JUVENILE NASOPHARYNGEAL ANGIOFIBROMA - THREE YEARS EXPERIENCE

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ABSTRACT : *Juvenile nasopharyngeal angiofibroma is a rare, highly vascular, histologically benign but locally invasive tumour, affecting almost exclusively adolescent males. This is a prospective study of 37 patients with juvenile nasopharyngeal angiofibroma treated during 3 years in our institution. The various pharyngeal and extrapharyngeal presentation staging and proper surgical approaches as per extensions are discussed here with reference to past reports.*

Key Words: *Juvenile nasopharyngeal angiofibroma, pharyngeal and extrapharyngeal - presentation, staging & surgical approach.*

INTRODUCTION

Juvenile Nasopharyngeal Angiofibroma (JNA) is a highly vascular neoplasm almost exclusively affecting adolescent males. Although histologically benign, its biological behaviour is very aggressive and destructive. Clearly, these tumours are not limited to the nasopharynx and usually have extension, for example, into the nose, paranasal sinuses, orbit, pterygomaxillary, infratemporal fossa and cranial cavity. Thus the term "nasopharyngeal" angiofibroma is misleading and inaccurate. The term "Juvenile" may be unnecessary in view of the fact that the neoplasm does occur in older patients too.

JNA accounts for 0.05% of all neoplasms of the head and neck, and it is generally accepted that its incidence is relatively higher in India than in US and Europe.

Epistaxis and nasal obstructions are the most common presentation, but various extrapharyngeal extension of the tumour produce features like rhinolalia, palatal bulging, proptosis, swelling of cheek, dysphagia etc.

Surgery has been the overwhelming choice for the primary treatment of JNA and the selection of proper approaches depend primarily upon the extension of the tumour. C.T. Scan and proper staging of the tumour play a key role in this regard. Recent advances in skull base surgery and improvement of diagnostic and invasive radiography have yielded high cure rates, low recurrence rates, minimal morbidity and mortality.

In the present study we have tried to evaluate the various clinical presentations of this tumour. The various surgical

approaches, which we have employed to deal with this tumour as per their extensions and staging, have also been reported and evaluated.

MATERIALS AND METHODS

The study was carried out in the Department of Otolaryngology, Medical College, Calcutta between June 1998 and May 2001. 37 patients were treated during this period and all were male in the age group 9 to 27 years.

Each patient was subjected to a thorough clinical examination for evaluation of different pharyngeal and extrapharyngeal presentations. Epistaxis and nasal obstructions were found to be the complaints occurring in most patients. Bulging of cheek, rhinolalia, nasal discharge were the other common complaints. The duration of these symptoms varied from 4 months to 8 years. A battery of investigation like conventional X-ray, nasal endoscopy, C.T. scan and angiography (selected

Fig. 1 : Clinical photograph of a patient of J.N.A. showing palatal bulge.

Table - I : Staging of J.N.A. (Chandler et al, 1984)

Stage I	Tumour confined to the nasopharynx.
Stage II	Tumour extending into the nasal cavity and/or sphenoid sinus.
Stage III	Tumour extending into one or more of the following: antrum, ethmoid sinus, pterygomaxillary and infratemporal fossae, orbit and/or cheek.
Stage IV	Tumour extending intracranially.

Table II : Distribution of Patients According to Clinical Presentations

Symptoms and signs	No. of patients	Percentage
Epistaxis	37	100.0
Nasal Obstruction	32	86.5
Nasal Discharge	12	32.43
Rhinolalia	21	56.75
Obligatory mouth breathing	11	29.72
Headache	9	24.32
Hyposmia	4	10.2
Otalgia	2	5.40
Dysphagia	3	8.10
Blindness	1	2.70
Nasopharyngeal mass	37	100.0
Nasal mass	24	64.86
Bulged soft palate	11	29.72
Cheek swelling	7	18.91
Proptosis	5	13.51
Conductive deafness	14	37.83
Extension of mass in oropharynx	3	8.10
Swelling in the temp region	2	5.40

cases only) were done to confirm the diagnosis. Till date a surgical extirpation of the tumour is the mainstay of treatment. So, a proper approach was selected as per extension of the tumour in each case. Mainly lateral rhinotomy and transpalatal approaches with their various modifications and combinations were used in most cases and the mass was sent for histopathological examination for a final diagnosis. The patients were followed up thereafter at regular intervals by nasal endoscopy and CT scan (if necessary).

