

Nasopharyngeal Angiofibroma - Evaluation and Management

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ABSTRACT

Objective: To present our experience with 24 patients of Nasopharyngeal Angiofibroma who underwent surgical resection.

Design: A descriptive study

Setting: Department of ENT, Postgraduate Medical Institute, Lady Reading Hospital Peshawar.

Patients and methods: This study included 24 consecutive cases of nasopharyngeal angiofibroma over a period of 5 years. All patients were admitted. Detailed History, clinical examination and relevant investigations were done. Contrast-enhanced CT scan was done in all cases and in selected patients MRI was also done. All the patients underwent surgical excision using various surgical approaches. Patients were regularly followed for any recurrence.

Results: All the patients were male and the average age at diagnosis was 16.5 (range 10-24) years. Nasal obstruction and recurrent epistaxis were the presenting complaints in all cases. Other signs and symptoms included hearing loss, headache, cheek swelling, proptosis, obvious nasal deformity and nasal mass. Contrast-enhanced CT delineated well the tumour and its extensions. All the patients underwent surgical resection of the tumour. In 12 patients tumour resection was done by lateral rhinotomy, in 6 sub labial midfacial degloving approach, in 3 transnasal transmaxillary approach via Weber-Fergusson incision and in 3 transpalatine approach was used. None of the tumours were embolized prior to surgery. The overall recurrence rate was 29.17%.

Conclusion: Nasopharyngeal angiofibroma is a rare, benign and extremely vascular neoplasm of the nasopharynx which occurs in adolescent males. Preoperative evaluation of tumour extent is best done by CT scan and MRI. Pre operative embolization is not obligatory. Surgical excision is the treatment of choice. Most of the tumours are excisable by lateral rhinotomy and midfacial degloving approach, with minimal chance of recurrence.

Key words: Angiofibroma, juvenile nasopharyngeal angiofibroma, nasopharyngeal tumour.

INTRODUCTION

Angiofibroma is an uncommon, benign and highly vascular tumour of adolescent males that originates in the sphenopalatine foramen in the posterolateral wall of the nose^{1,2}. It accounts for less than 0.5 per cent of all head and neck tumours³. The tumour is not encapsulated and may invade the surrounding structures with a potential to extend intracranially. From the sphenopalatine foramen the tumour extends medially into nasopharynx, paranasal sinuses and laterally into the pterygopalatine fossa and infratemporal fossa. Larger tumours can involve the orbit and may even invade the skull base or extend intracranially⁴. Grossly, angiofibromas are well defined lobulated tumours which are covered by nasopharyngeal mucosa. They are pinkish to red in colour and firm in consistency. Histopathologically, the tumour consists of proliferating, irregular vascular

channels within a fibrous stroma. Tumour blood vessels lack smooth muscle and elastic fibres and this feature are responsible for sustained bleeding following minimal manipulation⁵. The blood supply of nasopharyngeal angiofibroma comes primarily from the branches of the external carotid system, although feeders from the internal carotid artery could also contribute to its vascularity. The major arterial supply is usually from the ipsilateral maxillary artery. The cause of this tumour remains unclear. The most prevalent clinical features include partial to complete nasal obstruction and recurrent, spontaneous epistaxis associated with mass in the nose and nasopharynx. Other features include hearing loss, headache, facial deformity, proptosis, anaemia and palatal displacement^{3,6}. Diagnosis is based on CT and MR appearances that are sometimes confirmed by angiography. Because of its vascular nature, biopsy of these lesions is contraindicated. The most practical staging system is that proposed by Fisch. According to Fisch staging system, these tumours may be contained within the nasal cavity and nasopharynx (1), invade the paranasal sinuses or the

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pterygopalatine fossa (II), expand towards the infratemporal fossa, orbit and parasellar region (III) or present intracranial invasion (IV). Surgery is the treatment of choice for Angiofibroma. The options for approaching angiofibroma include lateral rhinotomy, midfacial degloving, transpalatal, Lefort- I Osteotomy, or of course the endoscopic approach. Infratemporal fossa approach, craniofacial resection and a frontotemporal craniotomy are used for advanced stage disease^{7,8}. Intraoperative bleeding may occur in any of the approaches and has been one of the main preoperative complications. Preoperative arterial embolization has been used to reduce the amount of bleeding during surgery with good results⁹. However, there is no consensus as to the routine use of preoperative embolization, given its high cost, inconsistent availability of the procedure and being a contributory cause of recurrence^{10,11,12}. External carotid artery ligation or temporary clamping can be used as an alternative to embolization. Radiotherapy is reserved for the unresectable lesion. Angiofibroma is notorious for a high rate of recurrence which is related to tumour growth at the time of surgery combined with incomplete surgical excision. In view of the very high rate of recurrent disease, prolong clinical and radiological monitoring is necessary for all these patients. Spontaneous regression of residual or recurrent disease is doubtful and cannot be relied upon^{13,14}.

