

Extended Endonasal Endoscopic Approach for the Resection of Craniopharyngioma-An Analysis of 40 Cases

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Received: July 02, 2018; **Published:** October 27, 2018

Abstract

Introduction: Extended endonasal endoscopic approach for the non-pituitary lesions of the sellar and suprasellar regions are not new in the field of neurosurgery. Following endoscopic approach of the pituitary adenoma surgery, the endoscopic neurosurgeon is eager to develop the skill for non-pituitary sellar-suprasellar lesions. Common sellar suprasellar lesions are pituitary adenoma, craniopharyngioma, tuberculum sellae meningioma and suprasellar germinoma. Traditional transsphenoidal approach gives exposure to the pituitary fossa, whereas extended approach provides exposure to the optic nerve, chiasm, ACOM (Anterior communicating) complex and basal frontal lobe, mammillary body, mid brain, 3rd nerve, basilar artery, and circle of Willis and laterally to the cavernous sinuses (Figure 3A-3C).

Methods: From November 2007 to February 2016 (over 8 years), there were 40 cases of craniopharyngioma operated by the extended endonasal endoscopic approach. Patient's history, clinical findings, pre-operative and post-operative visual acuity, visual field and radiological data were collected and analyzed. All patients underwent endoscopic extended transsphenoidal approach with or without nasoseptal flap technique for closure. 7 patients were given lumbar drain as a treatment for cerebrospinal fluid (CSF) leak.

Results: Age group of the patients varied from 10 to 60 yrs. Male were 19 (47.5%), female were 21 (52.5%). Gross total removal was achieved in 22 cases out of 40 (55.00%) and subtotal in 10 (25.00%) cases. Visual acuity and field of vision improved in all cases. One case (2.5%) of craniopharyngioma had prolonged period of unconsciousness probably from hypothalamic disturbance. CSF leak developed in 10 (25.00%) cases. All Patients required thyroxin and cortisol for replacement. Permanent diabetes insipidus (DI) developed in 10 cases (25.00%). No cases required traditional, open approaches following endoscopic resection. Three patients required permanent CSF diversion via a ventriculoperitoneal shunt after documentation of post-op hydrocephalus (HCP). There was one case of chemical meningitis, and two cases of confirmed bacterial infections. Craniopharyngioma can be successfully resected via a purely endoscopic, endonasal approach. Craniopharyngioma have a higher rate of perioperative hydrocephalus and postoperative CSF leak compared with other tumor types in the same area.

Conclusion: Extended trans-sphenoidal approach is an excellent alternative of skull base approach for the removal of most of the craniopharyngiomas. The endoscopic endonasal route provides a good visualization, especially of the subchiasmatic and retrochiasmatic areas, as well as of the stalk-infundibulum axis, and the third ventricle chamber. It gives better visualization, improved post-operative visual outcome for less manipulation and lower complication rate than craniotomy. However CSF leak and DI are known common complications which have to be managed promptly and appropriately.

Keywords: Endonasal Endoscopic Approach; Craniopharyngioma; Cerebrospinal Fluid (CSF)

Introduction

Craniopharyngioma is a benign epithelial tumor of the sellar region but can have significant neurological and endocrinological consequences and may require treatment that will cause further morbidity [1]. As craniopharyngiomas grow, they can cause significant neurological complications, including visual loss, pituitary insufficiency, and hypothalamic damage, and recurrence. The first descrip-

tion of a craniopharyngioma was credited to Zenker, who made this observation in 1857 [1,2]. Following this, Mott and Barrett, in 1899, documented the occurrence of these tumors and postulated that they arose from the hypophyseal duct or Rathke’s pouch [3]. This was subsequently partially confirmed in 1904, when Erdheim described the tumors histologically and suggested that they arose from remnants of the Rathke duct [3,4]. Finally, in 1932, Cushing introduced the term “craniopharyngioma,” which has been widely used thereafter.

The incidence of newly diagnosed craniopharyngioma is 0.13 to 2.0 persons per 100,000 populations per year. A bimodal distribution by age was noted with peak incidence rate in children (5 - 14 years) and among older adults (age 50 - 75 years) [5]. It accounts for 5% - 10% of brain tumors in children. No sex predilection exists, and they equally occur in males and females [5,6].

The clinical presentation can include a wide range of symptoms, which depend on the location of the tumor and involvement of adjacent structures [7]. Headache is the most common presenting symptom, followed by endocrine deficiencies and visual disturbances. Headache is usually due to either the tumor’s mass effect or hydrocephalus (from obstruction of the foramen of Monro, third ventricle, or aqueduct of Sylvius), which occurs in 15% - 30% of patients [6,7].

