Nasopharyngeal Angiofibroma: A 34 - Year Retrospective Review from a Single Institution

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Abstract

Background: Juvenile nasopharyngeal angiofibroma (JNA) is a rare, histologically benign but locally aggressive vascular tumor, classically affecting adolescent males and presenting with nasal obstruction and recurrent epistaxis. This study summarizes a large single-institution experience over three decades in a high-volume tertiary ENT center.

Materials and Methods: A retrospective review was performed of 364 histologically confirmed cases of nasopharyngeal angiofibroma treated at Government ENT Hospital, Hyderabad, India, over 34 years (1980–2014). Medical records were analyzed for demographics, clinical presentation, radiologic evaluation, staging (Radkowski system), operative approach, blood loss/transfusion requirements, duration of hospitalization, complications, recurrence, radiotherapy use, and mortality.

Results: Age at diagnosis ranged from 7–50 years (mean 18 years). Most patients were in the 11–20-year age group (292/364; 80.22%). Surgical excision was the primary treatment in 98.88% of cases, with approaches including lateral rhinotomy (140; 38.46%), transpalatal (117; 32.14%), combined transpalatal + lateral rhinotomy (55; 15.11%), endoscopic endonasal (31; 8.52%), and Weber–Ferguson (20; 5.49%). Endoscopic resections increased after 2000. Estimated blood loss was lower with endoscopic surgery (~300–350 mL) compared with open approaches (~1250–2000 mL), with shorter hospitalization (4 days vs 7–10 days) and fewer complications. Primary radiotherapy was used in 2 patients due to extensive intracranial extension; postoperative radiotherapy was used selectively for residual intracranial disease/intractable bleeding. Overall recurrence was most commonly seen within the first postoperative year; outcomes improved with adoption of endoscopic techniques in later years. Overall mortality was primarily related to severe perioperative or postoperative hemorrhage.

Conclusion: Surgery remains the gold standard for JNA. Over time, a clear institutional trend toward endoscopic endonasal resection was associated with reduced blood loss, fewer complications, shorter hospital stay, and improved disease control in appropriately selected stages. Careful preoperative staging and meticulous skull base clearance are critical to minimize recurrence.

Keywords: Juvenile nasopharyngeal angiofibroma; nasopharynx; endoscopic endonasal surgery; lateral rhinotomy; transpalatal approach; embolization; radiotherapy; recurrence.

Background.

Nasopharyngeal Angiofibroma (NPF) is a rare non malignant vascular tumor but potentially destructive in nature occurring in the nasopharynx of prepubertal and adolescent males. Etiology of this remains unknown. Great advances have been made in their diagnosis and treatment (Zito et-at2001). It accounts to 0.05% of all head and neck tumors. Incidence is relatively higher in India and Egypt than United States and Europe. Patterson (1965) stated an incidence in USA varied from 1:6000 to 1:16000 of ENT cases. Misra and Misra (1964) from North India stated it to be 1in 1107 cases. At our institution i.e. Government ENT Hospital, Hyderabad, India it is 1.08% of all head and neck tumors. The etiology is multifactorial. (Hormonal, desmoplastic response, non- chromaffin

