ORIGINAL ARTICLE

# Retrospective stuty of 356 cases of nasopharyngeal angiofibroma reported at Jinnah Postgraduate Medical Centre (JPMC) Karachi

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**Abstract**

**Objective: To determine the incidence and management of Nasopharyngeal angiofibroma at JPMC, Karachi from 1981-2011.**

**Design: Descriptive, Retrospective study.**

**Subjects and method: The study was conducted at JPMC, from 1981-2011(30 years). The data includes age of patients, gender, clinical presentation, staging, mode of treatment, status of pre-op embolization, surgical procedures performed and recurrence rate.**

**Results: Total number of cases were 356. Males were, 352 and females were 04. Minimum age was 13 years and maximum was 36 years and median age was 17.5 years. According to stage they were 39% Stage II, were 31% stage III, 08% stage IVa and were 02% stage IVb. Geographically, 48% were from hot/desert areas and 52%were from cold/high altitude areas. Commonest symptom was nasal obstruction, presnet in 98% patients and in 75% it was unilateral. Epistaxis was present in 70%. Surgical excision was performed in 349 cases while radiotherapy was done in 07 patients of stage IVb. Embolization was done in 278 patients while 78 patients were not embolized due to non availability of the facility earlier. Surgical approach commonly adopted was lateral rhinotomy in 277 patients. Midfacial degloving was performed in 30 patients, endoscopic excision was done in 13 patients and transpalatal rout was chosen in 29 patients. Recurrence was seen in 7.8% (n=28) cases. Follow-up was done after 03 months and 06 months and every year for the next 05 years.**

**Conclusion: This is a common vascular tumour in Pakistan. Reported from both hot and cold areas. Nasopharyngeal angiofibroma is rare in females but 04 were reported and their genetic analysis was done. Before preoperative embolization era, recurrence was uncommon but more blood transfusion was required. After preoperative embolization the recurrence has increased but blood transfusion has reduced. Earlier patients presented in late stage but now they present usually by stage II & III.**

**Keywords: nasopharyngeal angiofibroma, lateral rhinotomy**

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**Introduction:**

Among head and neck tumours Juvenile Nasopharyngeal Angiofibroma (JNA) accounts for 0.5% of all tumours [1]. It is a benign tumour [2]. It is non-encapsulated and composed of vascular tissue and fibrous stroma [3]. It principally affects young and adolescent males, females are affected very rarely. There are certain theories which support the etiology of nasopharyngeal angiofibroma including congenital cell rest cell rest theory, hormonal theory, hemartomatous theory, persistent stapedial artery theory, myoepithelial theory, chemodectoma and vascular theory [4]. The nasopharyngeal angiofibromas usually arise from sphenopalatine foramina [5]. It can extend into the nasal cavity, maxillary, ethmoid and sphenoid sinuses, pterygiod fossa, infratemporal fossa, cavernous sinus, eyes, pituitary fossa and optic nerve. Common presentation is nasal obstruction and epistaxis [6]. Other symptoms and signs are headache, rhinolalia clausa, ear blockage, serous otitis media, broadening of nasal

bridge, proptosis, swelling of cheek and involvement of II, III, IV and VI cranial nerves[7].

Accordingly to Fisch staging system: Stage 1 is tumour limited to nasopharynx. State II is tumours extension to pterygopalatine fossa and para nasal sinuses. Stage III is extension into infratemporal fossa and orbit (Stage IIIa is without intracranial extension and IIIb is intracranial but extradural involvement). Stage IVa is intracranial and intradural only while IVb is intracranial and intradural with infiltration of cavernous sinus, pituitary fossa or optic chiasma [8,9]. Preoperative embolization has been a controversial issue. Internal maxillary artery embolization has reduced blood loss by over 50% but at the same time recurrence rates have been increased due to the shrinkage of tumour [3].

