Original Article

Juvenile Nasopharyngeal Angiofibroma, Experience of 35 Cases with Different Surgical Approaches

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ABSTRACT

Objective: 1) To study the clinical presentation. 2) To study the outcome regarding recurrence, prognosis and complications of different surgical approaches of juvenile nasopharyngeal angiofibroma.

Study design: Observational study.

Place and duration of study: The study was conducted at department of Ear, Nose, Throat. Head & Neck Surgery, Quaid-e-Azam Medical College and Bahawal Victoria Hospital Bahawal Pur, from March 2005 to February 2009.

Patients and methods: All young male patients presenting in ENT OPD/COD with complaint of recurrent nose bleeding, nasal obstruction and nasopharyngeal mass were admitted in ward. Detailed history was taken and thorough clinical examination done. Routine investigations and CT scan done in all cases. Staging done according to Fisch staging systems. Sufficient quantity of blood arranged and patient prepared for surgery. Specimen removed was sent for histopathological examination to confirm clinical diagnosis. Follow up was done for 12-18 months to see the recurrence and complications.

Results: Total 35 young male patients clinically diagnosed as JNA and postoperatively confirmed by histopathology were included in the study. Age range was between 10-25 years, majority were between 12-20 years (n-31). All patients were male. All patients presented with epistaxis, nasal obstruction and nasopharyngeal mass. Other presenting symptoms were, anemia (n-29), nasal mass (n-27), ear blockage (n-27), nasal discharge (n-25), headache (n-22), snoring (n-15), speech defect (n-11) and proptosis (n-6). All patients under went surgery. Seventeen patients were approached through lateral rhinotomy, 12 through transpalatal approach and 06 through Weber Furguson approach. In 05 patients recurrence occurred and in 11 patients postoperative complications occurred.

Conclusion: Meticulous surgical approach depending on the stage of JNA reduces the risk of recurrence and complications.

Key words: Angiofibroma, Lateral rhinotomy, Transpalatal, Weber Furguson, Recurrence..

INTRODUCTION

Recognized since ancient time by Hippocrat, Juvenile nasopharyngeal angiofibroma (JNA) is a benign vascular tumour, constitute less than 0.5% of all head and neck tumous[1]. Although histologically benign but is locally aggressive tumour, which arise from posterolateral wall of nasopharynx, found exclusively in adolescent male but in literature female patients have been reported[2]. The exact site of origin is sphenopalatine foramen[3].

Classical presentation is adolescent male with recurrent epistaxis, nasal obstruction, nasal mass and chronic anaemia. The voice acquires a nasal intonation and if the swelling is large enough to force the soft palate down there may be an added plummy quality to it. Other presentations like proptosis, cheek swelling, visual impairment, hearing loss, tinnitus or neurological symptoms may be present in advanced cases. Failing vision has been reported[4] indicating tenting of the optic nerve.

Diagnosis is made on history and clinical examination. Angiography, CT scan and MRI are done to see the feeding vessel and extent of tumour. Pre operative biopsy is not recommended due to intractable bleeding[5].

Surgery is the treatment of choice for JNA. Selection of proper surgical approach depends primarily upon the extension of tumour[6]. Radiotherapy is reserved for advanced tumour[7].

PATIENTS AND METHODS

This observational study was conducted at department of ENT and Head & Neck Surgery Quaid-e-Azam Medical College / B.V. Hospital Bahawal Pur from March 2005 to February 2009. All the adolescent male patients presenting in ENT OPD or in casualty with history of epistaxis, nasal obstruction were admitted in ward. Proper history and clinical examination done. Routine investigations carried out. Those patients who were having nasopharyngeal mass were sent for CT scan and included in the study. Staging of tumour done according to Fisch staging system. Surgery was planned in all cases after arranging sufficient quantity of blood. Stage I tumour under went transpalatal approach, stage II tumours by lateral rhinotomy approach and stage III tumour by Weber Furguson approach. Specimen

removed was sent for histopathological examination. Patients were followed up for 12-18 months postoperatively, recurrence and complications were noted.

Those patients who were having postoperative histological diagnosis other than JNA were excluded from the study. Advance stage IV JNA patients and recurrent cases were also excluded from the study.

RESULTS

Total 35 young male patients clinically diagnosed as JNA and postoperatively confirmed by histopathology were included in the study. Age range was between 10-25 years with mean age of 16.7 years, majority (n-31) were between 12-20 years, 88.57%. One patient was below 12 years (02.85%) and 03 (08.57%) were above 20 years age. (Table No. I).

Table No.I: Age Wise Distribution

Age (years)	No. of cases	Percentage
<12	01	02.85
12-20	31	88.57
>20	03	08.57
Total	35	100

All patients were male, no female patient was reported.