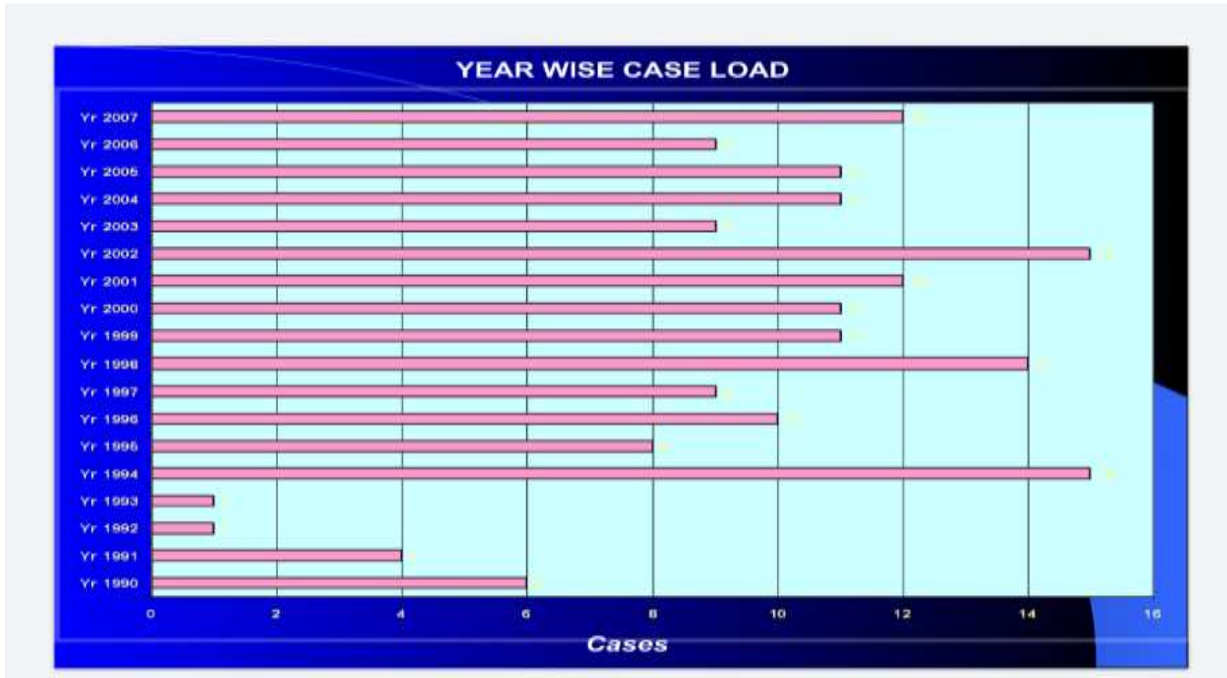
paraganglionic cells). The tumor starts adjacent to the sphenopalatine foramen. 80-90% of patients present with nasal obstruction, 45-60% with epistaxis and facial swelling in approximately in 18% patients. Most of the patients received medical care in the first year.Majority of them belonged to stage I or stage II (Fisch classification). Even though the number of cases of JNF cases per year are varying in number, on the whole it is showing a chronological increase.

Material and Methods.
The retrospective study was carried out in 364 cases of NPF over a period of 34 years (1980 to 2014) at Government Ear Nose Throat Hospital which is a tertiary hospital.All cases were histologically confirmed cases of NPF. The medical records of the patients were reviewed to know age,gender, race, occupation symptoms, site of tumor, tumor size, preoperative embolisations, use of radiotherapy if any, surgery adopted, type of anesthesia, number of units of blood transfused, hospital stay duration, post-operative sequelae, recurrences and deaths if any.

Results.
There were 364 cases of NPF confirmed by biopsy between the years 1980 and 2014. The ages at the time of diagnosis ranged from 7 to 50 years. The average age being 18years.

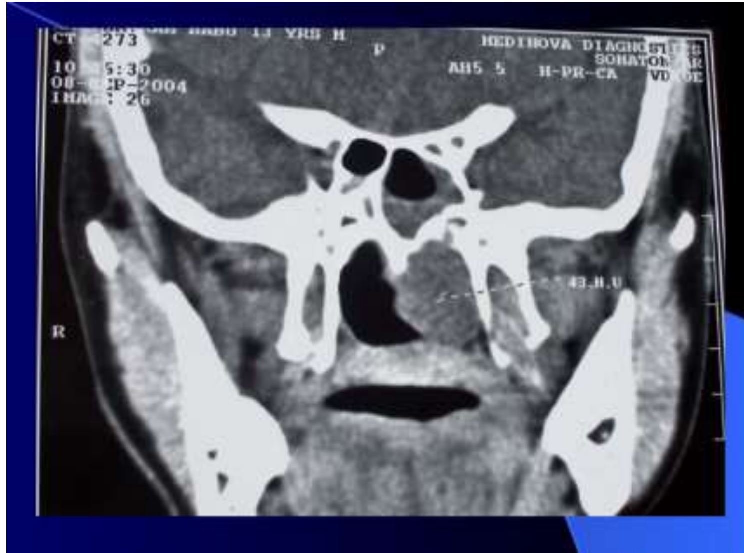
Diagnosis is by history, Physical exam , Radiological study CT scan, MRI and Angiogram. Biopsy is rarely indicated.
Characteristic presentation: Teenage or young adult male with,recurrent epistaxis and nasal obstruction

