

Original Article

Management of Juvenile Nasopharyngeal Angiofibroma: A Three Years Retrospective Study

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Abstract

Introduction: Juvenile nasopharyngeal angiofibroma is a pathologically benign yet locally aggressive and destructive vascular tumor. It comprises 0.5% of neoplasms in the head and neck typically affecting adolescent boys.

Material & Method: A retrospective study of surgically treated patients of juvenile nasopharyngeal angiofibroma was performed over a period of 3 years. The study discusses about most common presenting complaints, correlation of preoperative radiological and intraoperative staging and factors affecting recurrence of juvenile nasopharyngeal angiofibroma.

Result: 20 surgically treated patients were included in the study; all were male with average age of 15.7yrs having epistaxis and nasal obstruction as the most common symptom. Pre-operative staging was almost similar with intraoperative staging except one case which needed upstaging. Suitable surgical approach was adopted considering site of lesion. Two recurrent cases were also managed successfully.

Conclusion: Juvenile nasopharyngeal angiofibroma is a benign disease of male adolescents with bone eroding capacity. The planning of surgical approach for its excision has utmost importance which is based on extent of the lesion or staging done pre-operatively. Early pre-operative evaluation not only helps in better management but also prevents recurrence of JNA.

Keywords: Angiofibroma, Nasopharynx

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Introduction

Juvenile nasopharyngeal angiofibroma (JNA) or nasopharyngeal angiofibroma is an uncommon fibrovascular tumour representing only about 0.05% of head and neck tumours of prepubertal and adolescent males. The propensity of the lesion to cause significant degree of morbidity

commonly related to either intracranial extension or massive hemorrhage leading to acquisition of considerable importance in otolaryngology practice. Hippocrates described the tumor in 5th century BC and Friedberg first used the term angiofibroma in

1940^{1,2}. The smaller vessels in the central portion of the lesion typically lack muscular elastic laminae and the absence of muscular coat contribute to the capacity for massive bleeding that occurs with JNA³. Although angiofibroma is histologically benign, it may act in an aggressive fashion characterized by recurrences that may extend into and destroy the adjacent bony structures. JNA originating from

area surrounding sphenopalatine foramen commonly presents with nasal obstruction and epistaxis. The extent of JNA growth is studied clinically and radiologically by contrast enhanced computerized tomography (CT) scan and staged accordingly. Preoperative biopsy is at best avoided for fear of massive lethal bleeding. The condition is most commonly treated by surgical excision and the surgical approach is chosen according to the disease stage. Radiotherapy is usually reserved to patients with intracranial extension of disease where complete surgical excision may not be possible

Methods

This retrospective study on patients surgically treated for JNA whose diagnosis was based on histopathological examination of postoperative specimen. The study was conducted on 20 patients treated at a tertiary care hospital between 2011 and 2013. The 20 patients were given case numbers from 1 to 20. A detailed history followed by thorough clinical examination of head and neck region was done. The patients were staged both clinically and radiologically prior to surgical treatment using Radkowski et al. and Sessions et al. classification of staging respectively^{4, 5, 6}. The patients were treated surgically using various surgical approaches like trans-palatal, lateral rhinotomy and trans-maxillary approach depending on their JNA stage. The patients were followed up

post operatively for a period of 1 year minimum.

Result

The result of study shown in tables and figures.

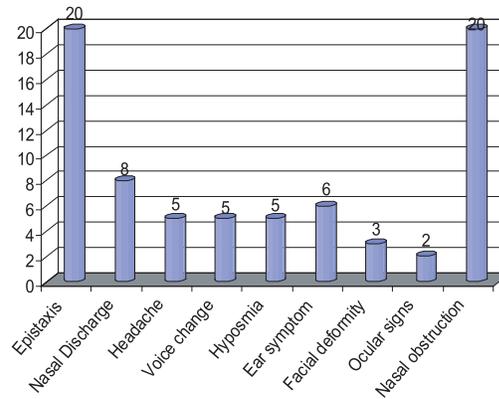


Fig-1: Summary of signs and symptoms observed in the study.

Table-I
Pre-operative staging with sites involved

| Stages | Cases Involved | Total Number |
|--------|----------------|--------------|
| IA | 1,7,8,11,12,18 | 06 |
| IB | 2,3,14,19 | 04 |
| IIA | 9,15 | 02 |
| IIB | 4,5,13,17 | 04 |
| III | 6,10,16,20 | 04 |

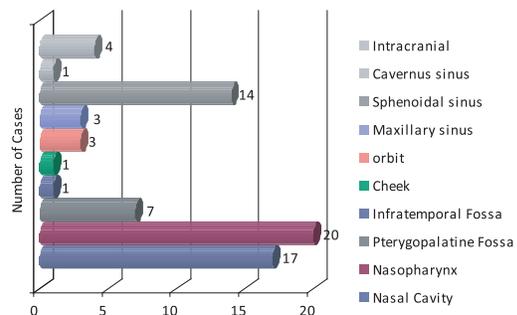


Fig-2: Subsites involved in different cases

Table-II
Surgical approach adopted

| Surgical approach | Cases Involved | Number |
|-----------------------|----------------------------|--------|
| Wilson's Transpalatal | 1,2,3,5,7,8,11,12,14,18,19 | 11 |
| Transmaxillary W.F | 4,6,10,13,16,17,20 | 7 |
| Lateral Rhinotomy | 9,15 | 2 |

Discussion

All the 20 patients were males and no female case is reported in the present series. This study includes patients between the ages 7–19 years with an average age of 15.7 years. Majority of patients, 15 out of 20, fell in the age group of 14–18 years, which happens to be age of most rapid growth of the patient as well as the JNA.