Table 1: *Demographic data*

| Variable | Value |
|---|---|
| Gender Distribution | |
| Male | 352 |
| Female | 4 |
| Age Distribution | |
| Minimum | 13 years |
| Maximum | 36 years |
| Mean | 17.5 years |
| Geographical Distribution | |
| Hot / Desert Areas | 48% |
| Cold / Mountain Areas | 52% |

Table 2: *Presentation of Juvenile Nasopharyngeal Angiofibroma*

| | |
|---|---|
| **According to stage** | |
| Stage I | 20% |
| Stage II | 39% |
| Stage II | 31% |
| Stage IV a | 08% |
| Stage IV b | 02% |
| **According To Symptoms** | |
| Nasal obstruction | 98% |
| Unilateral | 75% |
| Bilateral | 23% |
| Epistaxis | 70% |
| Cheek Swelling | 30% |
| Proptosis | 27% |
| Palatal Buldge | 18% |

Table 3: *Treatment modalities*

| | |
|---|---|
| Treatment mode | |
| Surgery | 349 |
| Radiotherapy | 07 |
| Pre operative Emborlization | 278 |
| Non Embolosed | 78 |
| **Surgical approaches** | |
| Lateral Rhinotomy | 277 |
| Midfacial Degloving | 30 |
| Transprlatal | 29 |
| Endoscopic | 13 |

The main treatment modality is surgical excision via, transpalatine, sublabial approach, extended lateral rhinotomy, mid facial degloving approach, extended Denkers approach, infratemporal and endoscopic approaches. Other treatment modalities are internal maxillary artery ligation, hormonal therapy for recurrent tumours and low dose radiotherapy for stage IVb tumours [7]. Recurrence up to 34% has been documented [10].

**Subject and methods:**

This retrospective study included a total number of 356 cases of Nasopharyngeal Angiofibroma collected from the Department of Ear, Nose and Throat-Head and Neck Surgery, Jinnah Postgraduate Medical Centre, Karachi from 1981-2011. The information regarding the cases was recorded on a proforma and the data included age of patients, gender, clinical presentation, staging of Nasopharyngeal Angiofibroma, whether embolization was done or not preoperatively, mode of treatment, surgical procedures performed and recurrence rate.

**Results:**

A total number of 356 cases were included in this study, majority were males (352) and only 04 were females. Mean age was 17.5 years. The demographic details are depicted in Table No.1. Jointly seventy percent of patients were in stage II and III at the time of presentation and nasal obstruction was the most common symptom (Table No.2). Majority of the patients were treated surgically but 07 patients who were in stage IVb were subjected to radiotherapy. Lateral rhinotomy was found to be most convenient and effective approach for tumor excision although some other surgical techniques were also adopted (Table No.3).

Overall recurrence was seen in 7.8% (n=28). Out of 78 non embolized patients 54 had a regu-

lar followup from 1-5 year and 03(5.5%) of them showed recurrence while out of 278 pre embolized patients 215 were followed up regularly for around a period of 1-5 year and 23 (10.69%) of them were presented again with recurrence. Two of the seven patients who were sent for radiotherapy reported with recurrence / residual tumor and five were lost to follow up.

**Discussion:**

This tumor was first described by Hippocrates in 5th century BC, but the term "Angiofibroma" was coined by Friedberg for the first time in 1940. Although the correct statistics are not available but in contrast to international literature it is not a very rare tumor in our country. The rate of incidence per year in our study is 11.86 patients per year at one centre. In another local study it is described as 12.5 cases per years [11]. A study conducted in Peshawar it was found out to be ten patients per year [12]. The highly vascular tumor is predominantly found in adolescent males. Genetic studies demonstrated a close relationship between these tumors and androgens receptor expression which indicates that these tumors are androgen dependent and thus to some extent explains its high prevalence rate in males. Juvenile Nasopharyngeal Angiofibroma (JNA) usually occurs between the age of 14-25 years in males although extremely rarely the tumor has been reported among females [13].In our series we have also come across 04 female patients. Fisch classification is commonly used for staging of JNA. Mostly the patients report in stage II or III, as in our series 70% of the patients were in stage II or III when they were reported. Other local studies have the same finding regarding stage of presentation [14]. Altitude and weather did not seem to have much effect on the incidence of disease as in our study 48% of patients were from desert area of Sindh and Baluchistan where the mereury hits more than $40^0$ Celsius in summers whereas around 52% of our patients belong to the areas of high altitude and cold weather. We could not found any study indicating geographical difference in prevalence of JNA.