Endocrine disturbances are related to direct compression from the tumor. Endocrine deficiency is most commonly with growth hormone (75%), followed by gonadotropin deficiency (40%), thyroid-stimulating hormone (25%), and corticotropin (25%). Growth failure can be seen in up to 93% of children with craniopharyngioma and is related to either growth hormone deficiency, hypothyroidism, or both. Adults have more varied presentation and may develop sexual or menstrual dysfunction. 88% of men experience decreased sex drive, while 82% of women have amenorrhea. Other endocrine dysfunction may lead to precocious puberty and obesity [8].

Large tumors in adults can cause psychiatric symptoms, memory loss, apathy, incontinence depression, and hypersomnia. Long-standing cognitive deficits and profound memory loss have been reported and suggest a worse prognosis. Visual deficits are caused by compression of the optic chiasm from suprasellar tumor growth. Classically, the tumor presents as a bitemporal hemianopsia, but it may also manifest as homonymous hemianopsia, scotoma and papilledema or optic atrophy [9].

Craniopharyngioma is surgically divided into 3 groups: seller, pre-chiasmatic, and retrochiasmatic. According to the grade of involvement of the third ventricle, we identified three main ventricular growth patterns: (1) stalk-infundibulum; (2) infundibulum-ventricular chamber; (3) stalk-infundibulum-ventricular chamber. These tumors occasionally grow into the third ventricle, causing hydrocephalus [10,11].

In relative to infundibular stalk, craniopharyngiomas are divided according to their suprasellar extension: Type I is preinfundibular; Type II is trans-infundibular (extending into the stalk); Type III is retro-infundibular, extending behind the gland and stalk, and has 2 subdivisions (IIIa, extending into the third ventricle; and IIIb, extending into the interpeduncular cistern); and Type IV is isolated to the third ventricle and/or optic recess and is not accessible via an endonasal approach [11,12] (Figure 1).

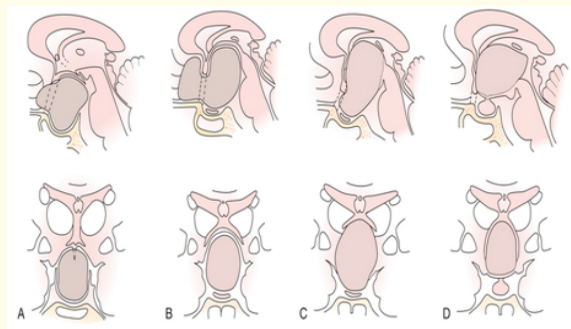


Figure 1: Classification of craniopharyngioma relative to infundibular stalk: 1A (Preinfundibular type 1), 1B (Trans-infundibular type II), 1C (Retro-infundibular type IIIa), 1D (Retro-infundibular type IIIb).

Grading in relative to hypothalamus: In relation to hypothalamus, craniopharyngioma is divided in 3 grades. Grade 0 means no hypothalamic involvement. Grade 1 means Abutting/displacing the hypothalamus. Grade 2 means Involving/Infiltrating the hypothalamus -marked by absence of hypothalamus on imaging (Figure 2A-2C).

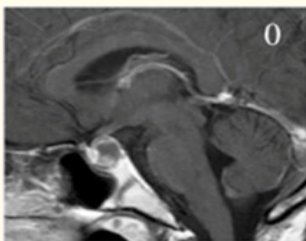


Figure 2A: Grade 0 = No hypothalamic involvement.



Figure 2B: Grade 1 = Abutting/displacing the hypothalamus.

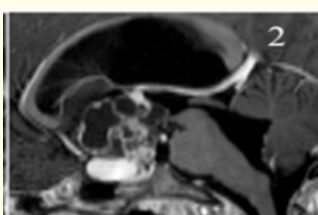


Figure 2C: Grade 2 = Involving/Infiltrating the hypothalamus-marked by absence of hypothalamus on imaging.

The arterial supply is usually from the anterior cerebral and anterior communicating arteries or from the internal carotid and posterior communicating arteries [12].

A craniopharyngioma does not receive blood supply from the posterior circulation, unless it is parasitized from the floor of the third ventricle. As these tumors enlarge, they may elevate and infiltrate the optic chiasm as well as the hypothalamic region. Occasionally, they extend into the pituitary fossa or posteriorly to the ventral pons, and, rarely, they invade the basal ganglia or the brain parenchyma [13].

When predominantly in the sella, these tumors erode the bony floor and enlarge the sella [13].

