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A Clinicopathological Profile and management of Juvenile Nasopharyngeal Angiofibroma in a Tertiary Care Centre in Central India

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Abstract: ***Aims and Objectives: To study the clinicopathological profile and to analyze the efficacy of pre operative embolisation and optimum surgical approach in management of angiofibroma. Material and Method: In present study, 23 patients diagnosed clinicallyand radiologically as a case of Juvenile Nasopharyngeal Angiofibroma. After clinical diagnosis, patients are subjected to Digitalsubtraction angiography followed by pre - op embolisation. Embolization was done using PVC particles and gelfoam and check angiography was done. Patients were treated surgically using Endoscopic or combined surgical approaches. Total intraoperative blood loss was calculatedafter surgery. Results: The mean age of the patients was found to be 17.4 years. The preoperative diagnosis was based on clinical findings and CT findings and the staging wasdone accordingly. The most common feeder was found to be internal maxillary artery in all the cases. Blood loss was minimal to moderate, only one patient had blood loss of around 2200 ml. Conclusion: Juvenile Nasopharyngeal Angiofibroma should be clinically diagnosed and managed by lesser invasive surgical approach, along with preoperative embolisation to minimise intraoperative bleeding. Drilling of the pterygoid wedge was done in all patients resulting in minimal recurrence.***

Keywords: Juvenile Nasopharyngeal Angiofibroma (JNA), Embolization, Angiography, Pathogenesis

1. Introduction

Juvenile Nasopharyngeal Angiofibroma is a highly vascular, benign but locally invasive and destructive fibrovascular neoplasm, with a tendency to local recurrence. It is a rare lesion occurring predominantly in adolescent male.[1, 2] and accounts for less than 0.5% of neoplasm of head and necktumors.[1, 2]. The reported incidence from 1/5000 to 1/50, 000 (one new case per 5000 to 50, 000 patients) of otolaryngological admissions in hospitals of different countries.3 The management of JNAsis a challenge owing to their rich vascular supply, complex adjacent anatomical structures and aggressive growth pattern[4]. Though JNAs being histologically benign, they may act aggressively by extending and destroying adjacent bony structures. The tumor is vascular and fibrous and commonly found in the nasopharynx.[1, 2] The major causes for concern are recurrent severe epistaxis and symptoms being progressive nasal obstruction with space occupying lesion of the nasopharynx.[1, 2]

JNA originating from postero superior margin of the sphenopalatine foramenis often slow growing with extension into pterygopalatine fossa, the paranasal sinus or infratemporal fossa. The tumor evaluation for staging and planning is done by contrast computed tomography (CT) and magnetic resonance imaging (MRI) to reliably assess tumor extent[5]. CT is best utilized for determining bony changes and magnetic resonance imaging (MRI) for soft tissue destruction[6]. Due to the vascular nature of JNA, angiography is often performed to identify the primary vessels that feed thetumor and allow for embolization to reduce intraoperativeblood loss[7]. The mainstay of treatment in JNA is surgery. Other treatment options include radiation therapy (external beam), chemotherapy and hormone therapy antiandrogen (flutamide) [8]. The use of endoscopic excision for JNA limited to nose and pterygopalatine fossa is reported by several authors[9, 10].

Being a highly vascular tumor, surgical excision of angiofibroma carries a significant risk of intraoperative bleeding. But due to emergence of newer interventional modalities and help of pre operative embolisation of feeders of tumor mass, intraoperative bleeding hardly troubles the head and neck surgeons. Recurrence is by far the most common complication despite such extensive and efficient management modalities.

The purpose of this study is to present a clear clinical and pathological picture of angiofibroma to emphasise on treatment modalities and blood loss during surgery and also to look for role of embolisation in reducing the blood loss.

2. Material and Method

A prospective hospital - based study carried out to analyze 23 consecutive cases of Juvenile Nasopharyngeal Angiofibroma (JNA) operated at Government Medical College, Nagpur, India. All the patients of adolescent age group having the history of nasal obstruction and epistaxis and on clinical examination showing nasal mass and mass in nasopharynx, come to ENT O. P. D were included in the study. Initially, a detailed case history, physical examination with emphasis in lesion of head and neck were done for all

the patients. Clinically all the patients seem to be JNA was subjected to radiological investigation (including X ray PNS, Xray Nasopharynx and CT PNS) and nasal endoscopy for confirmation of diagnosis. The patients were staged both clinically and radiologically prior to surgical treatment using Fisch classification of staging (Table 1).

