Juvenile Nasopharyngeal Angiofibroma: Combined Approach for Excision, Transpalatal and Endoscopic; A New Perspective

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Abstract Juvenile angiofibroma is a rare, highly vascular, histologically benign but locally invasive tumour, affecting predominantly male adolescents. To describe our experience in diagnosis of angiofibroma and treatment by combined endoscopic and transpalatal route. We analysed a case series of 50 patients of nasopharyngeal angiofibroma in Fisch stages I and II who presented at our institute during a period of four years 2011–2014, their clinical presentation, their treatment by combined transpalatal and endoscopic approach, intraoperative and post operative course, complications and tumour recurrence. Patients were aged 8–26 years. All patients were treated surgically by combined approach endoscopic and transpalatal. The mean operation time was 90 min, and the mean bleeding volume was 300 ml. Only 8 % patients came with recurrence and only 10 % came with palatal perforation (fistula formation). With gratifying results we advocate this novel technique for the treatment of early to intermediate JNA.

Keywords Angiofibroma · Transpalatal · Endoscopic

Introduction

Juvenile nasopharyngeal angiofibroma (JNA) is a benign vascular tumour forming about 0.05 % of all head and neck tumours [1], though higher incidence has been

reported from India [2]. Its propensity to bleed torrentially due to its highly vascular nature and its ability to spread along the base of skull makes the tumour surgically challenging. Grossly, these are pale red to blue smooth masses often lobulated, non-encapsulated, sessile or pedunculated arising in pterygopalatine fossa. They grow along the tissue planes following the path of least resistance. Various theories are described for their pathogenesis of which androgen dependence theory is widely popular. According to current opinions, juvenile angiofibroma is regarded as a developmental defect, affecting the embryonic vascular network surrounding the sphenoid bone [3].

The patient presents with recurrent epistaxis and nasal blockage with intranasal mass, cheek swelling, visual changes, hearing impairment and neurological deficits may be present sometimes [4].

Various management options for control of JNA, include surgery, radiotherapy, chemotherapy, hormonal therapy and electrocoagulation. Surgery remains the mainstay of treatment [4]. Surgical approaches range from extensive mid facial degloving to minimally invasive transnasal endoscopic excision. The various surgical approaches that have been tried are transpalatal, lateral rhinotomy, Weber Fergusson, midfacial degloving and the trans-zygomatic infratemporal approach reflect clearly the technical difficulty in completely excising all tumour extensions.

Open approaches give the benefit of wider exposure but with high morbidity, more hospitalisation, more bleeding, facial disfigurement, sometimes delayed bone growth in children [4] With the evolution of endoscopic surgery, better instrumentation and greater experience in the field, it is now imperative to aid conventional surgical techniques with the telescopes. Here we describe

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combined approach of transpalatal and endoscopic for its removal.

Materials and Methods

Between Jan 2011 and Mar 2015, 50 patients with JNA ranging from stage I to stage II-B (Fisch classification), all young males in the age group of 8–26 years, were diagnosed and operated by combined transpalatal and endoscopic approach.

Preoperative Evaluation

Clinical Evaluation

A thorough history and physical are performed.

In the history following nasal symptoms were evaluated i.e. epistaxis, nasal obstruction headache, unilateral rhinorrhoea and hyposmia.

Head and neck physical examination following were noted proptosis, vision changes, facial swelling, otitis media with effusion.

Fiberoptic nasopharyngoscopy is performed to look for vascular mass emanating from the posterior aspect of the middle and/or superior meatus, possibly filling the nasopharynx.

Radiologic Evaluation

CT scan with contrast is obtained to evaluate the extent of the tumour, the bony anatomy, widening of the sphenopalatine foramen and possibly anterior bowing of the posterior wall of the maxillary sinus, also known as the Holman-Miller sign.

Staging System

We use Fisch staging for the classification of the angiofibroma [5].

FISCH stage description of tumour involvement

- I* Limited to nasopharynx, bone destruction negligible or limited to sphenopalatine foramen.
- II* Invades pterygopalatine fossa or maxillary, ethmoid, or sphenoid sinus with bone destruction.
- III* Invades infratemporal fossa or orbital region.
- IIIA* No intracranial involvement.
- IIIB* Extradural, parasellar involvement.
- IV* Invades dura.
- IVA* Without cavernous sinus, pituitary, or optic chiasm involvement.
- IVB* With the above.

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 our institute between 2011 and 2014. Inclusion criterion included all patients suspected of angiofibroma in stage I, II Fisch. The patients were followed up for six months minimum clinically, endoscopically and radiologically for any recurrence and complication.

Variables analyzed included patient age, sex, symptoms, duration of surgery, volume of intraoperative bleeding, need for nasal packing after surgery, length of hospitalization, immediate and late complications and recurrence.