All patients presented with recurrent epistaxis, nasal obstruction and nasopharyngeal mass. Other presenting symptoms were, anemia 29 patients (82.85%), nasal mass in 27 patients (77.14%), ear blockage in 27 patients (77.14%), nasal discharge in 25 patients (71.42%), headache in 22 patients (62.86%), snoring in 15 patients (42.85%), speech defect in 11 patients (31.42%) and proptosis in 06 patients (17.14%) (Table No.2).

Table No.2: Clinical Presentation

Symptoms / signs	No. of cases	Percentage
Recurrent epistaxis	35	100
Nasal obstruction	35	100
Nasopharyngeal mass	35	100
Anemia	29	82.85
Nasal mass	27	77.14
Ear blockage	27	77.14
Nasal discharge	25	71.42
Headache	22	62.86
Snoring	15	42.85
Speech defect	11	31.42
Proptosis	06	17.14

Twelve patients (34.29%) presented in stage I, 17 (48.57%) in stage II and 06 patients (17.14%) presented in stage III (Table No.3).

Table No.3: Staging of the Cases

Stage	No. of cases	Percentage
Stage I	12	34.29
Stage II	17	48.57
Stage III	06	17.14
Total	35	100

All patients under went surgery. Seventeen patients (48.57%) were approached through lateral rhinotomy, 12 (34.29%) through transpalatal approach and 06 (17.14%) through Weber Furguson approach (Table No.4).

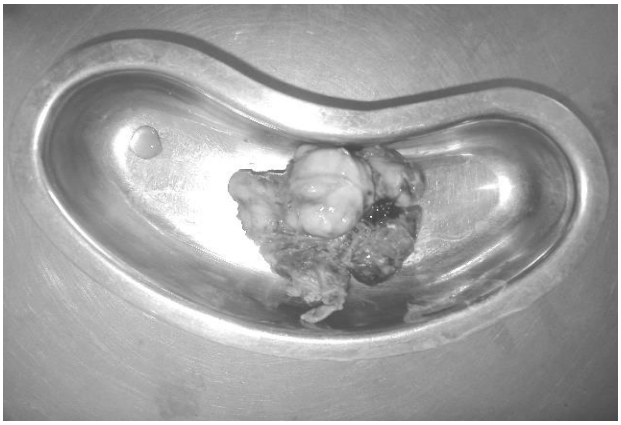
Table No.4: Surgical Approaches

Approach	No. of cases	Percentage
Lateral rhinotomy	17	48.57
Transpalatal	12	34.29
Weber Furguson	06	17.14
Total	35	100

Patients were followed up for 12-18 months postoperatively. In 05 patients (14.29%) recurrence occurred, and they underwent revision surgery. Twelve patients (34.28%) developed postoperative complications. Cheek anesthesia due to section of the infra orbital nerve occurred in all 06 patients (17.14%) operated by Weber Furguson approach, which decreased with the passage of time. Four patient (11.43%) developed post operative wound infection which was dealt with intravenous antibiotics. Palatal fistula was observed in 02 cases (05.71%), which was repaired secondarily. (Table No.5).

Table No.5: Recurrence / Complications

Recurrence / complications	No. of cases	Percentage
Recurrence	05	14.29
Cheek anesthesia	06	17.14
Wound infection	04	11.43
Palatal fistula	02	05.71



Juvenile Nasopharyngeal Angiofibroma removed after surgery

DISCUSSION

JNA is a benign but locally aggressive, highly vascular tumour of nasopharynx found exclusively in adolescent male. It constitute less than 0.5% of all head and neck tumours. Its incidence is more in subcontinent as compared to Europe and America. Midilli[2] found incidence of 02 patients per year in his study with mean age of 16. He also reported a female patient. Witt[8] found 01 patient per year and all were male. While Muhammad[9] described same incidence with age and sex but 12.5 cases per year. Marfani[10] described 08 cases per year, all were male with age range 12-17 and mean age15.4 years. Our study shows 8.8 cases per year with age range 10-25 and mean age 16.7 years which is near to local studies.

Although it is a unilateral tumour and arises from the posterolateral wall of nasopharynx, but in literature bilateral cases of JNA[11] has been reported. Rarely they originates outside the nasopharynx. Reports of primary extranasopharyngeal angiofibroma have appeared sporadically in the literature. One such unusual case of an angiofibroma arising from middle turbinate has been reported by Huang[12]. In our study all cases were arising from the nasopharynx and no extra nasopharyngeal origin was noted. Exact site of origin of JNA has been under discussion for the long time but with advancement of imaging techniques, most researchers agree that it arises from sphenopalatine foramen and from here it goes medially to nasopharynx, forward into the nose and paranasal sinuses. Growth in lateral direction invade pterygopalatine fossa, infratemporal fossa, superior orbital fissure, eyes and cranial cavity.