PATIENTS AND METHODS

We treated 24 patients of Angiofibroma during 5 years from January 2007 to December 2011. All patients were admitted. Detailed history, clinical examination and relevant investigations were done. CT scan with contrast was done in all patients and in some cases MRI was also done. Preoperative blood transfusions were given to those patients who were anaemic while 2 to 3 pints of fresh blood were arranged to substitute blood loss during surgery. Preoperative embolization was not used. Unilateral external carotid artery ligation or temporary clamping of the artery was done intraoperatively in majority of the cases. Surgery was selected as the primary treatment modality in all patients. The surgical approaches adopted in the present series included transpalatine approach, lateral rhinotomy, and transnasal transmaxillary approach using Weber-Fergusson incision or midfacial degloving approach. To confirm the nature of the excised tumour histopathology was done. None of the patients received adjuvant treatment such as hormonal therapy, chemotherapy, or radiotherapy.

Regular follow up was conducted at one to two months interval for 1 to 2 years.

RESULTS

All twenty four patients operated for nasopharyngeal angiofibroma were male with age ranging from 10-24 years. Peak incidence was observed between 10-20 years and a mean age of 16.5 years. 18 patients (75%) belonged to different districts of Khyber Pakhtunkhwa, 3 patients (12.5%) were from FATA whereas 3(12.5%) were Afghan refugees. All of them presented with recurrent spontaneous epistaxis and nasal obstruction (100%). Other symptoms included headache (25%) and decreased hearing (12.5%). Important signs on clinical examination included mass in the nose and/ or nasopharynx (100%), retracted ear drum (50%), cheek swelling (25%), palatal displacement (25%) broadening of the nose (25%) and proptosis (16.66%). On routine investigations 6 patients (25%) were found to have iron deficiency anaemia and 1-2 pints of blood was transfused to them preoperatively. All patients underwent contrast enhanced CT for the initial assessment and in 12 patients (50%) MRI was also obtained. The patients were staged according to Fisch classification. Six patients (25%) were at stage 1, 6 (25%) were stage II whereas 12 patients (50%) had stage III disease. None of the patients had stage IV disease in this study. All patients underwent surgical resection of the tumour without preoperative embolization. Unilateral external carotid artery ligation was done in 9 patients (37.5%) while in 3 patients (12.5%) temporary clamping of the artery was used to reduce intraoperative blood loss. Different surgical approaches were used for removal of the tumour. 12 patients (50%) were operated through lateral rhinotomy approach, 6 patients (25%) through midfacial degloving approach, 3 patients (12.5%) through transpalatine approach and 3 patients (25%) by transnasal transmaxillary approach via Weber- Fergusson incision. Mean blood loss during surgery was 1100ml which was replaced by giving 1-3 pints of blood intraoperatively. Postoperative complications included nasal crusting in 6 patients (25%), facial numbness in 4 patients (16.66%) and palatal fistulas in 2 patients (8.33%). Postoperative follow up showed recurrence of the tumour in 7 patients (29.17%). In patients with recurrent disease 2 patients (16.66%) had undergone lateral rhinotomy, 2 patients (33.33%) midfacial degloving approach, and 3 patients (100%) transpalatine approach.

Table I: Age distribution

Age (Years)	=n	%age
Less than 10	03	12.5
11-15	10	41.66
16-20	07	29.16
21-25	04	16.66
Above 25	0	

Table II: Sex distribution

Sex	=n	%age
Male	24	100
Female	0	0

Table-III: Clinical features

Clinical feature	=n	%age
Recurrent epistaxis	24	100
Nasal obstruction	24	100
Headach	06	25
Decreased Hearing	03	12.5
Anaemia	06	25
Mass in Nose / Nasopharynx	24	100
Retracted ear drum	12	50
Cheek Swelling	06	25
Palatal displacement	06	25
Broadening of nose	06	25
Proptosis	04	16.66

Table IV: Stage of the tumours

Stage	=n	%age
I	06	25
II	06	25
III	12	50
IV	0	0

Table V: Surgical treatment

Approach	=n	%age	Recurrence rate
Lateral Rhinotomy	12	50	2(16.66%)
Midfacial degloving	06	25	2(33.33%)
Transpalatine	03	12.5	3(100%)
Transnasal-Maxillary using Weber-Fergusson incision	03	12.5	0