Surgical Steps

The patient is taken for the surgery under general anaesthesia. After proper painting and draping ipsilateral greater palatine block is given by the infiltration of local anaesthesia. A lazy S shaped incision is given in the palate (Fig. 1). This incision saves one side of the greater palatine artery that prevents the flap necrosis. Through chisel and mallet palatine bone is removed (Fig. 2). It is ensured to leave a sleeve of 2 mm bone for the support of the palatal flap. Care was taken to elevate the upper mucosal flap sufficient enough from the hard palate to make tension-free suture of the incision line to prevent its breakdown and palatal fistula formation. The incision and the palatine bone removal exposes the tumour completely. We then inject mixture of 2 % lignocaine and 1:100000 adrenaline in the tumour. By sharp and the blunt dissection using the Cottle's elevator and bipolar cautery, we give the medial traction and the tumour is delivered first medially then inferiorly and finally from its origin at the sphenopalatine foramen. Any bleeding is stopped by the pressure and by the bipolar cautery. Any oozing is stopped by the

Fig. 1 Lazy S incision

Fig. 2 Removal of the palatine bone

placement of the surgical. Endoscopes were used to check the sphenoid sinus and nasal cavity. With the help of the endoscopes areas suspicious of recurrence like vidian canal are inspected and drilled. In stage II B, tumour tissue was sometimes detected with the help of the endoscopes in the sphenoid sinus which is then removed. Any concomitant nasal pathologies like deviated nasal septum, sinusitis are addressed simultaneously. Nasal and oral packs were almost never needed. The incision was closed by the absorbable sutures. Histopathological examination of the specimen was done in all the cases for confirmation of preoperative diagnosis. All the cases were confirmed as nasopharyngeal angiofibroma histopathologically.

Observations

Age

Age (years)	Number of patients	Percentage (%)
0–10	1	2
11–20	44	88
21–30	5	10

Symptoms

Common symptoms	Number of patients	Percentage (%)
Epistaxis	50	100
Nasal blockage	45	90
Headache	36	60
Facial deformity	24	40

Intra Operative Blood Loss

In our series the average intra operative blood loss is 300 ml.

The following table shows significant haemorrhage which required blood transfusion and nasal packing.

Haemorrhage	Number of patients	Percentage (%)
Uneventful	43	86
Significant bleeding	7	14

Post Operative Palatal Perforation

The patients were evaluated for palatal perforation at the time of the follow up. If palatal perforation was diagnosed it was repaired with good results.

Palatal perforation	Number of patients	Percentage (%)
Present	5	10
Absent	45	90

Results

The success was evaluated if the patient is free of the symptoms and on endoscopic and radiological investigation no sign of the disease is found at the end of the six months.

Result	Number of patients	Percentage (%)
Success	46	92
Failure	4	8

Discussion

In our study all of the patients were male with maximum incidence in adolescence (88 %).

Our results are in harmony with other studies which state that angiofibroma occur exclusively in males with median age of presentation being 14 [6]. The onset of nasopharyngeal fibroma always occurs during adolescence and is one of its most characteristic clinical features.

All of the patients presented with epistaxis (100 %), most of the patients presented with nasal blockade (90 %), with almost half of the patients presented with headache (60 %) and facial deformity (40 %). Results of our study matched with Panda et. al which stated that epistaxis and

nasal obstruction are the chief symptoms [7]. Ardehali et al stated nasal obstruction (93.75 %), epistaxis (62.5 %), and facial swelling (18.75 %) were the most common symptoms [8].

In our series the average intra operative blood loss is 300 ml. The blood loss was measured by measuring the suction bottles and soaked sponge. Significant haemorrhage was seen in 14 % of the patients, which either required blood transfusion or nasal packing to control the bleeding. Blood transfusion was required in about 10 % of the cases. Only about 15 % patients required nasal or oral packs to control the bleeding. Embolization was used in none of our patients.

According to Ardehali et al the mean blood loss in patients operated endoscopically was 1261 ± 893 cc. Martins et al used endoscopic approach when average blood loss was 300 ml [9]. Transpalatal approach have documented about 10 units of blood transfusion [10].

We had intraoperative mean time of 90 min. Other pure endoscopic techniques mean operation time was 120 min.[9].

Post operative palatal perforation was seen in 10 % of the patients during follow up. The palatal perforation was repaired under local anaesthesia with good results. Palatal perforation is a documented complication of the angiofibroma surgery with transpalatal route [11].

92 % of the patients had no recurrence of the disease within a follow up period of minimum 6 months. 8 % patients had occurrence of the disease which was in agreement of other studies [2, 12].

Conclusion

The disease almost exclusively affects adolescent male and symptoms like epistaxis and nasal obstruction can be regarded as almost universal.

This technique which is being used in our institution blends the traditional surgical approach with the edifices of modernity, thus giving advantage of both and negating the disadvantage of both.

With the evolution of endoscopic surgery, better instrumentation and greater experience in the field, it is now imperative to aid conventional surgical techniques with the telescopes.

In our described technique no pre operative embolisation is required, minimum instruments required, average surgical skills required. Normal nasal anatomy and physiology is maintained. The most significant advantage of this technique is that it allows meticulous dissection of the tumour under direct vision in the subperiosteal plane. This minimizes bleeding making it possible to visualize all tumour extensions and remove them completely.

Transpalatal exposure directly exposes the tumour and endoscopes give panoramic view of the tumour attachment, thus combined technique has advantage of both techniques leading to complete removal of the tumour preventing residual disease. With this technique, we therefore hope to see a decrease in the recurrence rate of the tumour.

There is also a significant decrease in the postoperative morbidity as the patient requires no nasal packing and is able to breathe, eat, and talk freely within hours of surgery. The absence of any facial scar is also a major advantage in young adolescent males.

Though, the limitation of this procedure is Fisch stage 3.

With gratifying results we advocate this novel technique for the treatment of early to intermediate JNA.

Compliance with Ethical Standards

Conflicts of interest None.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent Full written consent was taken from the patients regarding the nature of the treatment and further use of his photographs and details for the publishing.

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