Surgical management of craniopharyngioma can be transcranial or extended endonasal transsphenoidal approach depending upon location, size and extent of tumor, vascular encasement, degree of pneumatization of sphenoidal air sinus and surgeons choice. Extended endoscopic endonasal transsphenoidal approach recently been popularized for retrochiasmatic craniopharyngioma because of less visual pathway manipulation, maximum tumor removal and relatively safer than transcranial approach [13,14].

Extended endoscopic endonasal transsphenoidal approach provides visualization to the carotid arteries, pituitary gland, infundibular stalk, optic nerve and chiasm, mammillary bodies, basilar artery bifurcations, hypothalamus, 3rd ventricle and foreman of Monro [14], interthalamic adhesion, habenular commissure, posterior commissure, aqueductal sylvius (Figure 3A-3C).

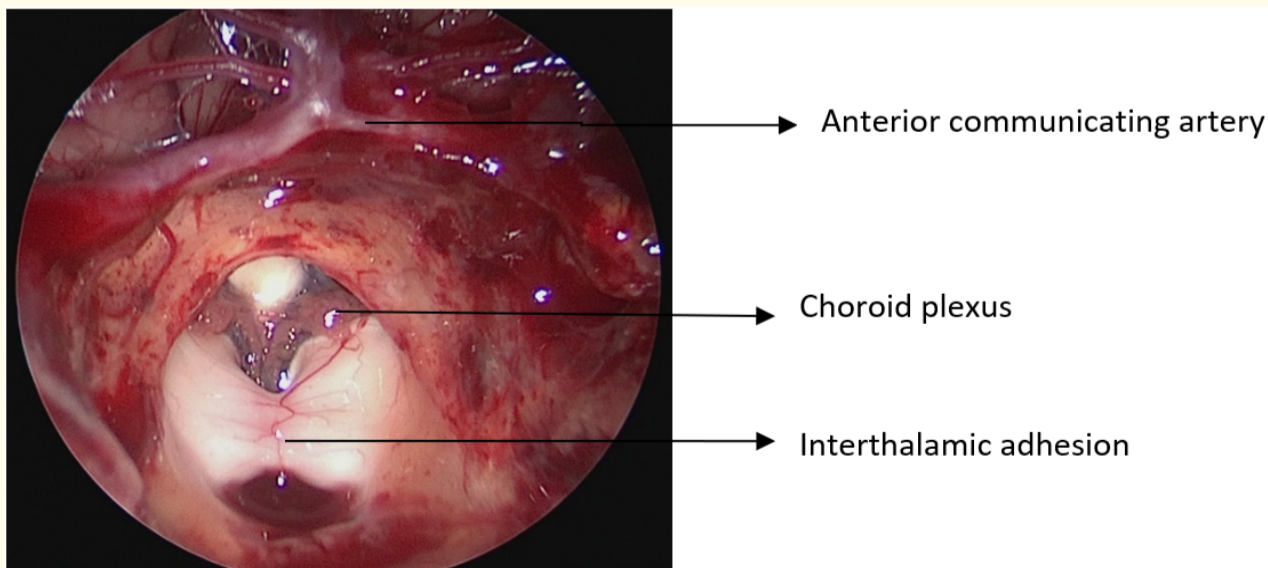


Figure 3A: Endoscopic view of anterior communicating artery, choroid plexus and interthalamic adhesion, after removal of tumor.