Table VI: Postoperative complications

Complications	=n	%age
Nasal Crusting	06	25
Facial Numbness	04	16.66
Palatal Fistula	02	8.33

DISCUSSION

Angiofibroma is benign, locally invasive and highly vascular tumour of adolescent males which originates in the posterolateral wall of the nose in the region of sphenopalatine foramen. It is a rare tumour and accounts for less than 0.5% of all head and neck tumours^{1,2,3}. In this study we treated 24 patients of angiofibroma. The mean age at presentation in our study is 16.5 years which compares favourably with

other studies. In some studies the tumour had presented in adults as well. All patients were male as in most of the studies, however, the disease have also been reported in females^{15,16}. Recurrent epistaxis (100%), progressive unilateral nasal obstruction (100%) and the presence of mass in the nose and nasopharynx (100%) were the most common clinical features as seen in most of the studies^{3,6}. In this study we received relatively advanced cases as early diagnosis is difficult due to illiteracy ad poor socioeconomic condition in our region. Therefore the features of advanced disease-facial deformity, headach, infratemporal fossa mass and proptosis were not infrequent in our study. CT scan with contrast in axial and coronal planes was the mainstay of diagnosis following positive clinical features. MRI was done in 12 patients (50%) with advanced stage III disease. Imaging delineated the tumour and its extension with erosion of bone behind the sphenopalatine foramen or the skull base. Biopsy was not done in any case. 12 patients (50%) were of stage III, 6 (25%) stage II while the other 6 patients (25%) belonged to stage I. None of the patients had stage IV disease. This compares favourably with other studies done in our country¹⁷. Pre operative anaemia was corrected by blood transfusion of 1-2 units. Pre operative embolization was not done because of its high cost, local unavailability and its association with increased recurrence rate¹⁸. Ligation of ipsilateral external carotid artery or its temporary clamping was adopted in 12 patients which significantly reduced intraoperative blood loss¹⁹. Lateral rhinotomy was the surgical approach which was used in 12 patients (50%). This approach has the advantage of access to the nose, maxillary sinus, ethmoids and nasopharynx. However it leaves a facial scar and removal of facial bones in prepubertal boys could lead to facial asymmetry. The approach is appropriate for stage II and stage III tumours. There are less chances of recurrence in this approach^{9,20}. In our study the recurrence with this approach is 16.66% only. Same approach was used by Azam et al and Zeb et al in Karachi, Tosun et al in Turkey for the same stage with good results²¹. Similarly midfacial degloving approach provides good exposure to the maxillary antrum, nose, pterygopalatine fossa and infratemporal fossa^{9,22}. This approach is an excellent alternative to lateral rhinotomy as it gives good exposure and there is no facial scar but needs extensive removal of facial bones. We have used this approach in 6 patients (25%). Midfacial degloving approach was used in majority of patients with advanced disease by cansiz et al¹. In our study the recurrence rate following this approach was 33.33%. We used transpalatal approach in 3 patients (12.5% with limited early stage

disease but all of them were found to have recurrence of their disease within two years of follow up. There was no recurrence in 3 patients who underwent transnasal-transmaxillary approach via Weber-Fergusson incision, Nowadays, the use of endoscopic surgery for resection of nasopharyngeal angiofibroma is growing but due to lack of experience with this approach we did not use it in our study. We believe that lateral rhinotomy and midfacial degloving approaches are appropriate for most of stage II and stage III tumors. For advanced stage III tumors lateral rhinotomy may be combined with lip splitting or Webr-Fergusson incision to give additional lateral exposure when necessary to minimize recurrence²². The complications rate in our patients was much less as compared to other studies. Angiofibroma has a high recurrence rate of 20-50% in literature. In our study the recurrence rate is 29.17% which is at par with other studies.

CONCLUSION

Although considered a rare tumour of adolescent males, angiofibroma is not uncommon in our region. Surgery has been the mainstay of treatment. Preoperative embolization is not necessary in all cases. Unilateral external carotid artery ligation or temporary clamping is a safe and effective means of facilitating surgery and reducing intra-operative bleeding. Lateral rhinotomy approach affords excellent exposure for complete removal of extensive angiofibroma. This approach may be combined with lip splitting or Weber-Fergusson incision to give additional lateral exposure of the tumour to reduce recurrence. Midfacial degloving approach is an excellent alternative to lateral rhinotomy.

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