RESULTS AND ANALYSIS

In the past three years 37 patients were diagnosed with

Fig. II : Soft tissue X-ray Nasopharynx in a case of J.N.A. showing nasopharyngeal mass.

Fig. III : CT scan of a case of J.N.A. showing nasal and nasopharyngeal mass with antral extension.

this tumour by one or more of the following criteria - clinical presentations, characteristic radiological and endoscopic appearances and postoperative histopathological examination. Surgical removal of the tumour was done in all cases employing different approaches as per the extensions and the results thus yielded have been presented below.

All the patients were exclusively male. The incidence of JNA in the present series was calculated to be 1 per 2340.52 OPD and Emergency cases.

Epistaxis (100%) and nasal obstruction (86.5%) were the most common symptoms in our series. Rhinolalia (56.75%), nasal discharge (32.43%), obligatory mouth breathing (29.72%) and headache were the others symptoms. Cases complaining of hyposmia (10.82%), otalgia (5.40%), dysphagia (8.10%) and blindness (2.70%)

Fig. IV : CT scan of a case of J.N.A. showing orbital extension with proptosis.

Fig. V : CT scan of a case of J.N.A. showing intracranial extension.

were also found.

Among the signs, a nasopharyngeal mass was seen in all the cases. Nasal mass was the next common sign (64.86%). Conductive deafness, palatal bulge and cheek swelling were noted in 32.13%, 29.72% and 18.91% cases respectively. Proptosis and extension of the mass down to the oropharynx were also noted. Some rare signs in the present series included swelling in the temporal region and blindness, seen in 2 cases and 1 case respectively.

The patients were staged according to Chandler et al (1984) classification given in table No. I. In the present series we did not find any stage I lesion. Only 9 cases of stage II lesions have been treated. On the other hand stage III tumours contributed the majority of the cases (23 out of 37) and Stage IV tumours were the second least common (5 out of 37).

Table III : Distribution of Patients According to Stage of Tumor and Surgical Approaches Employed for Removal

Stage	No. of patients	Approaches	Recurr- ence
I	0	-	-
II	9	<ul style="list-style-type: none"> • Lat Rhinotomy (2) • Transpalatal (6) • Retracting the soft palate (1) (Small recurrent tumour) 	2
III	23	<ul style="list-style-type: none"> • Lat. Rhinotomy (9) • Transpalatal (2) • Weber Fergusson incision with sublabial extension followed by Medial maxillectomy (7) • Weber Fergusson incision with sublabial extension and medial maxillectomy(1) + Transpalatal incision • Transpalatal incision + Sublabial extension (4) 	5
IV	5	<ul style="list-style-type: none"> • Weber Fergusson incision with sublabial extension and medial maxillectomy (1) +Transpalatal incision • Extended Lateral Rhinotomy (Lip splitting) and Medial Maxillectomy (2) +Sublabial extension • Combined Intracranial-extracranial approach (2) 	2

Lateral rhinotomy was the most common approach (24.72%) chosen in our series followed by transpalatal approach (22.62%). Various combinations were employed in 17 cases to deal with the extensions.

Stage II tumours in our series were mostly removed by transpalatal approach (6 out of 9) and Stage III tumours mostly required lateral rhinotomy (9 out of 23) and other combined approaches. 3 of our stage IV tumours were managed by removing the extracranial part employing the extensive approaches mentioned in the table No. III.