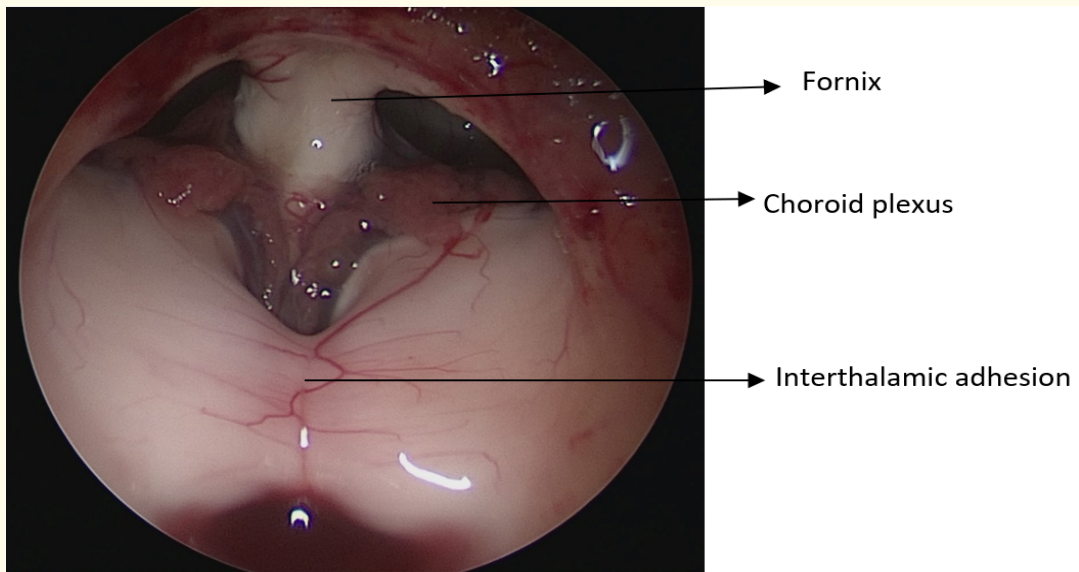


Figure 3B: Endoscopic view of fornix, choroid plexus and interthalamic adhesion.

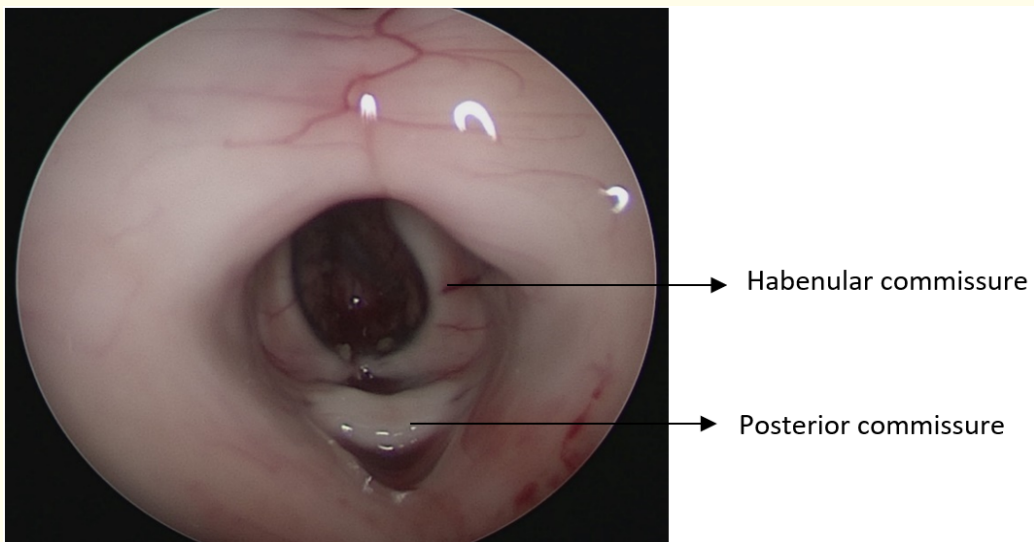


Figure 3C: Endoscopic view of habenular commissure and posterior commissure.

Methods

From November 2007 through February 2016 (over 8 years), there were 40 cases of craniopharyngioma done by extended endonasal endoscopic approach for seller and suprasellar craniopharyngioma. Patient’s history, clinical findings, preoperative and postoperative visual acuity, visual field and radiological data were collected and analyzed. All patients underwent endoscopic extended transsphenoidal approach with or without nasoseptal flap technique for closure. Seven patients were given lumbar drain as a treatment for CSF leak.

Results

There were 40 patients studied retrospectively. Among them 19 patients were males, 21 were females. Age of the patients ranged from 10 years to 60 years, with a mean age of 28.97 years.

All the patients complained of headache and vomiting. Anosmia and personality or behavioral changes were the next common manifestations. Visual impairment was found in all cases. All patients underwent preoperative and postoperative CT (Computed tomography) scan and MRI (magnetic resonance imaging) of brain. Maximum tumor diameter was 4.5 cm and mean diameter was 3.42 cm.

Retrochiasmatic craniopharyngioma in 25 patients (62.5%), subchiasmatic in 8 patients (20.00%) and pre-chiasmatic in 3 patient (7.5%), pre and retrochiasmatic in 4 patients (10.00%) (Table 1). In all the cases surgery was performed with the help of endoscopic extended endonasal approach.

Types of craniopharyngioma	Number of Cases (40)	Percentage
Retrochiasmatic	25	62.5%
Subchiasmatic	8	20.00%
Pre-chiasmatic	3	7.5%
Pre and Retrochiasmatic	4	10.0%

Table 1: Types of Craniopharyngioma (N = 40).

Extent of Resection: Extent of resection was determined using pre-operative and post-operative volumetric analysis of CT scan images. The comparison was performed by chief surgeon. Evaluation of the series has shown that 22 (55.00%) of the 40patients underwent gross-total resection (Table 2). Ten cases (25.00%) of the patients underwent sub-total. 5 cases (12.5%) near total and 3 cases (7.5%) partial total resection were done. All the 18 patients who underwent subtotal, near total and partial total resection were followed-up with early postoperative CT scan (Figure 4A-4D) and neurological evaluation.