Year wise Case load

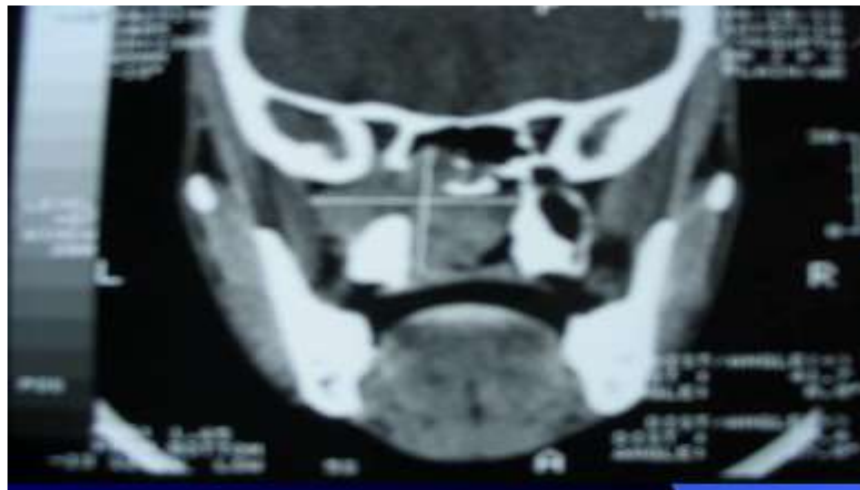


Age incidence (364 patients).		
Age Group	**No. of Patients**	**Percentage (%)**
0-10	19	5.22
11-20	292	80.22
21-30	38	10.44
31-40	9	2.47
41-50	6	1.65

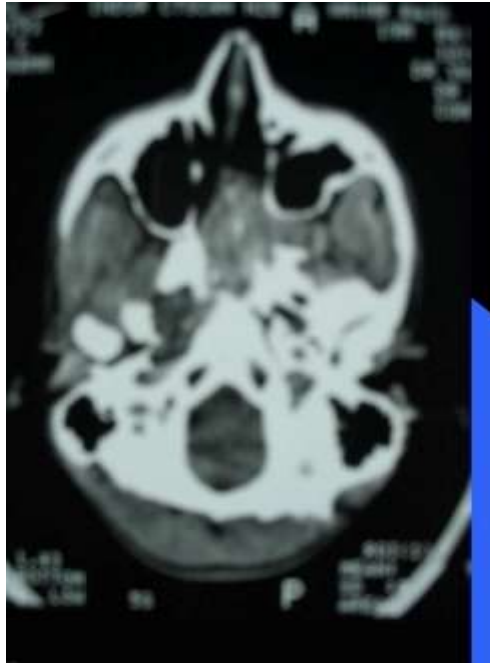
CT scan showing the NPF occupying the posterolateral aspect of nose on Left side



CT scan revealed widening of left sphenopalatine foramen, lesion fills left choana and extends into sphenoid sinus

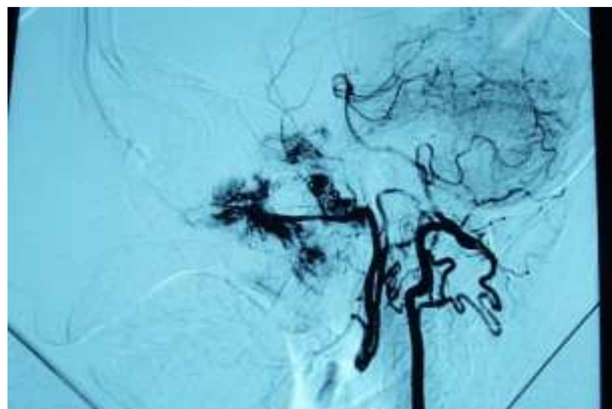


Soft tissue window with contrast (AxialCT)- shows homogeneous enhancement, widening of left sphenopalatine foramen and extension into Nasopharynx & Pterygopalatine fossa

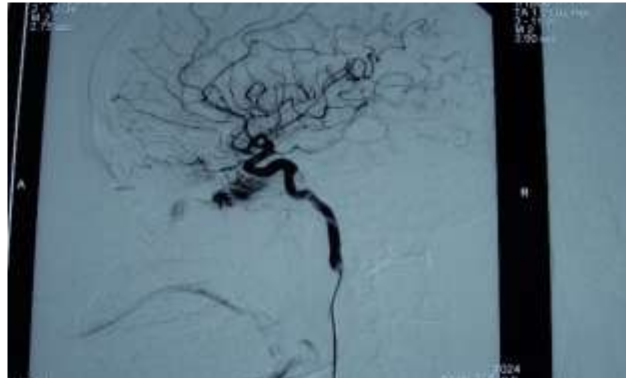


PLANE CONVENTIONAL ANGIOGRAM

DSA PRE EMBOLISATION



DSA POST EMBOLISATION



Surgical Procedures from 1980 - 2014

(N=364 patients)