Table 1: Fisch Staging System

Types	Details
I	Tumor limited to the nasopharyngeal cavity; bonedestruction negligible or limited to sphenopalatine foramen.
II	Tumor invading the pterygopalatine fossa or the maxillary, ethmoid or sphenoid sinus with bone destruction.
III	Tumor invading the infratemporal fossa or orbital region • Without intracranial involvement • With intracranial extradural (parasellar involvement).
IV	Intracranial intradural tumor • Without infiltration of the cavernous sinus, pituitary fossa or optic chiasm • With infiltration of cavernous sinus, pituitary fossa or optic chiasm.

After diagnosis of JNA patients were advised to undergo angiography and preoperative arterial embolization prior to surgery. Then saline was injected to prevent reflux into the intracranial circulation. PVA particles along with gel foam for embolisation of arterial feeders and check angiography was done. Check angiography reveals decreased in blushing of tumor mass and then procedure is terminated.

The surgery of patients was planned within 48 hours of embolisation and patients were treated surgically using various surgical approaches like lateral rhinotomy, endoscopic or combined approach depending on their JNA stage. During surgery general anaesthesia was given with throat packing done. All patients received labetalol to maintain intraoperative blood pressure between 90/60 mmhg so as to reduce intraoperative blood loss.

After surgery patient is packed with anterior nasal pack and post nasal pack which was kept for 48 hrs. The intraoperative blood loss is calculated by measuring amount of blood collected in suction machine bottle and subtracting amount of Normal saline used for flushing purposes. Additionally, weight of blood - soaked mops was measured and deducted from weight of normal mops and accordingly the blood loss was calculated (1gm=1ml). Then total intraoperative blood loss is calculated by addition of blood lost in suction and blood loss in mops. Blood transfusion was given to patient according to intraoperative blood loss and post operative haemoglobin status. The patients were followed up postoperatively for a period of 3 months minimum.

3. Observation and Results

During the period under study, a total of 23patients were studied. Allpatients were male, with a mean ageof 17.4 years (12 - 22 years). The staging outlined in Table No.2 shows that 12 out 23cases had stage II (52.10%) lesions, 05 patients had stage IV (21.70%) and 4patients were of stage I (17.30%). Nasal obstruction was most common complaints and seen in all cases. The next most common symptom was epistaxis (22 cases i. e., 95.65%). Other symptoms were hyponasal speech, rhinorrhoea, headache, facial pain and eye swelling.1 patient did not have complaint of epistaxis which may be attributed to smaller size of the tumor mass as all these patients have small size of tumor on CT PNS and belongs to stage I.

On angiography we found that internal maxillary artery was the most common feeder and found in all the cases. Along with Internal maxillary artery concurrent supply with Internal Carotid artery was found in 2 patients. None of the patient shows supply from ascending pharyngeal arteries and vertebral arteries. After embolisation of feeding vessels all patients underwent surgical excision. Total surgical excision was a treatment of choice in our study and in most of the patients the mass was removed by Endoscopic approach used in stage II patients. Stage III and stage IV patients were managed by combined approach (Endoscopic + Lateral Rhinotomy (Figure 1).

Table 3 and 4depicted the intraoperative blood loss andblood transfusion required during surgery respectively. Blood loss was not so severe due to preoperative embolisation and most of the patients require 1 - 2 units of blood transfusion. Only one patient had blood loss around 2200 ml which was due to concurrent supply from branches of Internal Carotid artery which could not be embolised.

Recurrence and residual tumor were found in 1 and 0 cases respectively. This could be attributed to better views through endoscopic approach and mandatory preoperative embolisation in all patients.

4. Discussion

The patients in second decade of life which emphasise on the importance that Juvenile Nasopharyngeal Angiofibroma are a disease of adolescent age group. All the 23 patients were males between the ages 12–22 years with an average age of 17.4yrs and no female case was reported in the present series which was comparable to the observations made in the literature.