Figure 4A: Suprasellar craniopharyngioma (Type IIIB) (Pre-op, Sagittal view).

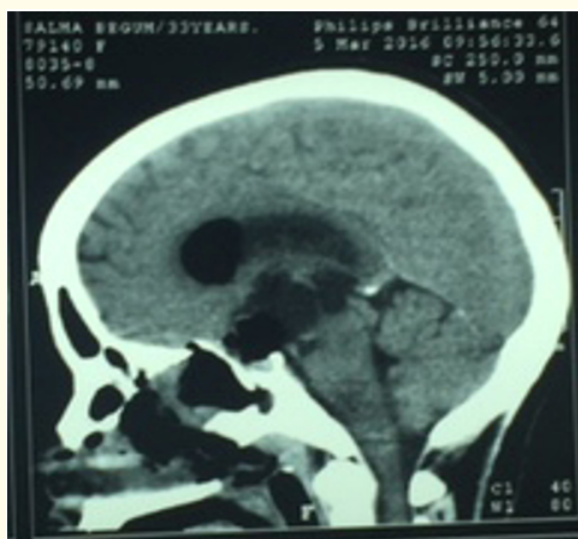


Figure 4B: Suprasellar craniopharyngioma (Type IIIB) (Post-op, Sagittal view).

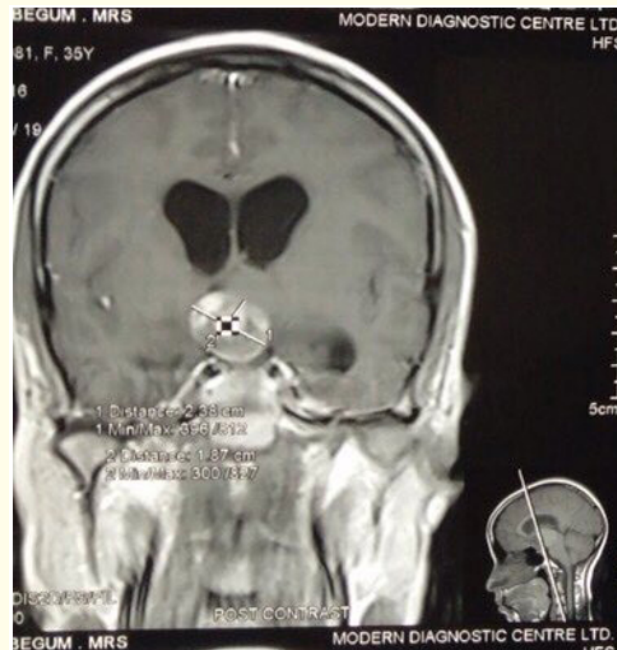


Figure 4C: Suprasellar craniopharyngioma (Type IIIB, pre op) coronal view.



Figure 4D: Suprasellar craniopharyngioma (post op).

The follow up period ranged from 7 month to 16 months. Two cases of tumor were found within this short period of follow up which were re-explored by endoscopic extended endonasal approach. CSF leak was found in 10 cases.

There were mortalities in six cases which was caused by CFS leak, meningitis, meningoencephalitis and septicemia. Those six patients who died due to CSF leaks were treated previously by antibiotic therapy and lumbar drain.

Small subdural hygroma developed in one case and small amount of tumor bed hematoma was seen in one case which resolved over time as was evidenced in later scan. Steven Johnson syndrome developed in one case following Phenytoin therapy.

Outcome was good having GOS (Glasgow outcome score) 5 in 28 patients (70.00%), GOS 4 in 9 patients (22.5%) and GOS 3 in 2 (5.00%) patient during the follow up period (Table 2).

Size of Tumor (cm)	No. of Cases	Percent	Extent of Tumor Removal	No. of Cases	Percent
5 - 10	10	25.0 %	Gross Total	22	55.00%
11 - 15	9	22.5 %	Subtotal	10	25.00%
16 - 20	11	27.5 %	Near Total	5	12.5%
> 21	10	25.0 %	Partial Total	3	7.5 %

Table 2: Shows size, Extent of tumor removal, complication and outcome of craniopharyngioma (N = 40).

