

## Juvenile angiofibroma and its management

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### ABSTRACT

This prospective longitudinal study was done to determine extent of tumor and to highlight the importance of lateral rhinotomy approach for nasopharyngeal angiofibroma. Patients with a diagnosis of nasopharyngeal angiofibroma who underwent surgery in Tribhuvan University Teaching Hospital, Maharajgunj, Kathmandu from April, 2004 to Jan 2009 were included in the study. Extent of tumour and surgical approaches were noted in a special form prepared for the study. Fisch Staging system was followed for the extent of tumor. A total number of patients were 23 and all of them were male. Age ranged from 15-30 years with mean age of 18.25 years. All the patients underwent contrast enhanced CTScan. Two patients had stage I tumor, 9 patients had stage II disease, other 12 patients had stage III tumor. Twenty patients were operated by lateral rhinotomy approach. In one patient with the tumour hanging in the oropharynx with a pedical attached to the lateral wall of nasopharynx was removed by peroral route and in another patient with tumour confined to nasopharynx and posterior part of nose was removed by transpalatal approach. One patient who had tumour extension into infratemporal fossa were operated by lateral rhinotomy with inferior sublabial extention approach. Eleven patients underwent preoperative embolization with gelfoam 48 hours prior to surgery. Most of them had stage III disease and were managed surgically by lateral rhinotomy approach. Lateral rhinotomy approach with or without extension of incision can be used to remove tumours in majority of patients.

**Keywords:** Nasopharyngeal angiofibroma, extent of tumour, surgical approaches.

### INTRODUCTION

Juvenile Angiofibroma is a uncommon, benign and extremely vascular tumour that arises in tissue within the sphenopalatine foramen. Rarely it is found at other sites in the nasal cavity and paranasal sinuses. It accounts for less than 0.5% of all neoplasms of the head and neck tumours.<sup>1</sup> Though it develops almost exclusively in the adolescent male, there are reports of tumour being found in children and elderly patients.<sup>2</sup> Juvenile angiofibroma is locally invasive. The vascular nature of angiofibroma causes significant problem in its management. Angiofibroma presents as a well defined lobulated mass that is covered by nasopharyngeal mucosa. Recurrent severe nasal bleeding accompanied by progressive unilateral nasal obstruction are the classical symptoms.

Histologically the tumour consists of proliferating, irregular vascular channels with a fibrous stroma. Tumour blood vessels lacks smooth muscle and elastic fibers, this causes sustained bleeding.

The preferred way of treating the angiofibroma is surgical, though radiotherapy and chemotherapy is been tried for extreme unresectable tumour, but this also needs surgical treatment later on. Many surgical approaches are described in literature but, it all depends upon the extent of tumour and the surgeon's experience.

Though this kind of tumour has been managed in this tertiary centre from the beginning, the study in Nasopharyngeal angiofibroma has not been carried out so far in our context.

### PATIENTS AND METHODS

All the patients diagnosed having nasopharyngeal angiofibroma were operated in the department of ENT and Head and Neck Surgery, Tribhuvan University Teaching Hospital, Maharajgunj by single surgeon. The extent of tumour and surgical approaches were noted in special form. We followed Fisch staging system for the extent of tumour because it is more practical, it defines clinically which tumour needs limited resection and which tumour needs more extensive approaches. The staging system is as follows:

- a. Stage I - Tumours limited to the nasopharyngeal cavity with bone destruction negligible or limited to the sphenopalatine foramen
- b. Stage II - Tumours invading pterygomaxillary fossa, or maxillary, ethmoid or sphenoid sinuses with bony destruction
- c. Stage III - Tumours invading infratemporal fossa or orbital region
  - i. Without intracranial involvement.

**Table-1:** Distribution of age of patients

Age Group (Yrs)	No. of Patients
11-15	6
16-20	11
21-25	3
26-30	3

**Table-2:** Extent of tumour (According to Fisch)

Extent	No. of Patients
I	2
II	9
IIIA	9
B	3
IVA	0
B	0

- ii. With intracranial extradural (parasellar) involvement.
- d. Stage IV – Intracranial intradural tumour
- i. Without infiltration of the cavernous sinus, optic chiasmal region, and/or pituitary fossa
- ii. With infiltration of the cavernous sinus, optic chiasmal region, and/or pituitary fossa

**RESULTS**

Total number of patients operated from April, 2004 to Jan., 2009 were 23 and all the patients were male. Age ranged from 15-30 years but more patients presented at the age group of 16-20 years (Table-1). In 13 patients tumour was located in the left side while in 10 patients it was in the right side (Fig. 1).

All the patients underwent contrast enhanced CTScan before surgery.

**EXTENT OF TUMOUR BY CTSCAN**

Twelve patients had stage III disease. Nine patients had stage IIIA tumour, in 5 patients having erosion of orbital floor while in 4 patients tumour extended to infratemporal fossa. Three patients had stage IIIB tumour having erosion of cribriform plate with minimal intracranial extension but with intact dura. Out of 9 patients with stage II tumour, 4 patients had tumour involving nose, nasopharynx and extending minimally to pterygopalatine fossa, 3 patients had tumour occupying whole pterygopalatine fossa and in 2 patients tumour had extended to ethmoid. Two patients had stage I tumour, limited to nasopharynx and posterior part of nose (Table-2).