S.No	Type of Surgery	No. of Patients
1.	Transpalatal	117 (32.14%)
2.	Lateral Rhinotomy	140 (38.46%)
3.	Trans palatal + Lateral Rhinotomy	55 (15.11%)
4.	Endonasal endoscopic approach	31 (8.52%)
5.	Weber-Ferguson approach	20 (5.49%)
6.	Trans oral avulsion	1 (0.27%)

Alternative approaches to Nasal cavities and paranasal sinuses.

Weber Ferguson with Lynch extension
Weber Ferguson with lateral subciliary extension
Weber Ferguson with subciliary extension and supraciliary extension

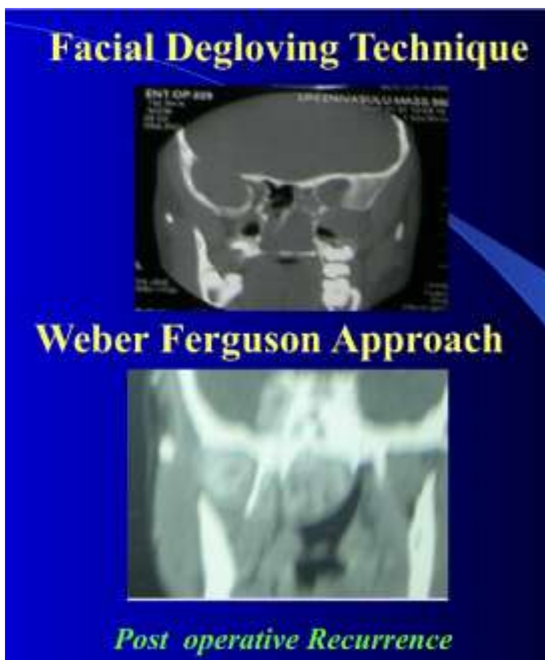
The time elapsed between the beginning of symptoms and the diagnosis was 6 -12 months. Majority of the patients obtained medical care in the first year.

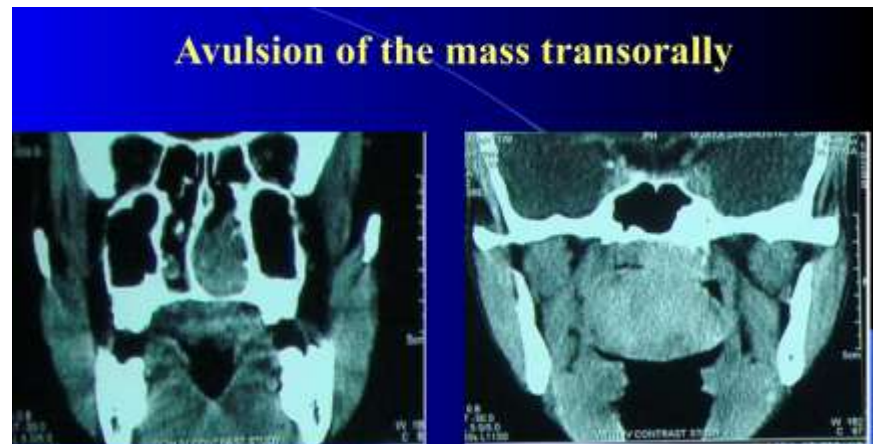
Radkowski staging system according to the regions involved was used. When the tumour was limited to the nasopharynx (Stage I) and those with superior spread into the ethmoid or sphenoid sinuses (Stage IIA) the tumours were removed by a trans palatal route alone or in combination with lateral rhinotomy approach. Tumors with lateral extensions into the pterygopalatine or infratemporal fossae or the cheek (Stage IIB), and those with simultaneous superior and lateral spread were subjected to trans maxillary excision. Endonasal endoscopic excision was under taken in 31 (8.52%)patients. The endoscopic excisions were gaining ground from the year 2000 at our center

Most of our cases presented late, either in stage 2 or stage 3. All patients were treated surgically, mean operation time was 3 hours. The mean blood loss was 1250-2000 ml. in open approaches and 300-400 ml in endoscopic approach. Nasal pack was kept for 5 days and average length of hospitalization was 7-10 days in open approach and 4 days in endoscopic approach. The average length of follow up was 2 years (range 2-5 yrs). Some patients defaulted due to long distance of travel, financial issues and late presentation of symptoms. Some of the well motivated patients are still under follow up. Majority of the cases (98.88%) underwent surgical excision.

