagnosed and operated for JNA from August 2004 to July 2013 were called by letters or phones. Patients who responded to letters or phones, gave their consent and came for regular follow up.

Prospective study

All the patient from August 2013 to August 2014 who were diagnosed and/or operated for JNA in SRN Hospital and gave their consent.

For retrospective cases name, age, sex, religion, socio-economic-status, address, phone no, chief complain, history of past illness, family history, personal history, general examination, ENT examination, CT or magnetic resonance imaging scan finding, date of operation, staging, and type of surgery were recorded from the case sheets. All the retrospective cases were examined at their first visit and then examined monthly for three consecutive months.

Patient's new complains, the traditional nasal examination and nasal endoscopy were done at every follow up visit.

In patients whom recurrence was seen and/or operated for recurrence were examined monthly till the end of the study.

For prospective cases, detailed medical history including name, age, sex, chief complain etc., were recorded, and then general examination and head and neck examination were done at the time of admission. Special emphasis was given to nasal examination which include traditional nasal examination with head mirror, bull's eye lamp, Killian's nasal speculum, tongue depressor, posterior rhinoscopy mirror, nasal endoscopy by 0, 25, 70 degree nasal endoscope. A pre-op CT scan was advised in all patients.

Patients were watched during their post op hospital stay for any kind of post op complication and were advised a post op CT scan. All the patients were re-examined monthly till the end of this study. In our study we used Radkowski classification.²

Table 1: Radkowski classification of JNA.²

Characteristics	
Ia	Limited to nasopharynx and nose area.
Ib	Extension into one or more sinuses.
IIa	Minimal extension into pterygopalatine fossa.
IIb	Occupation of pterygopalatine fossa without orbital extension.
IIc	Infratemporal fossa extension without cheek or pterygoid plate involvement.
IIIa	Erosion of skull base (middle cranial fossa or pterygoid).
IIIb	Erosion of skull base with intracranial involvement.

RESULTS

Total number of patients included in the study was 39 over a period of 10 years from August 2004 to August 2014. Minimum age was 12 years and maximum age was 21 years and 92% of patients belong to age group 11 to 20 years. We found that all the patients (100%) were male.

Table 2: Comparison of symptoms in percentage with no of patients.

S. no.	Sign/symptoms	No. of patients (n=39)	%
1	Epistaxis	36	92.4
2	Nasal obstruction	35	89.7
3	Nasal mass	4	10.2
4	Facial swelling	7	18
5	anosmia	2	5.1
6	Change of voice	6	15.38

Table 2 shows that 92.4% of the patient of JNA had complain of epistaxis, 89.7% had complain of nasal obstruction, 18% had facial swelling, 10.2% had visible nasal mass, 5.1% complained of anosmia and 15.38% had complain of change in voice.

Table 3: Distribution of patients according to Radkowski's staging in JNA patients (n=38).

S. no.	Stage	No. of patients*	%
1	Ia	3	7.8
2	Ib	3	7.8
3	IIa	4	10.5
4	IIb	7	18.4
5	IIC	2	5.2
6	IIIa	15	39.4
7	IIIb	4	10.5

*CT scan of one patient was missing and he was not included in this table.

Table 7: Recurrence according to staging and type of surgery.

S. no.	Radkowski's	No. of patients (n=20)*	Type of surgery	Result
1	Ia	2	TP	No recurrence
2	Ib	2	TP	No recurrence
3	IIa	1+	LR+MM	No recurrence
		1	TP	
4	IIb	1+	TP	Recurrence
		1+	TP	No recurrence
		1+	TP	No recurrence
		1	Endoscopic	No recurrence
5	IIC	1	LR	No recurrence
6	IIIa	6	LR	One residual
7	IIIb	1+	TP	Residual
		1+	LR	Recurrence
		1	LR+MM	Recurrence

*CT scan of one patient was missing and he was not included in this table however he showed recurrence. TP= Transpalatine; LR= Lateral rhinotomy; MM= Medial maxillectomy.

Table 5 and 6 shows that most common approach in both prospective and retrospective cases was transpalatal followed by lateral rhinotomy.

Table 4: Distribution of patients according to type of surgery done in JNA patients (n=39).

S. no.	Type of surgery	No. of patients	%
1	TP	20	51.28
2	LR	14	35.89
3	LR with MM	2	5.1
4	Endoscopic	2	5.1
5	LR with maxillary swing	1	2.5

TP= Transpalatine; LR= Lateral rhinotomy; MM= Medial maxillectomy.