Fig. VI: Photograph of a specimen of extensive J.N.A. removed by Weber-Fergusson incision.

Fig. VII : Clinical photograph of a postoperative case of J.N.A. (removed by transpalatal approach) showing palatal fistula.

On an average we required 2.08 units of blood per patient, during the operation. But the amount may vary according to the patient profile, bleeding potential of the tumour and operative approach.

Main postoperative complications in our series were nasal crusting (7), nasal deformity (flattening of ala) (3), cheek numbness (4), palatal fistula (3), epiphora (2) and ectropion (2). The last two complications were the result of Weber-Fergusson incision. Out of the 3 cases of palatal fistula, two healed subsequently and one was repaired by palatal flap.

DISCUSSION

The tumour incidence varies from rare (i.e. 1 in 4000 – 7000, Harma, 1958) to very rare (1 in 50,000, Batsakis JG, 1979). But, it is relatively commoner in contemporary Indian studies (Bhatia et al, 1967, Gupta and Gupta, 1971

and Patil 1982). In our study, the incidence rate was 1 per 2340.52 otorhinolaryngological cases. The incidence seems to be quite high, but it may be due to Berksonian bias, as our hospital is a major referral center in the state of West Bengal.

The tumour predominantly affects male adolescents. The age range varies in different studies: 7 - 21 years (average 14 years) in Briant TDR et al (1978) study, 7-29 years (median 15 years) in Bremer JW et al (1986) study. In our study the age range was 9-27 years with the mean of 15.9 years.

The triad of nasal obstruction, a nasopharyngeal mass and recurrent epistaxis may result from multiple conditions, but their presence in an adolescent male almost always indicate the presence of an angiofibroma (Bremer JW, Neel HB, III, et al, 1986). In our series, epistaxis and nasopharyngeal mass were present in all the 37 cases and nasal obstruction was present in 32 (86.5%) cases. Rhinolalia and nasal mass were the most common presentations probably due to extra-nasopharyngeal extension of the tumour.

Among the investigations, CT scan of nose, nasopharynx and paranasal sinuses is considered an integral part of the pre-operative diagnostic evaluation (Bremer JW et al, 1986). So, CT scan (both plain and with contrast) was done in each and every patient.

Surgical resection is considered the treatment of choice for this tumour, reserving radiation therapy for unresectable lesions. Among the other adjunctive treatments - hormones like androgens and estrogens; chemotherapeutic agents like doxorubicin hydrochloride & dacarbazine and embolisation of the feeding vessels from external carotid artery has shown some role in some of the studies.

The choice of best surgical approach is the most critical decision in the care of a patient of nasopharyngeal angiofibroma. Stage I and II are approached through a transpalatal or lateral rhinotomy procedure. The transpalatal procedure is used for smaller lesions and it avoids a facial incision, but it provides a limited exposure. The lateral rhinotomy approach provides good exposure of nasal cavity, paranasal sinuses and pterygopalatine fossa; but exposure beyond these regions are limited and direct visualization of the ICA can not be maintained during all stages of the resection. In the present series, stage II Angiofibroma was removed via transpalatal approach.

The major problem in the evaluation and treatment of stage III and stage IV tumours is that these tumours can spread into the paranasal sinuses, pertinent cranial nerves, Eustachian tube, ICA, skull base, and in some cases into the intracranium and cavernous sinus; so stage III or IV nasopharyngeal angiofibromas require a surgical procedure that will encompass the tumour and allow for full exposure of the above mentioned areas. There is no unified opinion on how to treat these tumours; however, the infratemporal fossa approach, type C, has the advantage of all these qualities and will facilitate complete visualization of the ICA during tumour manipulation. Additionally, the use of operating microscope allows for greater precision in tumour resection and safety in manipulating the vital structures mentioned above (Fisch U et al, 1989). The sequelae of infratemporal fossa approach are acceptable. The only permanent dysfunction is a conductive hearing loss. The only complication directly related to infratemporal fossa approach consisted of wound infection in a very rare instance (less than 1 percent). Sardana (1965) has used this approach combined with transpalatal approach when the JNA extends laterally through pterygomaxillary fissure. In the present series, lateral rhinotomy was mainly chosen to deal with these tumours and for greater extensions more radical approaches like Weber-Fergusson incision and various combinations were undertaken. Here, in the case of extensive stage III tumours, transmaxillary approach following Weber-Fergusson incision gave a very good exposure and the tumours, which we had removed employing this approach did not recur. In some cases the help of endoscope was also taken to remove the extension of the tumour in the ethmoids and sphenoid sinuses and endoscopic excision of JNA was also tried in limited number of cases.