Two cases (1.12%) were subjected to primary radiotherapy due to extensive intra cranial extension. Paris etal (2001) suggested selection of the surgical approach should be based on the tumor stage.

Surgical procedure





Radiotherapy was given in two postoperative cases (1.1%) as there was residual intracranial extension and in two (1.1%) cases of intractable post-operative bleeding.

Use of combined modalities of treatment helps achieve decreased intra operative blood loss and cure (Rao & Shewikar – 2000). Piquet etal (1985) reported a low recurrence (10%) in most of his cases where transmaxillary approach was adopted. Howard etal (2001) reported a low recurrence rate by meticulous removal of JNA by

infiltrating pterygoid canal and basisphenoid with novacaine. The most common surgical exposure used was a combination of transpalatal and transantral approaches with least recurrences.(Roberts et al 1989).

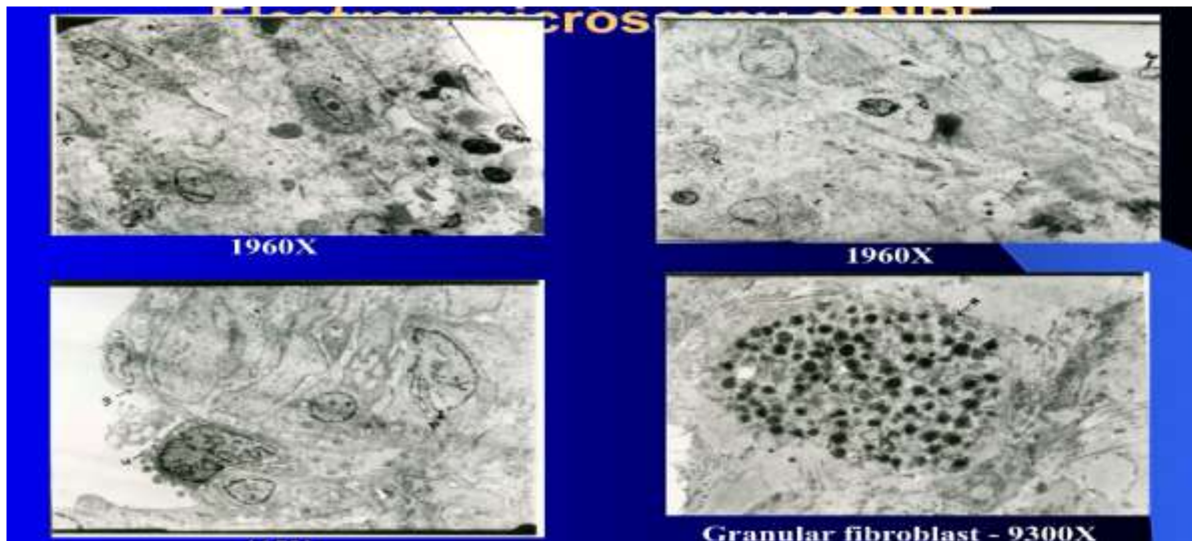
The average operating time was 3-4 hours and the estimated blood loss ranged from 1000 to 1200ml with minimal loss around 500ml in enmass excision and a blood loss of around 2200 ml in situations where the tumor was removed in piece meal. Over all around 4-5 units of blood were transfused (intraoperative + post operative). Earlier studies of Brinquin etal (1986) reported a blood loss of 1300 – 2800 ml and Wu Shan (1983) reported 300 – 500ml blood loss by injecting novacaine solution in the base of the tumor. All patients were given postnasal pack, which was removed on third postoperative day.

Histology

- Myofibroblast is cell of origin
- Fibrous connective tissue with abundant endothelium lined vascular spaces
- Pseudocapsule of fibrous tissue
- Blood vessels leak a complete muscular wall

HIGH POWER MAGNIFICATION

ELECTRON MICROSCOPY PICTURE



Patients were hospitalized for a mean duration of 7 days. Radiotherapy was given post operatively for two cases (0.58%) of intracranial extention and in two cases (0.58%) for intractable post operative bleeding.

PRE AND POST RADIOTHERAPY

COMPLICATIONS

Long term morbidity included nasal crusting, malodorous nasal discharge and occasional fascial dysthesia in transfacial approaches.