Table 5: Type of surgery done in prospective cases.

S. no.	Type of surgery	No. of cases (n=8)	%
1	TP	4	50
2	LR	2	25
3	Endoscopic excision	1	12.5
4	LR+MM	1	12.5

TP= Transpalatine; LR= Lateral rhinotomy; MM= Medial maxillectomy.

Table 6: Type of surgery done in retrospective cases.

S. no.	Type of surgery	No. of cases (n=31)	%
1	TP	16	51.61
2	LR	12	38
3	LR+TM	1	3.2
4	Endoscopic	1	3.2
5	LM+maxillary swing	1	3.2

TP= Transpalatine; LR= Lateral rhinotomy; TM= Total maxillectomy.

Table 7 shows that all the patient of stage I was operated by transpalatine route without any recurrence. 1 patient of stage IIa was operated by lateral rhinotomy with medial maxillectomy approach and 1 patient by transpalatine route. Both the patients did not report any recurrence. 3 patients of stage IIb were operated by transpalatine approach of these only one reported recurrence. One patient (stage IIb) was operated by endoscopic approach

without any recurrence. 6 patients of stage IIIa were operated by lateral rhinotomy approach out of which one was left with residual tumor. 1 patient of stage IIIb was operated by transpalatine route one patient by lateral rhinotomy approach and one by lateral rhinotomy with medial maxillectomy approach. All the patients in stage IIIb showed residual or recurrent tumor regardless of this route of surgery.

Table 8: Time taken by the tumor to recur after previous surgery.

S. no.	Time taken by the tumor to recur after previous surgery	No of patients (n=10)*	%
1	0-6 month	2	20
2	7-12 months	5	50
3	13-18 months	1	10
4	More than 1.5 years	2	20

Table 8 shows that recurrence was seen within 1 year in 70% of the cases and in 20% case it was seen after 1.5 years.

Recurrence was seen in only 6 patients but total no. of recurrences was 10 because one patient had 2 recurrences and one patient had 4 recurrences.

Table 9: Complications related to surgery of JNA.

S. no.	Complications	No. of cases (n=21)	%
1	Septal perforation	7	33
2	Permanent palatal perforation	2	9.5
3	Facial disfiguration	2	9.5
4	Hypertrophic scar	2	9.5

Table 9 shows that out of total patients operated for JNA, 33% suffered from septal perforation, 9.5% from permanent palatal perforation, 9.5% from facial disfiguration, 9.5% from hypertrophic scar.

DISCUSSION

JNA is a disease of young men. In our study all the patients were male and the mean age at the time of diagnosis was 15.74 years and the age ranged between 11-22 years. In the present study most of the patients (92%) belonged to age group of 11-20 years. Almost similar data was published in by Lara et al.¹⁶ Our study showed that all the patients were male. Genetic studies have demonstrated a close relation between JNA and androgen receptor expression, suggesting that this tumor is androgen dependent. This is the reason of male predominance and early age of onset between 11-22 years. Similar result were found in various studies.^{5,17-19} Most frequent sign and symptoms in our study were epistaxis (92.4%), nasal blockage (90%), and facial swelling (18%). Tang et al also found similar triad of symptoms with little difference in frequencies.²⁰

Midilli et al found in their study of “analysis of 42 cases and important aspects of endoscopic approach” that about half of the patients belonged to stage IIA to IIC.²¹ Twelve patients underwent endoscopic removal of the tumor (28.5%). Lateral rhinotomy was used for 10 patients (23.8%), degloving approach for 7 patients, transpalatal approach for 6 patients (14%), combined approach for another 6 patients and finally midfacial splitting for 1 patient.

In our study 34% cases belonged to stage IIa to IIC and 50% to stage IIIa and IIIb. Most common surgical approach was ‘trans palatine approach’ used in 51% cases and ‘lateral rhinotomy approach’ used in 35% cases. Only 5.1% cases were operated by endoscopic approach.

High incidence of stage III tumors in our study may be due to delay in making the diagnosis of JNA which led to enlargement and intracranial extension of tumors. Endoscopic approach was not used very frequently in our setup because of lack of embolization facility.

Our study showed that in last 10 years the most common approach in our institute was trans palatine and lateral rhinotomy. In retrospective cases, these routes of surgery were employed in 75% cases (TP=50%, LR=25%) and in prospective cases it was seen in 90% cases (TP=52%, LR=38).

