Original Article

Angiofibroma

Clinical Profile, Conventional Surgical Approaches and the Outcome of the Surgery in Juvenile Nasopharyngeal Angiofibroma

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ABSTRACT

Objective: To study the clinical profile of angiofibroma, various conventional surgical approaches and the outcome of surgery.

Design: Prospective, analytical study.

Place and Duration of Study: This study was conducted at the Department of Otorhinolaryngology and Head & Neck Surgery, Bolan Medical Complex Hospital, Quetta from January2008 to December2012.

Materials and Methods: This study included 31 patients of juvenile nasopharyngeal angiofibroma over a period of 5 years. All patients were treated by conventional surgical approaches following Fisch staging system. Twenty one (21) patients were operated by lateral rhinotomy approach, Three (3) patients by transpalatal approach, five (5) patients by Weber-Fergusson approach and two (2) by Mid-facial degloving approach. The patients were followed up for a period of three (3) years for any recurrence.

Results: All patients were male with mean age 15.61±2.64 years. The patients presented with recurrent epistaxis, nasal obstruction and nasopharyngeal mass apart from other symptoms and signs. Majority of the patients (80.64%) came with stage II and IIIA disease. Lateral rhinotomy approach was used in majority of cases (67.74%). Recurrence was observed in 5 cases (16.12%). Recurrence rate was less (one out of 21cases) with lateral rhinotomy approach in comparison with other approaches. In transpalatal route recurrence was observed in two (2) cases out of three (3) cases, while in Weber-Fergusson approach it was one out of five (5) and in Mid-facial degloving it was one out of two (2) cases.

Conclusion: Juvenile nasopharyngeal angiofibroma is a disease of male adolescents. The patient most commonly presents with recurrent epistaxis and nasal blockage with nasopharyngeal mass. Surgery is the treatment of choice. Lateral rhinotomy approach gives an excellent exposure for most of these tumours with less chance of recurrence.

Key Words: Juvenile nasopharyngeal angiofibroma, Clinical profile, surgical approaches, Recurrence.

INTRODUCTION

Juvenile nasal angiofibroma (JNA) is a rare, highly vascular, locally aggressive tumour that primarily affects male adolescents.[1]It accounts for approximately 0.5% of all head and neck tumours.[2] It originates from sphenopalatine foramen[3] and invades nasopharynx. From here it may spread to the nose, paranasal sinuses, pterygopalatine fossa, infratemporal fossa, orbit or skull base and intracranial. Grossly, the tumour is pale red to blue smooth mass often lobulated, noncapsulated, sessile or pedunculated and covered by nasopharyngeal mucosa.

The patient presents with recurrent epistaxis and nasal blockage with intranasal mass.[4] Cheek swelling, visual changes, hearing impairment and neurological deficits may be present sometimes. Diagnosis is made mainly by history, clinical examination and imaging studies. Preoperative biopsy is at best avoided for fear of massive lethal bleeding. There are a variety of staging criteria developed when evaluating juvenile angiofibromas which include those developed by Sessions, Chandler, Fisch, and Radkowski.[5-9] The Fisch staging is the most robust and practical.[10] Treatment options for juvenile nasopharyngeal angiofibroma include surgery, radiation therapy, chemotherapy and hormonal therapy.[11] Surgery is the treatment of choice for nasopharyngeal angiofibroma.[4] Preoperative angiography and embolization minimizes the intraoperative blood loss.[12] In surgical treatment of juvenile nasopharyngeal angiofibroma the possibility of recurrences and residual tumours is always there. The present study focuses on the clinical profile of Juvenile nasopharyngeal angiofibroma, various conventional surgical approaches for this tumour and their outcome.

MATERIALS AND METHODS

The study was conducted in ENT and Head & Neck Surgery Department of Bolan Medical Complex Hospital, Quetta over a period of 5 years from January 2008 to December2012. Total patients were 31. All patients underwent a complete workup and contrast enhanced CT scan. Magnetic resonance imaging (MRI) was performed in 2 cases. Since the facility of Angiography and preoperative embolization were not available, therefore, our patients received blood transfusion during surgery and postoperatively.

RESULTS

Fisch staging system was followed to stage the disease. The patients with extensive intracranial involvement and those with recurrent disease were excluded from the study. All the patients were treated surgically using various conventional surgical approaches like Transpalatal, Lateral rhinotomy, Weber-Fergusson and Mid-facial degloving approach. Lateral rhinotomy approach was employed in majority of the patients as most frequent approach. Postoperative specimens were sent for histopathological examination to confirm the diagnosis and all of them were reported as angiofibroma. The Patients were followed-up postoperatively for a period of 3years. During follow-up, the symptomatic patients underwent a new contrast enhanced CT scan to assess the presence and extent of the recurrence.