**SURGICAL MANAGEMENT**

All patients underwent lateral rhinotomy approach for excision of tumour except one patient who had stage I tumour and underwent transpalatal approach for removal

of tumour. One patient underwent lateral rhinotomy approach with sublabial extention. Eleven patients underwent preoperative angiography with embolization using gelfoam 48 hours prior to surgery. The specimen were sent for histopathological examination and all of them were reported as angiofibroma.

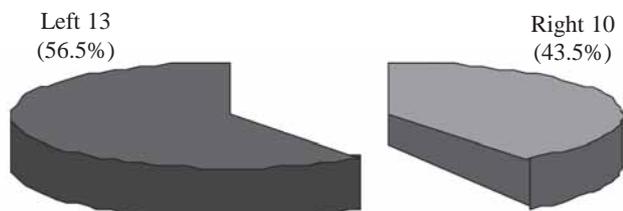
**DISCUSSION**

As mentioned earlier angiofibroma is a rare, benign an extremely vascular tumour of adolescent males. According to studies by Nong *et al* and Mistry *et al* the age of the patients with nasopharyngeal angiofibroma ranged from 13-24 years with mean age of 16.3 years.<sup>3,4</sup> However, Cansiz *et al* reported that patients age ranged from 9-26 years with mean age of 14.9 years.<sup>5</sup> FG Ondrey et al. have mentioned the average and median ages for the presentation being 12.5 and 14 years respectively.<sup>6</sup> In our study more patients were the age group of 16-20 years with mean age of 18.25 years.

A contrast enhanced CTScan and MRI with gadolinium would be the gold standard radiographic evaluation in order to see the extent of nasopharyngeal angiofibroma.<sup>3</sup> According to the extent of tumour surgical approach is planned. All of our patients underwent contrast Enhanced CTScan.

Angiography and selective embolisation should be done at least 48 hours prior to surgery to reduce preoperative bleeding if it is available. In our study we found that the patients who underwent embolization had significant reduction of blood loss during surgery. Those patients with embolization had blood loss about 100-200ml during surgery but one patient who was embolized but had extensive extention into the sphenoid sinus bled around 1 litre, those without embolization had blood loss about 500-800ml.

There is no recurrence so far. We encountered synechia on the opposite nasal cavity in all patients. It occurred due to nasal packing. Eight patients complained epiphora from the operated side which recovered later on. It may be due to damage of nasolacrimal duct during surgery.



**Fig. 1.** Side of tumour

Table-3: Surgical approaches

Surgical Approach	No. of Patients
Lateral rhinotomy	21
Extended lateral rhinotomy	1
Transpalatal	1

One patient who underwent excision of tumour by transpalatal approach developed palatal fistula which was repaired by secondary suture. The lateral rhinotomy scar healed properly.

In the study by Mistry *et al* more than 90.0% of the patients had stage III or IV disease.<sup>4</sup> In another study by Cansiz *et al* out of 22 patients of nasopharyngeal angiofibroma, 3 had stage I tumours, 8 had stage II tumours, 6 had stage III and 5 had stage IV tumours.<sup>5</sup> In our study of 23 patients, 2 patients had stage I tumour, 9 patients had stage II tumour and 12 patients had stage III tumour.

Because nasopharyngeal angiofibroma can involve multiple anatomic sites at the skull base, surgical treatment options can be applied on more or less stage specific basis. Additionally, the experience and surgical preference of the surgeon may affect which approach to choose for a particular tumour. Endoscopic approaches have been suggested for the small stage I and II tumours. Transpalatal approaches have been used to remove angiofibroma limited to nose and nasopharynx. Other surgeons approach lesions of limited extent transfacially through a lateral rhinotomy with medial maxillectomy approach. Midfacial degloving techniques may also be used for tumours with limited extent and by this approach external scar can be avoided.<sup>6</sup> Z Yi *et al* recommended the transantral approach via midfacial degloving with another approach combined, if necessary as the best choice for removal of a huge juvenile nasopharyngeal angiofibroma.<sup>7</sup> According to OH Shaheen also a tumour confined exclusively to the postnasal space should be removed transpalatally.<sup>8</sup> For tumours which encroach on the nasal fossa and just spill over into the pterygopalatine fossa a lateral rhinotomy combined with resection of the medial antral wall may suffice to deliver the tumour. For larger tumours which invade the infratemporal fossa access is improved by a combined transnasal transantral approach, and this can be achieved via a Weber Furguson incision or a facial degloving approach.<sup>9</sup> Combined transpalatal route with a

gingivobuccal incision (Sardana's approach) can be used for access to the pterygomaxillary region.<sup>10</sup> In our setting majority of the patients were treated by lateral rhinotomy with medial maxillectomy approach. There is no recurrence till date of follow up in all the patients.

Juvenile angiofibroma is a very vascular tumour so it should be operated by experienced surgeon in a hospital where there is facility for CTScan or MRI, angiography with embolization facility. There can be massive bleeding during surgery and the chances of residual tumour with recurrence is also high if we operate blindly. It is important to assess the extent of tumour before surgery and plan the approach accordingly. We have excised almost all the tumour by lateral rhinotomy approach without difficulty, only in one case we needed sublabial extension. So we would like to highlight the importance of lateral rhinotomy approach. By this approach we can remove the anterolateral wall and also the posterior wall of maxillary sinus as much as required to visualize the extent of the tumour for complete excision.

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