Krekorian et al (1969, 1977 & 1979), Standefer J et al (1983) and Gill et al (1976) have recommended the use of a combined "intra-extracranial approach" to nasopharyngeal angiofibromas with intracranial extension. This requires the use of a fronto-temporal craniotomy with intradural inspection and resection of the tumour if present in this region. Following the craniotomy, resection of the extracranial part of the tumour is performed. Craniotomy is combined with lateral rhinotomy or transpalatal dissection; but with the difficulty of prolonged operation time and increased chance of infection & CSF leak. Total maxillectomy approach can also be used, but it results in extreme debility. A major benefit of infratemporal fossa approach is the ability to safely manage intracranial

tumour extension. The use of microsurgical techniques is the prerequisite for safe tumour removal. Taneja et al (1997) advocated staged surgical treatment for such cases while Tandon et al (1988) utilised craniotomy-rhinotomy with modified Weber-Fergusson incision. Removing the extracranial part of the tumour and then leaving the small intracranial part to undergo spontaneous involution have managed the 3 cases of stage IV tumours in the present series. Extended Weber-Fergusson incision with transpalatal and sublabial extensions was employed in one case and Lateral rhinotomy with sublabial extension was chosen in 2 other cases.

There is limited need for radiation therapy with the use of the infratemporal fossa approach in extensive nasopharyngeal angiofibromas. The role of radiotherapy should be limited to patients with Stage IV lesions infiltrating the cavernous sinus (Fisch et al, 1989). If radiation is unsuccessful in arresting tumour growth, a neurosurgical procedure would be required.

'Recurrent' lesions should be more properly defined as persistent disease resulting from an incomplete previous resection, which can be proved by histopathological investigations. Complete surgical removal is possible in almost all cases if an adequate approach and microsurgical techniques are used (Fisch et al, 1989). Regarding the recurrence different studies have reported different figures, ranging from 6% or less (Fisch et al 1989) to 20% or more (Jones GC et al, 1986). But the high rate of recurrence (50% or so) of stage IV tumours was universally agreed. In the present series, 2 cases of recurrence (40%) was detected out of 5 stage IV tumours so far treated, and the overall recurrence rate was 24.4%.

CONCLUSION

Juvenile nasopharyngeal angiofibroma is a very rare benign tumour of nasopharynx that may, because of its strategic anatomic location impinge on several vital structures of skull base and invite a challenge to the Otolaryngologists. The extensive study and analysis of this large series of patients with JNA along with a review and comparison of the reports in the literature have allowed us to draw the following conclusions :-

- the tumour almost exclusively affects adolescent male
- symptoms like epistaxis and nasal obstruction can be regarded as almost universal.
- the size of the tumour in the nasopharynx does not necessarily represent the true extent and size of the

- tumour. It may be only" the tip of the iceberg".
- Extra-nasopharyngeal extensions are very common and were seen in all the cases - nasal extension being commonest. They are mainly responsible for recurrence.
 - Contrast CT scanning is pathognomonic of angiofibroma and permits accurate staging which assists in planning for the surgical approach estimating prognosis, and reporting results.
 - Surgery should be tailored according to the extent of the tumours with emphasis on wider exposure for better tumour clearance and avoiding piecemeal removal, all leading to fewer recurrences.

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