Management of Juvenile Nasopharyngeal Angiofibroma: A Five Year Retrospective Study

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Abstract Juvenile nasopharyngeal angiofibroma is a pathologically benign yet locally aggressive and destructive vascular lesion of head and neck region typically affecting adolescent boys. The present article is a retrospective study of surgically treated patients of juvenile nasopharyngeal angiofibroma over a period of 5 years. The study discusses about most common presenting complaints, correlation of preoperative radiological and intraoperative staging and factors affecting recurrence of juvenile nasopharyngeal angiofibroma.

Keywords Angiofibroma · Nasopharynx · Neoplasm staging · Recurrence

Introduction

Juvenile nasopharyngeal angiofibroma (JNA) or nasopharyngeal angiofibroma is an uncommon fibrovascular mass arising in the nasopharynx of prepubertal and adolescent males and exhibiting a strong tendency to bleed. The propensity of the lesion to cause life threatening complications by way of massive bleeding has led to acquisition

of considerable importance in otolaryngology practice. Hippocrates [1] described the tumor in 5th century BC and Friedberg first used the term angiofibroma in 1940 [2]. Pathologically, it is characterized by haphazardly arranged vascular channels surrounded by dense paucicellular fibrous tissue. The smaller vessels in the central portion of the lesion typically lack muscular elastic laminae and the absence of muscular coat contributes to the capacity for massive bleeding that occurs with JNA [3]. 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.

Materials and Methods

The present study is a retrospective study on patients surgically treated for JNA whose diagnosis is based on histopathological examination of postoperative specimen. The study was conducted on patients treated at a tertiary care hospital between 2004 and 2009. Though 21 patients were surgically treated for JNA during the period, only 13 were included in our study to fulfill the postoperative review period of minimum 3 years. The 13 patients were

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given case numbers from 1 to 13. 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. [4] and Sessions et al. [5, 6] classification of staging respectively. The patients were advised to undergo preoperative arterial embolization prior to surgery. The patients were treated surgically using various surgical approaches like trans-palatal, endoscopic trans-nasal, lateral rhinotomy and trans-maxillary approach depending on their JNA stage. The patients were followed up post operatively for a period of 3 years minimum.

Discussion

All the 13 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 years [7]. Majority of patients, 9 out of 13, 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 1) [8–10]. Epistaxis was the most distressing symptom of all which made the patients to seek medical advice early. Six patients came for ENT consultation on first episode of epistaxis though other symptoms were already present for a long duration. The amount of bleeding varied from 10 to 100 ml in each episode and was spontaneous, self limiting. 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 11 out of 13 cases but only 2 patients have actually complained about it. It was mucoid to mucopurulent. A history of head ache was given by 6 patients (case nos. 1, 2, 4, 6, 7 and 12), but it was secondary to sinus infection of either maxillary or sphenoid sinus. Nasal intonation of voice, rhinolalia clausa, was observed in 5 cases (case nos. 1, 6, 7, 9 and 12). It was due to mass in nasopharynx obstructing choana. History of hyposmia was evident in 5 patients (case nos. 1, 3, 6, 9 and 12), though it was not a presenting complaint. Four 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. True facial deformity was not a complaint in any of the patients but case no. 1 had cheek swelling due to infratemporal spread of JNA. 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 10 patients with attachments to posterior end of turbinates in 8 cases and posterior end of septum in 2 patients. After occupation of nasopharynx the NPF tissue invaded sphenoid sinus in about 8 patients. Pterygopalatine fossa was invaded in 3 patients. Orbital involvement through infra-orbital fissure (case no. 1) and through breach of lamina

Table 1 Summary of extension of JNA into various sites in all the patients of the study

Site of extension	Case no.												
	1	2	3	4	5	6	7	8	9	10	11	12	13
Nasopharynx	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Nasal cavity		✓	✓	✓	✓	✓	✓	✓	✓			✓	✓
Pterygopalatine F.	✓	✓						✓					
Infra temp. fossa	✓												
Cheek	✓												
Maxillary sinus					✓		✓					✓	
Sphenoid sinus	✓	✓	✓			✓		✓	✓			✓	
Orbit	✓					✓							
Cavernous sinus	✓												
Intra cranial	✓					✓							
Stage [#]	III [®]	IIA	IB	IA	IA	III [®]	IA	IIB	IB	IA	IA	IB	IA

The sign '✓' indicates presence of JNA at that site

[®] indicates stage IIIA according to Radkowski [4]

[#] Indicates according to Sessions staging [5]

Fig. 1 Contrast enhance CT scan showing JNA extending into left sphenoid sinus eroding the lateral wall of sphenoid and entering the cavernous sinus area with intracranial extradural extension

papyracea (case nos. 6 and 8) is seen totally in three patients. Case nos. 1 and 6 showed cavernous sinus and minimal intracranial extension, which was extradural (Fig. 1). Case no. 1 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. 5, 7 and 12 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 3 of

the thirteen patients (case nos. 4, 10, and 13). During surgery, it was found that case no. 4 had JNA extending into sphenoid sinus, case nos. 10 and 13 were having JNA extension into pterygomaxillary fossa hence they were upstaged accordingly. However, this upstaging did not prevent total excision surgically with the approach that was planned prior to surgery. Incidentally these three patients were having a CT scan which was taken more than a month prior to surgery and JNA should have expanded by the time they were taken up for surgery (Table 2). They could not obtain a repeat scan with in 1 week prior to surgery because of financial constraints that they had.