The significance of staging of tumor lies in their application for proper surgical approaches so as to excise tumor with ease. We have staged the tumor according to Fisch classification for proper management and staging of tumor was done on the basis of CT PNS findings. In our study 12 patients belong to stage II.4 cases had intracranial extension of tumor. The importance of this lies in the fact that such patients might require neurosurgical intervention and considerable precaution is taken during excision of tumor to avoid CSF leak. Any CSF leak occurring as a complication of surgery can be better managed using neurosurgical backup. In present study none of the patients having

intracranial extension develops CSF leak. The stage III patients were managed by lateral rhinotomy approach. In the study12patients belong to stage II and 4 patients belong to stage I which was managed by endoscopic approach thus avoiding external incision.

According to literature review most surgeons afraid for surgical excision of tumor considering the torrential bleeding occurring during the excision of tumor. If the tumor excision was attempted it was associated with high morbidity. But with the introduction of preoperative embolisation of feeding vessels the tumor can be excised with comparably less bleeding. In our study all patients underwent preoperative embolisation prior to surgical excision and shows Internal Maxillary artery as a primary feeding vessel of tumor. Some patients show supply from ipsilateral Internal Carotid Artery (which couldnot be embolised). We reported that internal maxillary artery was the most common vessel supplying Juvenile Nasopharyngeal Angiofibroma. Similarly, Glenn Roberson et al[11] and Zeba Ahmed et al[12] also shows internal maxillary artery as a common feeding vessel

In the present study blood lossranges from 250 ml to 2200 ml, majority of the patients had intraoperative blood loss between 500 - 750 ml. One patient had blood loss of more than 2000 ml. The average blood loss was 740 ml which was comparable to other studies mentioned in literature[11 - 19]. The blood transfusion requirement was maintained on the basis of intraoperative blood loss, intraoperative vitals and post operative haemoglobin. Twenty - two patients in our series require blood transfusion. Most of the patients require either 1 or 2 units of blood transfusion. So, considering this we can conclude that preoperative embolisation plays a vital role in decreasing the intraoperative blood loss and thus associated morbidity.

5. Conclusion

In conclusion, Juvenile Nasopharyngeal Angiofibroma is a disease which can be clinically diagnosed and should be managed by surgery, without much bleeding intraoperatively due to preoperative embolisation and minimal recurrence.

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Table 2: Stage distribution of patients according to Fisch Classification

Stage	No of patients	Percentage
Stage I	04	17.30%
Stage II	12	52.10%
Stage III	02	08.60%
Stage IV	05	21.70%

Table 3: Blood loss during surgery

Blood loss in ml	No of patients	Percentage
250 to 500 ml	08	34.70%
500 to 750 ml	09	39.10%
750 to 1000 ml	01	04.30%
1000 to 1250 ml	02	08.60%
1250 to 1500 ml	00	00%
1500 to 1750 ml	01	04.30%
1750 to 2250 ml	01	04.30%

Table 4: Blood transfusion required during surgery

No of blood transfusion (in Units)	No of patients	Percentage
0	01	04.34%
1	07	34.70%
2	10	43.40%
3	03	13.00%
4	02	08.60%

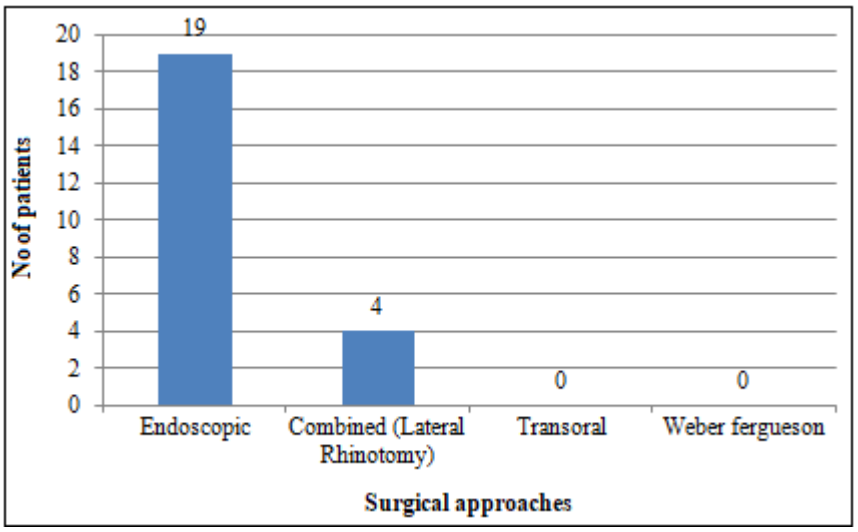

Figure 1: Diagrammatic presentation of various surgical approaches used for surgery