Post-op visual outcome: Visual improvement was satisfactory. Postoperative visual acuity and visual field improved in 25 cases (62.5%) (Table 3) (Figure 5A, 5B and Figure 6A, 6B). Our findings of visual improvement after operation is somewhat similar to findings of 61% by Chakrabarti, et al [4]. The visual outcome (for both acuity and fields) was better in younger patients and those with a shorter duration of symptoms. Patients with lesser degrees of preoperative visual acuity compromise had better postoperative visual acuity outcome. However, the severity of preoperative visual field defects did not seem to predict postoperative field outcome, and even patients with severe preoperative field defects often had striking postoperative improvement.

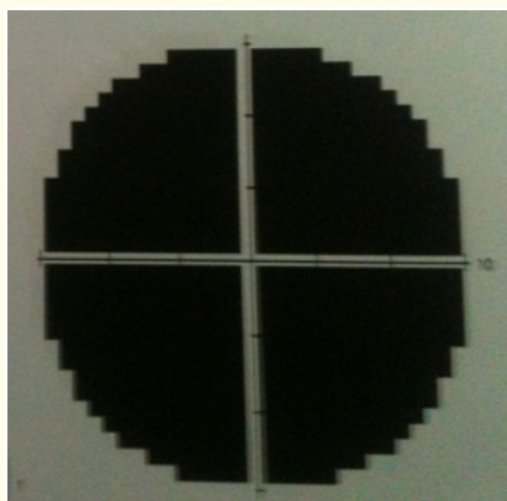


Figure 5A

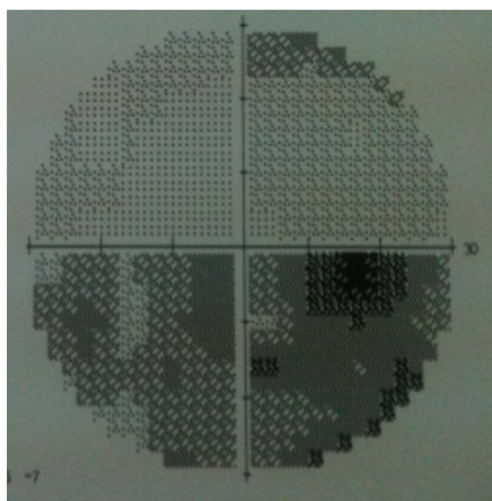


Figure 5B

Figure 5: Pre op (A) and Post op (B) picture of visual field of right eye showing significant improvement.

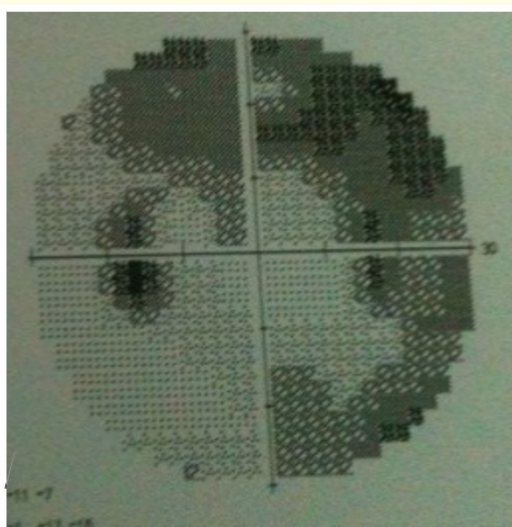


Figure 6A

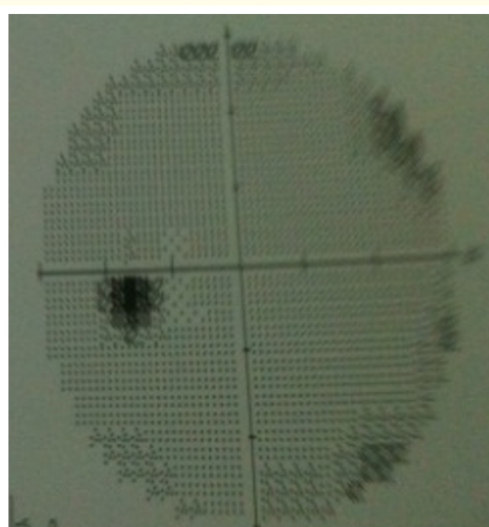


Figure 6B

Figure 6: Pre op (A) and Post op (B) picture of visual field of left eye showing significant improvement.

Pre-operative Visual status	No. of Cases	Percent	Post-operative Visual status	No. of Cases	Percent
Bi temporal field effect	22	55 %	Improved	25	62.5 %
Unilateral blindness and Contra lateral temporal field effect	10	25 %	Not improved/ Static	10	25 %
Bi lateral blind effect	8	20 %	Deteriorated	5	12.5 %

Table 3: Preoperative and postoperative visual status (N = 40).

All patients were complicated by temporary D.I which became permanent D.I in 10 cases. Post-operative VP Shunt was done 4 cases.

