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NASOPHARYNGEAL ANGIOFIBROMA?
DOES TUMOR STAGE INFLUENCE RECURRENCE IN NASOPHARYNGEAL ANGIOFIBROMA?

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ABSTRACT… Juvenile nasopharyngeal angiofibroma (JNA) is an uncommon tumor constituting less than 1% of all head & neck tumors. Tumor has an aggressive local behavior if left untreated. Surgery is the mainstay of treatment with no common consensus on a single approach. Tumour stage and surgical approaches are the major determinants of tumour recurrence. **Objectives:** To evaluate the influence of stage of tumor in recurrence in nasopharyngeal angiofibroma. **Study Design:** Descriptive study. **Setting:** Department of ENT and Head and Neck Surgery, PIMS, Islamabad and Ayub medical institution, Abbottabad. **Period:** Jan 2010 to Jan 2014. **Materials and Methods:** Consisting of 34 diagnosed cases of nasopharyngeal angiofibroma. CT-scan was done in all patients and were treated surgically except one patient who was irradiated. All patients were followed up for one year. **Results:** Among 34 patients, 24 patients were classified as stage III, 4 were in stage II and 5 were in stage IVa and one in stage IVb. 17.6% (6/34) of patients had disease recurrence. Stage IVb was treated by radiotherapy while the rest were treated surgically. Patients were followed up for one year both by clinical examination and imaging. Recurrence was found in 5 operated patients and residual disease in stage IV b. 1 (20%) patient of stage Iva disease and 4 (16%) patients of stage III disease had disease recurrence. **Conclusion**: Disease recurrence/ residual is directly related to the tumour stage in nasopharyngeal angiofibroma.

Key words: Tumor stage, Juvenile nasopharyngeal angiofibroma, recurrence

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INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) was described by Hippocrates 5000 years B.C. but was first named by Friedbirg in 1940. It is an uncommon tumor constituting less than 1% of all head & neck tumors.[1] Tumor has an aggressive local behaviour if left untreated.[2] Investigations include CT Scan, MRI and Angiography.[2,3,6] Surgery is the mainstay of treatment.[3,4] Preoperative embolization is considered to reduce the intraoperative blood loss but its cost, availability and a need of neuroradiological expertise makes it a non-feasible option in our setup.[5] Various surgical approaches are in practice depending upon the tumor stage and surgeon's choice and expertise.[3,6] No single technique has been accepted as a gold standard. Intraoperative blood loss, adequate access to the tumor, facial dysthesia and tumor recurrence are additional major concerns about surgical management.[6,7] Transpalatine approach is losing popularity because of its limited access and subsequent recurrence.[7] Lateral rhinotomy with medial maxillectomy (LRMM) offers maximum possible access even to stage III tumors with minimum facial dysthesia. Midfacial degloving approach carries more postoperative morbidity in terms of facial edema and nasal vestibular stenosis.[8,9] More aggressive techniques e.g. maxillary swing by Weber-Fergusson incision and craniofacial resection through type D-1 (infratemporal) approach produce significant facial scarring and disassembly, although effective for tumours having more lateral and intracranial extension.[10,11]

The purpose of this study was to evaluate the role of surgical approaches in determining recurrence of nasopharyngeal fibroma so that the least recuuence approach should be adopted in our setup.

MATERIALS AND METHODS

This Descriptive study was conducted at the ENT Department, Pakistan Institute of medical sciences (PIMS) Islamabad and Ayub Medical Institution (AMI) Abbottabad from Jan 2010 to Jan 2014. The inclusion criteria were patients with nasopharyngeal angiofibroma. Patients with history of bleeding disorders, those not willing for follow-up and history of previous surgery were excluded from the study. The diagnosis of angiofibroma was made on the basis of detailed history, examination and investigations. CT scan was done in all patients to stage the disease. The purpose and benefits of the study were explained to all patients and a written informed consent was obtained.

All patients were treated surgically by lateral rhinotomy approach with medial maxillectomy, midfacial degloving approach and transpalatine approach. Patients were kept in regular follow up for one year. Follow-up record of included patients was analyzed with special attention to the tumour recurrence based upon nasal symptoms, anterior and posterior rhinoscopy and CT scan of nose and paranasal sinuses. Data was collected using a proforma designed for the purpose. The data was stored and analyzed in SPSS version 11.

RESULTS

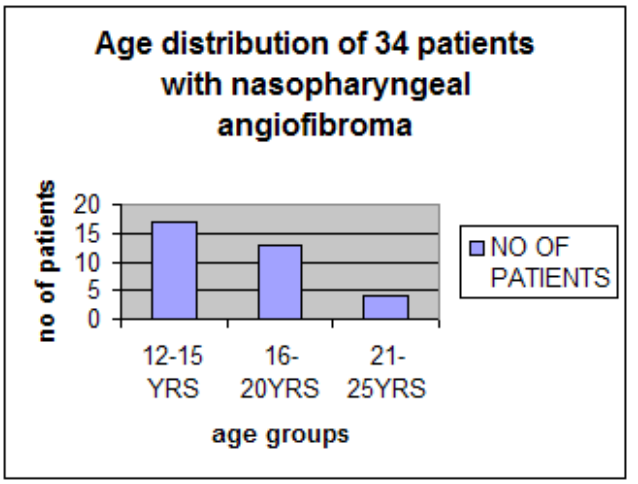
A total of thirty four patients were included in this study from Jan 2010 to Jan 2014. All were males with a mean age of 17.5 years. The minimum age was 12 years while the maximum was 23 years. Most of the patients presented with epistaxsis and nasal obstruction; unilateral in 22% and bilateral 78%. Incidence of signs and symptoms is shown in table-I.