Nasal obstruction is present in almost all patients as it is mentioned in many studies describing the presenting symptoms of JNA. Around 70% of patients had epistaxis and often an episode of severe nose bleed compelled them for consultation. Facial swelling, unilateral proptosis and palatal bulge were among the other symptoms. The presenting symptoms are usually universal and without much difference in frequency among them [15].

Diagnosis is made on a high suspicion index on clinical examination and then confirmed by contrast CT Scans which is the main diagnostic tool in cases of JNA. Occasionaly MRI are used to know the extent of angiofibroma intracranialy. All these tools are helpful in the staging of the tumor and provide a roadmap for further planning of ultimate surgical excision[16].

Surgery remains the main stay of treatment for definitive cure of JNA. A variety of approaches have been adopted by different surgeons according to the stage of disease, expertise of the surgeon himself and facilities available. Larger tumors of stage III and IVA have been resected by approaching through leteral rhinotomy, which has been the most popular approach among the surgeons. Sometimes extended Webers Furguson incision was also used to remove the tumors with intracranial extensions. Majority of our patients (277) were subjected to lateral rhinotomy as we found it convenient and practically feasible in terms of accessibility, removal, better hemostatic control, minimal chances of residual disease and acceptability of post-operative scar.

Limited or smaller tumours can be successfully removed endoscopically. The advocates of endoscopic removal of angiofibroma claims the shortend duration of surgery and a minimal blood loss then open surgery with additional advantage of avoiding unnecessary destruction of facial skeleton [17,18,19]. In our series 13 patients were operated endoscopically.

Since JNA is vascular tumor and possesses a dynamic potential to bleed, measures had always been sought to minimize blood loss. Surgery prior to the era of pre-operative embolization was a nightmare for the surgical teams. Heavy blood loss and massive transfusion led to a number of operative and post-operative complications along with increased morbidity. The advent of super selective pre-operative embolization revolutionarise the surgical management. Many studies highlighted the unmatched superiority

of preoperative embolization over the procedures performed without it [20,21].

Studies comparing the surgeries done with and without embolization demonstrates a markedly significant reduction in per operative blood loss to as low as 1/4th [20]. Our study results are accordingly placed where the blood loss was reduced from 1200-1500 ml to 300-500 ml after the embolozation being started and consequently minimizing the blood products transfusion and hence the chances of transfusion hazards.

To further state, all researchers are not convinced on pre-operative embolozation in every case. They tend to reserve it for larger tumors because they opine that tendency to recur and potential to leave the tumor behind are much greater in patients undergoing preoperative embolozation [22,23]. We faced recurrence in 7.8% of our subject, suggesting a reasonably good success rate. The recurrence rate is variable at different centres as described in literature. Up to 25% of relapse has been reported in certain studies [22].

Radiotherapy was offered only in patients who were in stage IV b without much success. Although in one case the radio therapy was used to down size the tumor and later it was esxcised surgically. The case report was published in a local journal [24].

The search for betterment never ends and so is the case in management of JNA. The current trend to remove the notorious tumor is endoscopically assisted open surgical techniques [25]. The results are encouraging with more radical and definitive removal and lesser number of relapses.

**Conclusion:**

In the developing countries like ours this is a common vascular tumour reported from both hot /desert and cold/high altitude areas. Nasopharyngeal Angiofibroma is rare in females but 4 cases were reported and their genetic analysis was done.

Temporary embolization done preoperatively was proved to be a better option for reduced introperative blood loss. The presence of tumour in the pterygopalatine fossa and basi sphenoid after embolization are sometime missed and not completely removed and thereby, increasing the recurrence rate. Nasopharyngeal angiofibroma in 80s and early 90s presented with late stages but afterwards patient presented in early stages (according to the Fisch classification of nasopharyngeal angiofibroma).

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