The common presenting symptoms were recurrent epistaxis (100%), nasal obstruction (100%) and nasal discharge (87.09%). Nasal mass (83.87%), snoring (80.64%), headache (45.16%), voice change (38.70%), hyposmia (38.70%), and hearing impairment (29.03%), swelling of cheek (16.12%) and diplopia (16.12%) were also present in some patients. On clinical examination a pinkish or bluish mass was found in nasopharynx of all the patients, while anaemia, nasal mass, mucopurulent nasal discharchage, palatal bulge, nasal deformity, serous otitis media, conductive deafness, facial asymmetry and proptosis were other signs (Table 1). Four patients had stage I disease, 13 patients stage II tumour, 12 patients stage IIIA and 2 patients stage IIIB tumour (Table 2). Intracranial extension was present in two patients but it was extradural. Majority of the patients had stage II and stage IIIA tumour as shown in Table2. In 17 (54.84%) cases the tumour was right side and in 14 (45.16%) cases tumour was on left side. Lateral rhinotomy approach was employed in 21 patients. Three patients underwent Transpalatal approach. Weber-Fergusson approach was used in 5 cases and Mid-facial degloving approach employed in 2 cases (Figure2). Complete resection was possible in 29 cases (93.54%). In two cases there was residual disease, and they were treated by radiotherapy. Recurrence was observed in 5 cases(16.12%), 2 of them were operated by transpalatal route , one by lateral rhinotomy, one by Weber-Fergusson approach and one by Mid-facial degloving approach. All of the recurrences were observed within 2years after surgery. Over all cure rate was 77.42% (n-24). No mortality occurred in this series (Table3). No major postoperative complication occurred except in one case there was massive postoperative nasal bleeding, which was managed by ipsilateral external carotid artery ligation. Minor postoperative complications were observed in some patients, who included epiphora (6.45%), facial bruising (6.45%), wound infection (9.67%), nasal crusts (12.90%), facial numbness (9.67%) and palatal fistula (6.45%).

Table No1: Clinical features of angiofibroma.

Symptoms	No. of cases	Percentage	Signs	No. of cases	Percentage
Epistaxis	31	100%	Nasopharyngeal mass	31	100%
Nasal obstruction	31	100%	Anaemia	28	90.32%
Nasal discharge	27	87.09%	Mucopurulent nasal discharge	27	87.09%
Nasal mass	26	83.87%	Nasal mass	26	83.87%
Snoring	25	80.64%	Palatal bulge	25	80.64%
Headache	14	45.16%	Nasal deformity	18	58.06%
Voice change	12	38.70%	Serous otitis media	9	29.03%
hyposmia	12	38.70%	Conductive hearing loss	9	29.03%
Hearing impairment	9	29.03%	Facial asymmetry	5	16.12%
Swelling of cheek	5	16.12%	Proptosis	5	16.12%
Diplopia	5	16.12%			

Table No.2: Staging of the cases (According to Fisch staging system).

I	4	12.91%
II	13	41.93%
IIIA	12	38.71%
IIIB	2	6.45%
IVA	0	0
IVB	0	0

Table No.3: Outcome of the Surgery

Outcome	No. of patients	Percentage
Completely cured	24	77.42%
Recurrence	5	16.12%
Residual disease	2	6.46%
Died	0	0
Total	31	100