Complication	No. of cases	Percentage
C.S.F leak	10	25.00%
Meningitis	8	20.00%
Post op VP Shunt	4	10.00%
Pneumocephalus	5	12.5%
Temporary D.I	40	100%
Permanent D.I	10	25.00%
Hyponatremia	4	10.00%
Hypernatremia	5	12.5%
Septicemia	6	15.00%
Tumor Bed Hematoma	1	2.50%

Table 4: Table of complication (N = 40).

Most of the patients had good recovery 26 cases (65.00%). Moderately disabled (GOS-4) were 6 cases (15.00%). Severely disabled (GOS-3) were 2 cases. No patients had GOS score of 2. Six cases (15.00%) had GOS-1. See table 5.

GOS Outcome	No. cases	Percentage
GOS-5 (Good Recovery)	26	65.00%
GOS-4 (Moderately Disabled)	6	15.00%
GOS-3 (Severely Disabled)	2	5.00%
GOS-2 (Vegetative Scale)	00	00%
GOS-1(Dead)	6	15.00%

Table 5: Post-operative outcome (N = 40).

Endocrine outcome

Pre-op endocrine presentation: Typical endocrinological findings were hypocortisolism, hypothyroidism and hypogonadism were found in most of the cases. Some of the common clinical findings are shown in table 6.

Symptoms	No of patients
Visual symptoms	40
Neurological symptoms	40
Endocrine symptoms	25
General symptoms (lassitude, depression)	1

Table 6: Common clinical presentations.

Post-op endocrine outcome: Postoperative anterior pituitary dysfunction improved in 4 cases (10.00%) out of 40 cases. There were 100% cases of temporary D.I. and about 10 cases of permanent D.I.

Complications: Permanent DI developed in 10 (25.00%) patients, pneumocephalus in 5 patients (12.5%). Small amount of tumor bed hematoma in 1 patient (2.5%). Severe Hyponatremia developed in 4 patients (10.0%). Hydrocephalus developed in 4 patient (10.0%) for which VP shunt was inserted in all the 4 cases.

Discussion

The extended transsphenoidal approach extends operative exposure beyond the sella by removing the tuberculum sellae and a portion of the planum sphenoidale. The advantages of the extended transsphenoidal approach over a traditional craniotomy are the avoidance of frontal or temporal lobe retraction or sylvian fissure dissection and the potential associated brain injury [14].

Approximately transcranial cranial base procedures result in some form of retraction injury to the brain. However, it has generally been thought that an enlarged sella was required to safely reach a suprasellar lesion through a transsphenoidal approach [15].

In our study all 40 patients underwent a detailed neuro-ophthalmological examination before and after surgery. 25 (62.5%) of these patients had improved resolution of their visual defect and 10 (25.00%) had not improved but static. Five (12.5%) patient's deficit deteriorated. 16 of 19 patients visual disturbances improved while 3 remained static as described by Maira, *et al* [19].

Kassam, *et al.* showed that all 16 patients underwent a detailed neuro-ophthalmological examination before and after surgery. The conditions of 2 patients without preoperative visual deficits were unchanged postoperatively. The remaining 14 patients had progressive visual deficits preoperatively. Six (43%) of these patients had complete resolution of their visual defect, and seven (50%) had improvement but not complete resolution. One patient deficit remained stable. One patient had visual worsening 4 days after surgery secondary to hydrocephalus. His vision improved following the placement of a VP shunt [16].

In an international publication of endocrinological results of 30 patients of craniopharyngioma treated transcranially developed postoperative diabetes insipidus in 60% cases, adrenal failure in 53.3% and hypothyroidism in 36.7% cases [17].

Microscope-based removal of purely suprasellar craniopharyngiomas and meningiomas has been associated with a 20 to 33% rate of CSF leak. For craniopharyngiomas, postoperative rates of DI and panhypopituitarism occur in roughly 70%. In our study permanent DI developed 10 cases (25.00%) out of 40, using the endoscopic endonasal approach [18,19].

We did gross total resection of 22 cases (55.00%) of the craniopharyngiomas with an overall risk of postoperative CSF leak of 10 cases (25.00%). While 10 patient developed CSF leak in a series of 57 patients as described by Maira, *et al* [19].

We found that the visualization provided by the endoscope is outstanding for the extended approach to purely suprasellar pathology. This advantage can potentially minimize the risk of morbidity to vital neurovascular structures and also decrease the risk of CSF leakage because closure is more secure, aided by improved visualization [20,21].