Fig 1. Pressure necrosis at the columella due to knot placement for retaining the post nasal pack

Fig 2. Wound dehiscence at the right medial canthus in Weber Ferguson approach

Fig 3. Inadequate closure for right lower eyelid due to inappropriate placement of the incision below the lower eye lid margin in Weber- Ferguson approach.
Fig 4. Late presentation - advanced stage with intracranial extension. Tracheostomy performed and Radiotherapy adviced (1984-1986)

Two cases (0.58%) succumbed to surgery due to profuse bleeding (Table death) . Two patients (0.58%) died due to profuse bleeding in the post operative period due to coagulation disorder (DIC). One patient had torrential bleeding on third post operative day due to major vessel breakdown.

Pre–operative embolisation was given in 19 cases (16.85%) and noticed reduced bleeding in 50% of the cases in the intraoperative period. Embolisation was done within 24-48 hours prior to surgery.

EMBOLISATION

Type of Material	No. of cases	Blood Loss	Transfused
PVP	12	1000ml.	2-3 units
Gel foam	7	1000ml	2-3 units

Comparison of Endoscopic and open Surgical Approaches

Surgical Technique	Endoscopic approach	Open approach
Average blood loss	300-350 ml	1250-2000ml.
Complications	1	42
Lengh of stay	4 days	7-10 days
Recurrence rate	1.44% (follow up from 2007-2014)	13.8%

Recurrence is otherwise a persistent disease or residual disease. The key determinant of recurrence is high tumor growth rate at the time of surgery coupled with incomplete exicision. Lloyd et.al (1959). Recurrence is more in tumors having extra nasopharyngeal extension and also in the ones that have more fibrous elements in on histological examination. 18.40% our cases had recurrence and it was within one year from the time of surgery. Our recurrence rate is high compared to the recurrence reported by Liu et al (16.7%) (2002). Majority of our recurrence were in transpalatal approaches (85%). The mean time for tumor recurrence after operation is 6 months (6-12 months).There was no statistical difference in recurrence rate between embolised and non embolised patients. Our results are in agreement with those of Petruson etal (2002). All the embolisations in our series were uneventful. The recurrence had no correlation with the age of the patient, duration of symptoms, peri operative treatment or surgical approaches. However it strongly correlated with the tumor stage (p less than 0.05).
The post operative follow up is carried by having frequent physical examination and diagnostic nasal endoscopy. Follow up CT scan or MRI is performed at 3 months, 6 months and at 3 years and then yearly for next 5 years.

UNUSUAL PRESENTATION IN FOUR CASES

Case 1: In one case there ws total loss of vision preoperatively (confirmed by Ophthalmologist). The vision returned back to normal after susrgical removal of the tumor. Electroretinogram was not performed in the preoperative period. Recommendation Electroretinogram would have helped us in confirming the vision status pre and postoperative period.
Case 2: Male patient around 30 years presented with nasal block and nose bleed. In view of age and profuse bleeding from nose a diagnosis of NPF was considered and patient was subjected to angiography and embolization. During angiography it was found that the tumor had a feeder vessel from ophthalmic artery and embolization was abandoned and the case was sent for radiotherapy. After completion of radiotherapy it was found that the tumor did not regress (did not respond to radiotherapy). Hence a biopsy wad planned from the nasal mass. The histological examination revealed it to be a Neuroblastoma. There was a delay in diagnosis as it was thought to be a NPF initially.

Case 3: Case presented with a pedunculated mass in the nasopharynx which was confirmed to be NPF by CT scan with contrast. In view of peduncle it was avulsed through the transoral route with Rose position i.e the operative position that is routinely used for adenotonsillectomy. Post operative period was uneventful.

Case 4 : Two brothers in the same family presented with NPF. (Genetic predisposition).

Conclusion

Surgey is the gold standard with a trend towards endoscopic approaches.Recent development of different approaches to nasopharynx has facilitated complete removal of the tumor. Most tumor can be removed by the traditional – modified transpalatal and lateral rhinotomy approaches. Transpalatal excision of tumor gave good results in stage 1 and stage II irrespective of availability of carotid angiography and embolization. Lateral rhinotomy gave good exposure and the recurrence rates are less compared to transpalatal approaches. Overall lateral rhinotomy approach is giving good results and the cosmetic results (minimal facial scar) are fairly good. The correlation between the Histopathology and the recurrence rate will help in choosing the right surgical approach and also post operative follow-up to detect the recurrence at the earliest.

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