Herman et al in their study found that the skull base was invaded in two-thirds of the patients (66%), and the rate of recurrence was 27.5%.²² Extensions to the infratemporal fossa, sphenoid sinus, base of pterygoids and clivus, the cavernous sinus (medial), foramen lacerum, and anterior fossa were correlated with more frequent recurrence. This was the reason of high recurrence in stage III. A series of 33 patients had been reviewed retrospectively by Combe et al.²³ All the patients in this study were treated by simple or extended lateral rhinotomy as a primary or secondary procedure. The final long term disease control rate was 97% but during the treatment period the overall symptomatic

recurrence rate was 50%. However, amongst those treated primarily, the recurrence rate was 34%.

Leong included 72 patients from 15 studies in a review.²⁴ The mean follow-up period was 47 months. Most patients had a craniofacial procedure. Recurrence was reported in 13 patients (18%), which were detected between 7 and 26 months during the follow-up period. Overall, 86% of the cohort was free of disease.

Our study showed that overall recurrence was about 29% and out of total recurrence, 66% cases belonged to stage III. 44.4% cases that originally belonged to stage III had recurrence.

Finding in our study was corroborated by Combe et al and Herman et al. However Leong found only 18% recurrence in his study. Herman also noticed that patient who initially had intracranial extension (stage III) reported high recurrence rate later (as in our study).

Our study shows that recurrences were seen within 6 month in 20% of the cases, 60% cases reported between 7-18 months and in 20% case it was seen after 1.5 years. Lenog also found in his study that most of the recurrence was seen within 7-26 month.

Hosseini et al in their study 'analyzed thirty-seven patients of JNA and found that among different surgical approaches used, the transpalatal resulted in 1 recurrence out of 14 patients treated with this approach when the lesion was limited to the nasal cavity, nasopharynx and paranasal sinuses (stage I).²⁵ No recurrence was observed with the use of this approach with lesions with minimal extension to the pterygopalatine fossa (stage IIA). But among three patients with intracranial extension who were treated with this approach, two resulted in symptomatic recurrence; however, using the Lefort I surgical technique, no evidence of recurrence was observed in the two patients in stage III who were treated with this approach. Involvement of the orbit, middle cranial fossa and base of the pterygoid by the primary JNA results in a higher incident of recurrent tumor. Among different surgical techniques, the lowest recurrence rate is seen either in the transpalatal approach when the tumor is limited to the nasopharynx with extension to the nasal cavity or paranasal sinuses or with the Lefort approach when skull base invasion is present.

Our study also supported the Hosseini et al study and showed that transpalatine approach was most appropriate for stage I, IIa and IIb tumors, reporting only one recurrence out of total 6 cases. Lateral rhinotomy approach provides good exposure and ensures complete removal of tumors in stage IIc and IIIa, as we had only one recurrence out of 7 cases. Transpalatine or lateral rhinotomy alone or with medial maxillectomy approach was not sufficient for stage IIIb tumors.

Our study also reported complications related to surgery

for JNA. 33% patients suffered from septal perforation, 9.5% from permanent palatal perforation, 9.5% from facial disfigurement and 9.5% from hypertrophic scar. No data regarding surgical complication are available.

Our study showed that 43.5% patients required at least 2 units of whole blood for surgery. 39 patients' required total 79 units of blood which means on an average 2.02 units of blood were required for every patient. Similar data was reported by Tuna et al who found that the intra-operative transfusion requirement was 2.3 units.²⁶

Our study also showed that blood requirement for JNA surgery was maximum for stage III. About 3 units of bloods were required per patient for stage IIIb and about 2.4 units of bloods were required for stage IIIa.

CONCLUSION

Our study we concluded that, most of the patient (92%) belonged between age group 11 to 20 years and all the patients were males. Epistaxis and nasal obstruction were the most common symptoms associated with JNA. About 50 % cases belonged to stage III (intracranial extension) at the time of diagnosis. About 51% cases were operated by transpalatal route, and 35% cases by lateral rhinotomy. Most of the recurrent tumor belonged to stage III tumor. Most appropriate surgical approach for stage I, IIa and IIb was transpalatine approach. Lateral rhinotomy approach was best suited for tumors of stage IIc and IIIa. Complete excision of intracranial extension of tumor was difficult and causes recurrence of diseases.

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

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