Of all the symptoms, epistaxis and nasal obstruction are the only two, which were present in all the patients (Chart-I)^{8,9,10}. Epistaxis was the most distressing symptom of all which made the patients to seek medical advice early. The amount of bleeding varied from 10 to 100 ml in each episode and was spontaneous. All patients have given a history of aggravation of nasal block with upper respiratory tract infections and more so during winter months. Patients tolerated nasal obstruction better than epistaxis. Nasal discharge was seen in eight patients. Nasal intonation of voice, rhinolalia clausa, was observed in 5 case. It was due to mass obstructing posterior choana. History of hyposmia was evident in 5 patients (case nos. 4, 5,6,10 and 20), though it was not a presenting complaint. Six patients had conductive hearing loss because of serous otitis media caused by eustachian tube block. In these patients mass was encroaching on to tubal openings causing physical obstruction. Facial deformity was complained by 3 cases. Case nos. 1 and 6 had orbital proptosis due to intra orbital extension of JNA. They complained of diplopia on lateral gaze. Their visual acuity was normal.

The extent of growth of JNA was studied both clinically using nasal endoscopy, posterior rhinoscopy and radiologically by contrast enhanced CT scanning or magnetic resonance imaging (MRI) of para nasal sinuses. The lesion was extending into nasopharynx in all the patients, thus in this study it is the most common site of extension (Table 1). Nasal Cavity extension of JNA was seen in 17 patients with attachments to posterior end of turbinates in 13 cases and posterior end of septum in 4 patients. After occupation of nasopharynx the nasopharyngeal angiofibroma tissue invaded sphenoid sinus in about 14 patients. Pterygopalatine fossa was invaded in 7 patients. Orbital involvement through infraorbital fissure (case no. 1) and through breach of lamina papyracea (case nos. 6 and 8) is seen totally in three patients. Case nos. 10 showed cavernous sinus involvement and minimal intracranial extension, which was extradural (Fig. 1). Case no. 10 also had Infratemporal fossa and cheek extension. True maxillary sinus involvement was not seen in any patient but soft tissue density seen on CT scan in case nos. 7, 16 and 20 is because of infection caused by osteomeatal block.

Intra operatively staging was done based on extension of JNA found intraoperatively and had to be upstaged in 1 of the twenty patients (case nos. 8). During surgery, it was found that case no. 8 had JNA extending into sphenoid sinus. However, this upstaging did not prevent total excision surgically with the approach that was planned prior to surgery.

Incidentally this patient were having a CT scan which was taken more than a month prior to surgery and JNA should have expanded by the time and he was taken up for surgery (Table II). They could not obtain a repeat scan with in 1 week prior to surgery because of financial constraints that they had.

The surface of lesion was breached intra operatively due to the massive extent and size of the JNA in case nos. 6, 8 and 16 thus causing excess hemorrhage.

All the patients were treated by surgical excision with an approach carefully selected according to the patients' clinical and radiological assessment of disease status. Principally four approaches were used in this study.

11 out of twenty patients (case nos.1,2,3,5,7,8,11,12,14,18 and 19) were treated by Wilson's transpalatal approach (Table-II). This approach gave excellent visualization of entire nasopharynx, and gave good exposure of sphenoid sinus. Thus this approach is best utilized for JNA confined to nasopharynx and sphenoid sinus.

Two patient with stage IIA disease (case no. 9, 15) was taken up for lateral rhinotomy as the mass was extending into pterygopalatine fossa and sphenoid sinus. This approach with partial medial maxillectomy can give good exposure of pterygopalatine fossa and easy manipulation of maxillary artery.

Transmaxillary approach using Weber-Fergusson's incision was used in case nos. 4, 6, 10, 13, 16, 17 and 20 who were having orbital extension. Case no 6, 10, 16 and 20 also had minimal extradural intracranial extension. The intracranial part in case no. 6 and 20 came out along with its extension in sphenoid, where as in case no. 16 was left in situ as it was encroaching cavernous sinus. Radiotherapy with 35 Gy was administered

to case no. 16 post operatively to deal with left out intracranial extension of JNA which slowly regressed there after a period of 1 year^{11,12}.

Postoperatively, all patients were called for regular follow up for nasal endoscopic examination at monthly interval, and a repeat scan was performed at six monthly interval in whom it was felt necessary. In our study we dealt with 2 patients with recurrent disease¹³. Case no. 16 had history of surgery for JNA at the age of 5 years and he presented to us with recurrent disease at the age of 16 years. He was cured of his recurrence by way of surgery and radiotherapy. Case no. 6 who was 18 year old had symptoms for a long duration of 8 years and presented late with stage III. He developed minimal recurrence 1 year after the primary surgery and was excised of recurrence by endoscopic approach.

Conclusion

Juvenile nasopharyngeal angiofibroma or nasopharyngeal angiofibroma is an uncommon disease of male adolescents. It presents most commonly with nasal obstruction and intermittent moderate to severe nasal bleeding. This benign lesion has great potential for growth in all directions, eroding bony confines. The planning of surgical approach for excision of JNA is based on extent of the lesion or stage. Radiological investigations, like contrast enhanced CT or MRI, are helpful in staging the JNA provided they are done as close to surgery date as possible. Age of the patient and stage of the JNA at presentation are the two most important factors in predicting the recurrence of JNA. As younger the age of the patient and later the stage of JNA, are the higher the chances of recurrence. Hence early diagnosis not only helps in better management but also prevents recurrence of JNA.

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