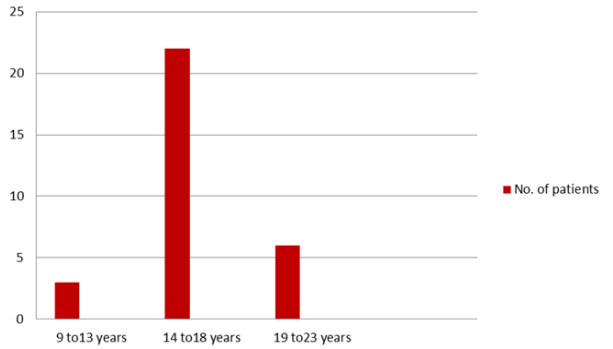

Figure No.1: Age Distribution

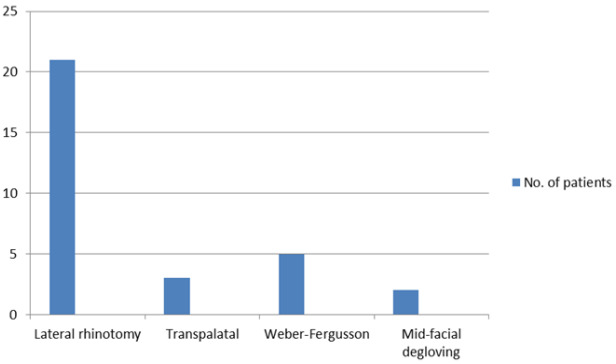

Figure No.2: Surgical approaches for angiofibroma

DISCUSSION

A young male presenting with symptoms epistaxis, nasal obstruction and a nasopharyngeal mass is strongly diagnostic of juvenile nasopharyngeal angiofibroma. In present study all the patients were male and no female case was reported. The mean age of the patients was 14.6years. For juvenile nasopharyngeal angiofibroma the most common age group is the second decade of life.[13, 14] As reported earlier, the incidence of epistaxis and nasal obstruction is more than 90% in patients with angiofibroma.[15] In this study all patients presented with epistaxis, nasal obstruction and nasopharyngeal mass. Other clinical features depend on size and extent of the tumuor. Expansion and extension of the tumour may lead to facial deformity, proptosis, palatal bulge, headache, deafness, nasal mass, voice change, snoring and nasal deformity. Most of these clinical features were observed (Table 1). Diagnosis is made mainly by clinical features, however, modern imaging techniques allow accurate diagnosis and staging of juvenile nasopharyngeal angiofibroma.[6] Various management options for control of juvenile nasopharyngeal angiofibroma include radiotherapy, chemotherapy, hormonal therapy and surgery.

Surgery is the treatment of choice for juvenile nasopharyngeal angiofibroma.[17]Surgical approaches for angiofibroma range from extensive mid-facial degloving to minimally invasive transnasal endoscopic excision. An endoscopic approach is feasible for early stage lesions (Fisch I and II) and conservative external approaches are still useful in advanced stages (Fisch III and IV). The open approaches proved helpful with respect to exposure, safety, cosmetic outcome and low morbidity. Preoperative angiography and embolization minimize intraoperative blood loss[12], which was not available at our center. So we used blood transfusion during surgery. The conventional open approaches for angiofibroma include transpalatal, lateral rhinotomy, transmaxillary via mid-facial degloving, Weber-Fergusson, LeFort 1 osteotomy and Maxillary swing approach. We have used lateral rhinotomy approach in majority of our patients and recurrence was observed in one patient. Lateral rhinotomy approach is effective for the exposure of the nasopharynx paranasal sinuses, pterygopalatine fossa, and medial parts of the infratemporal fossa. Lateral rhinotomy approach with or without extension of incision can be used to remove juvenile angiofibromas in majority of patients.[18] Lateral rhinotomy approach to nose and nasopharynx gives an adequate exposure in almost all the cases of juvenile nasopharyngeal angiofibroma.[19] Surgical treatment, specially the lateral rhinotomy approach and its extensions, is recommended as the best method of managing angiofibroma in most patients.[20]Transpalatal approach provides access to the nasopharynx, sphenoid, sphenopalatine foramen and posterior nares. According to Hosseini SM et al. the lowest recurrence rate is seen either in the transpalatal approach when the tumour is limited to the nasopharynx with extension to the nasal cavity or Para nasal sinuses or with LeFort 1 approach when skull base invasion is present.[20] However, poor results were observed by us with the transpalatal approach. This approach was used in 3 cases of juvenile nasopharyngeal angiofibroma. In one patient there was residual disease and 2 patients came with recurrence. Mid-facial degloving approach provides good exposure to the maxillary antrum, nose, pterygopalatine fossa and infra temporal fossa. When an open approach is used, a midface degloving technique affords excellent exposure even for advanced disease.[21] Mid-facial degloving approach was used in 2 patients. In one case there was residual disease and in other case recurrence was observed. Weber-Fergusson approach was employed in 5 patients and only in one patient recurrence was observed. The main outcome measure was regular follow-up for a period of 3 years. Recurrence of the tumour was observed in 5 cases (16.12%) within two years of surgery. Recurrence was less in patients with lateral rhinotomy approach as compare to other surgical approaches. The incidence of recurrence is in range of 6 to 16.66%.[22,23] Age of the patient and stage of the juvenile nasopharyngeal angiofibroma at presentation are the two most important factors in predicting the recurrence .[24] As younger the age of the patient and later the stage of angiofibroma, are the higher the chances of recurrence. Hence, early diagnosis not only

helps in better management but also prevents recurrence of juvenile nasopharyngeal angiofibroma.

CONCLUSION

Juvenile nasopharyngeal angiofibroma is an uncommon disease of male adolescents. It presents most commonly with recurrent episodes of epistaxis, nasal obstruction and a nasopharyngeal mass. Surgery is the treatment of choice for Juvenile nasopharyngeal angiofibroma. Lateral rhinotomy approach is effective for most of the juvenile nasopharyngeal angiofibromas with less chance of recurrence. Recurrence of juvenile nasopharyngeal angiofibroma is observed within 2years of surgery.

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