Signs & symptoms	No. of cases
Epistaxsis	34(100%)
Nasal obstruction	34(100%)
Mass Nasopharynx and nose	34(100%)
Cheek swelling	13(38%)
Proptosis	4(12%)
Retracted drum	19(56%)
Conductive deafness	14(41%)
Forward palatal displacement	20(59%)

Table-I. Signs & Symptoms in patients with angiofibroma

Stage of disease	No. of Cases	No. of Recurrences
Stage III	24 (70.6%)	4/24 (16.7%)
Stage Iva	5 (14.7%)	1/5 (20%)
Stage II	4 (11.8%)	0
Stage IVb	1 (2.94%)	1/1 (100%)

Table-II. Stage of the disease and number of recurrences



CT scan was done in all patients to stage the disease. Among 34 patients 1 had cavernous sinus involvement and this single patient was not operated and was given Radiation. Three patients were treated by transpalatine, 5 patients by mid facial degloving while 25 patients were treated with lateral rhinotomy approach with medial maxillectomy approach. Patients were followed up regularly and CT scan was done when recurrence was suspected. The recurrence was seen in 1 (20%) patient of stage Iva disease and in 4 (16.7%) patients of stage III angiofibroma. The one patient (100%) in stage IVb who was irradiated had residual tumour on CT scan. So in 5 operated patients recurrence were found and one had the residual disease. All 5 (15.15%) operated patients with recurrence were given postoperative radiotherapy with a dose of 30-36 Gy. Recurrence rate increases as stage of the disease advances like 16.7% recurrence in stage III, 20% in stage Iva and 100% recurrence/residual in stage IVb. No recurrence was seen in stage II disease.

DISCUSSION

Juvenile nasopharyngeal angiofibroma (JNA)

is the most common of all benign tumours of nose and nasopharynx exclusively affecting young adolescent males.[11,13,14,15] It accounts for 0.5% of all head and neck tumours.[12] There is a common consensus on its origin from outer margin of sphenopalatine foramen close to the medial pterygoid plates.[11] Grossly it has typical bilobed dumbbell appearance occupying nasal fossa and post nasal space with possible extensions into sphenoid sinus, pterygopalatine and infratemporal fossa, erosion of greater wing of sphenoid with retroorbital extensions at times giving a characteristic frog-face appearance.[14]

Microscopically tumour mainly comprises vascular spaces lacking surrounding muscular coats lacking contractile ability.[16] This explains heavy epistaxsis and massive intraoperative blood loss associated with tumour.[17,18] Clinically the triad of progressive nasal obstruction, recurrent heavy epistaxsis and a post nasal mass in a young adolescent male is nothing but angiofibroma until proved otherwise.[15]

Advent of contrast enhanced CT and MRI delineates tumour extension into the surrounding tissues helping in staging the disease and selecting a suitable surgical approach.[19,20,21] Endoscopic removal is advocated in early stage I and stage II disease.[22,23,24] Although surgery is the mainstay of treatment with external beam radiotherapy reserved mainly for intracranial extension, there is no common consensus on a single surgical approach probably because of variable presentation of patients at different stages and surgeon's expertise.[25] We adopted lateral rhinotomy approach with medial maxillectomy through Moure's incision in most of our patients followed by midfacial degloving and transpalatine approach. Cure rate with this approach is 85 % in our study which is consistent with most of the published series.[26,27,28]

Recurrence of the disease is one of the major issue which is especially true in patients with advanced disease having intracranial extension. A proper surgical approach, surgical technique and postoperative radiotherapy greatly reduces the risk of recurrence in such cases. We defined recurrence in our study as persistence of tumour within six months of surgery in terms of recurrent epistaxsis, anterior rhinoscopic findings and/or CT scan of nose and paranasal sinuses. Recurrence/ residual rate in our study is 17.6% (06/34).It varies in different series i.e. 50% in Conley and Williams[29], 2% in Chatter Jee and Soni[28], 6.1% in IH Jaffery and SH.Zaidi[10], 10% in Gohar MS[15], 27.8%, 19%and 10% in older and newer series of Isteraj S.[14] Recurrence rate increases with advance stage disease as in study of Bleier BS.[30] Endoscopic nasal and postnasal examination and CT scan are reliable tools for detection of recurrence. We integrate history, clinical findings and CT scan to determine recurrence of the disease. Regular follow up of the patients is mandatory to detect recurrence at right time.

CONCLUSION

JNA is fairly common in our part of world. More lateral and upward extensions are best delineated on contrast enhanced CT scanning. Residual or recurrent disease after surgery is directly related to the tumour stage and to the surgical approach. Lateral rhinotomy with medial maxillectomy is highly effective even in advanced stage JNA and rate of recurrence increases with stage of the disease.

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"Everyone should be respected
as an individual,
but no one idolized."

Albert Einstein

PREVIOUS RELATED STUDY

Muhammad Saeed, Muhammad Ajmal. NASOPHARYNGEAL ANGIO-FIBROMA (Original) Prof Med Jour 9(2) 111-115 Apr, May, Jun, 2002.

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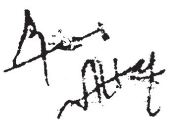
1	Dr. Raza Muhammad	Principal author	
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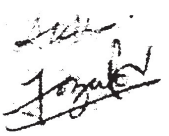
3	Dr. Akhtar Zaman	Co-author, help in data collection	
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