Preoperative embolization using absorbable gelfoam was done 24–48 h prior to proposed time of surgery in 8 out of 13 patients (case nos. 1, 3, 6, 7, 8, 9, 11 and 13) in this study. Other patients either refused or could not afford preoperative embolization. There were no embolization associated complications [11]. In these entire patients, ipsilateral maxillary artery was found to be the major feeding vessel. The intraoperative bleeding was considerably reduced as a result of embolization except in cases 1 and 6. The surface of lesion was breached intra operatively due to the massive extent and size of the JNA in case nos. 1 and 6 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.

Six out of thirteen patients (case nos. 4, 5, 9, 10, 11 and 12) were treated by Wilson's transpalatal approach (Fig. 2). Four patients were having Sessions stage IA disease and

Table 2 Summary of preoperative (pre-op.) staging, surgical approach adopted and intra operative (intra-op.) stage found in each patient of the study

Case no.	Age	Pre-op. stage [#]	Embolized	Surgical approach	Intra-op. stage
1	15	III [®]	Yes	Transmaxillary W.F.	III [®]
2	14	IIA		Lateral rhinotomy	IIA
3	16	IB	Yes	Endoscopic transnasal	IB
4	18	IA		Wilson's transpalatal	IB
5	7	IA		Wilson's transpalatal	IA
6	17	III [®]	Yes	Transmaxillary W.F.	III [®]
7	16	IA	Yes	Endoscopic transnasal	IA
8	12	IIB	Yes	Transmaxillary W.F.	IIB
9	19	IB	Yes	Wilson's transpalatal	IB
10	18	IA		Wilson's transpalatal	IIA
11	12	IA	Yes	Wilson's transpalatal	IA
12	14	IB		Wilson's transpalatal	IB
13	14	IA	Yes	Endoscopic transnasal	IIA

Transmaxillary W.F transmaxillary approach by Weber Fergusson's incision

[®] indicates recurrence

[#] According to Sessions et al. staging [5]

Fig. 2 Wilson's transpalatal approach showing nasopharynx exposure through palate

two had stage IB. 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.

One patient with stage IIA disease (case no. 2) 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. 1, 6 and 8 who were having orbital extension. Case nos. 1 and 6 also had minimal extradural intracranial extension. A modified Weber-Fergusson's incision was used to approach the JNA in pterygopalatine fossa, orbit, sphenoid and cavernous sinus. Partial maxillectomy was done in case no 1 and 6. Orbital decompression was done to resolve proptosis. The intracranial part in case no. 6 came out along with its extension in sphenoid, whereas in case no. 1 was left in situ as it was

encroaching cavernous sinus. Radiotherapy with 35 Gy was administered to case no. 1 post operatively to deal with left out intracranial extension of JNA which slowly regressed there after a period of 1 year [12].

In the present series 3 patients (case nos. 3, 7 and 13) were treated with endoscopic approach [13–17]. Two patients (case nos. 7 and 13) were in stage IA disease and case no. 3 was in stage IB disease. Preoperatively, all the three patients were embolized and this step seems to be helpful for endoscopic approach to prevent intraoperative hemorrhage. Everything was kept ready preoperatively to convert endoscopic approach to transpalatal approach as and when it is felt necessary. The advantages felt with endoscopic approach were better and magnified view of various extents and attachments of JNA and the surgical dissection can well be limited to subperiosteal plane. This approach obviates the need for any skin incision and hence no cosmetic defect is expected. The amount of bleeding was considerably less compared to other approaches. Post operatively the follow up was easier with endoscopic approach. The duration of surgery reduced considerably and there was no need for a prolonged post nasal pack. Hence the patient can take oral feeds early and with less morbidity. The disadvantages are it is technically difficult in untrained hands and visibility drastically reduces if endoscope comes in contact with blood.

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 disease [18]. Case no. 1 had history of surgery for JNA at the age of 5 years and he presented to us with recurrent disease at the age of 15 years. He was cured of his recurrence by way of surgery and radiotherapy. Case no. 6 who was 17 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.

Chart 1 Summary of signs and symptoms observed in the study. Numbers in brackets indicate the number of patients having the particular symptom or sign

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 hamartomatous 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.

Conflict of interest None.

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