Clinical presentation of adolescent male with recurrent epistaxis, nasal obstruction and nasal mass is so typical that clinician has no hesitation to make its diagnosis. Other presentations may be like deafness, nasal discharge, headache, change of voice or proptosis. In our study recurrent epistaxis, nasal obstruction and nasopharyngeal mass were present in 100% cases. Other presenting symptoms were, anemia 29 patients (82.85%), nasal mass in 27 patients (77.14%), ear blockage in 27 patients (77.14%), nasal discharge in 25 patients (71.42%), headache in 22 patients (62.86%), snoring in 15 patients (42.85%), speech defect in 11 patients (31.42%) and proptosis in 06 patients (17.14%), which is almost same as studies conducted by Mohammad[9] and Marfani[10]

Preoperative biopsy is contraindicated for fear of bleeding. Postoperatively the tissue removed is sent for histopathology to confirm the clinical diagnosis. CT scan and MRI are needed to see the extent of tumour for staging. Carotid angiography is done to see the feeding vessel and its embolization. In our centre the facility of carotid angiography is not available, so no comments can be given about the feeding vessels.

We staged the tumour after clinical examination and CT scan and / or MRI. In our study stage I tumours were found 34.29%, stage II tumours 48.57% and stage III were 17.14%. According to Marfani[10], 04% cases were in stage I, 24% in stage II and 74% in stage III. While according to study conducted by Mohammad[9], 08% were in stage I, 24% in stage II and 48% in stage III and 20% were in stage IV. The difference observed in stages of the tumour may be due to geographical variation and early or late presentation of the patient.

JNA has always presented management challenges to ENT surgeons because of its vascular nature, site of occurrence and local tissue destruction. Surgery, radiotherapy, chemotherapy and hormone therapy are different treatment modalities for JNA, but surgery is the treatment of choice. Different surgical approaches are used for its resection. These approaches are, lateral rhinotomy, transpalatal, Weber Furguson, midfacial degloving, maxillary swing and infratemporal approach.. We used only three approaches in our study i.e. lateral rhinotomy, transpalatal and Weber Furguson approaches. Pre operative angiography and arterial embolization of feeding vessel 24-72 hours prior to resection has significantly reduced intraoperative blood loss and facilitate resection of large tumours due to shrinkage[13]. We did not have the facility of angiography, so we did not use embolization.

Recent advancement in endoscopic surgery also gave concept of this type of surgery in resection of JNA, but this approach is reserved for tumours involving nose and paranasal sinuses with minimal extension into pterygopalatine fossa[14], but Onerci[15] has tried endoscopic approach for tumours with minimal intracranial extension with minimum morbidity and low recurrence rate. As we do not have the facility of endoscopic surgery, so we only used three classical surgical approaches in our study. These classical surgical approaches are still practized with good results for larger tumours and where endoscopic facility is not available. By using these approaches we have recurrence rate of almost 14.2%, while Hirani[16] has shown recurrence rate of 9.4% in his study, and Witt[8] has shown recurrence rate of 14% and Marfani[10] 7% with surgery in his study. Variation in recurrence rate may be due to difference in stage of tumour, surgical approach, experience of surgeon and facilities available.

In our study complications occurred in 11 (31.42%) cases. Cheek anesthesia seen in 06 (17.14%) cases, wound infection in 03 (08.57%) and palatal fistula in 02 (05.71%) cases. Main complication was cheek anesthesia which was due to section of infraorbital nerve, which is inevitable in Weber Furguson approach. In our study palata fistula occurred in 02 patients out of 12 cases (16.66%) who were operated by transpalatal approach, while Hassan[17] in his study has shown palatal

fistula in 01 casae out of 03 (33.33%) cases. This difference may be due to correction of preoperative anemia and better management of palatal wound. Wound infection in our study is 11.43% which is comparable to other sinonasal surgery.

CONCLUSION

Nasopharyngeal angiofibroma is not uncommon in our region. Angiofibroma should be suspected whenever adolescent male present with nasal obstruction epistaxis and nasopharyngeal mass. Surgery is the treatment of choice. Surgery of JNA is always a challenge due to its vascularity and recurrence. Classical surgical approaches are best to combat this challenge. Pre operative CT scan and / or MRI of paranasal sinuses helps in staging of the tumour Meticulous surgical approach is planned according to the stage of tumour and it reduces the rate of complication and recurrence. We recommend these approaches for operable cases.

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