Despite the minimally invasive approach and the use of the endoscope, these endoscopically treated cases are not without morbidity. However, removal of these lesions using a traditional microscope-based transcranial or transsphenoidal also has a potentially high morbidity and mortality rate. The success of this maneuver requires meticulous closure with dural graft inlay, either fascia lata or Dura-Guard, rigid buttressing with either vomer or a metal plate and the use of sealants, such as fibrin glue [22].

Conclusion

Extended trans-sphenoidal approach is an excellent alternative of skull base approach for the removal of selected group of the craniopharyngioma. The endoscopic endonasal route provides a good exposure, especially of the sub- and retrochiasmatic areas, as well as of the stalk-infundibulum axis, and the third ventricle chamber. It gives better visualization, improved postoperative visual outcome because of less manipulation of visual apparatus and low complication rate than craniotomy. However CSF leaks and D.I. are known common complications which have to be managed promptly and appropriately.

Bibliography

1. Cappabianca P, et al. "Extended Endoscopic endonasal transsphenoidal approaches to the suprasellar region, planum sphenoidale and clivus". In De Diviitis E, Cappabianca P (eds): Endoscopic Endonasal Transsphenoidal Surgery. New York, Springer-Verlag/Wien (2003): 176-188.
2. Cavallo LM, et al. "Endoscopic endonasal surgery of the midline skull base: Anatomical study and clinical considerations". *Neurosurgery Focus* 19.1 (2005): E2.
3. Frank G, et al. "Transsphenoidal endoscopic approach in the treatment of Rathke's cleft cyst". *Neurosurgery* 56.1 (2005): 124-129.
4. Chakrabarti I, et al. "Long-term neurological, visual, and endocrine outcomes following transnasal resection of craniopharyngioma". *Journal of Neurosurgery* 102.4 (2005): 650-657.
5. De Diviitis E, et al. "Endoscopic transsphenoidal approach: Adaptability of the procedure to different sellar lesions". *Neurosurgery* 51.3 (2002): 699 -705.
6. Dusick JR, et al. "The extended direct endonasal transsphenoidal approach for Non adenoma to us suprasellar tumors". *Journal of Neurosurgery* 102.5 (2005): 832- 841.
7. Fahlbusch R, et al. "Surgical Treatment of craniopharyngiomas: Experience with 168 patients". *Journal of Neurosurgery* 90.2 (1999): 237-250.
8. Jho HD and Ha HG. "Endoscopic endonasal skull base surgery. Part 1-The midline anterior fossa skull base". *Minimally Invasive Neurosurgery* 47.1 (2004): 1-8.
9. Kaptain GJ, et al. "Transsphenoidal approaches for extracapsular resection of midline suprasellar and anteriorcranial base lesions". *Neurosurgery* 49.1 (2001): 94-101.
10. Kassam A, et al. "Expanded endonasal approach: The rostrocaudal axis. Part 1. Crista galli to the sella turcica". *Neurosurgery Focus* 19.1 (2005): E3.
11. Kassam A, et al. "Expanded endonasal approach: The rostrocaudal axis. Part II. Posterior clinoids to the foramen magnum". *Neurosurgery Focus* 19.1 (2005): E4.
12. Kim J, et al. "Transsphenoidal Supra diaphragmatic intradural approach". *Minimally Invasive Neurosurgery* 43.1 (2000): 33-37.
13. Kitano M and Taneda M. "Extended transsphenoidal approach with submucosal posterior ethmoidectomy for parasellar tumors". *Journal of Neurosurgery* 94.6 (2001): 999-1004.
14. Landolt AM and Zachman M. "Results of transsphenoidal extirpation of craniopharyngioma". *Neurosurgery* 28.3 (1991): 410-415.
15. Laufer I, et al. "Endonasal endoscopic extended transsphenoidal, transplanum approach for suprasellar lesions". *Journal of Neurosurgery* 106.3 (2007): 400-406.
16. Amin B Kassam. "Outcomes following endoscopic, expanded endonasal resection of suprasellar craniopharyngiomas: a case series". *Journal of Neurosurgery* 109.1 (2008): 6-16.
17. Honegger J, et al. "Surgical treatment of craniopharyngiomas: Endocrinological results". *Journal of Neurosurgery* 90.2 (1999): 251-257.
18. Laws ERJ. "Transsphenoidal microsurgery in the management of craniopharyngioma". *Journal of Neurosurgery* 52.5 (1980): 661-666.
19. Maira G, et al. "The role of transsphenoidal surgery in the treatment of craniopharyngioma". *Journal of Neurosurgery* 100.3 (2004): 445- 451.
20. Mason RB, et al. "Selective excision of adenomas originating in or extending into the pituitary stalk with preservation of pituitary function". *Journal of Neurosurgery* 87.3 (1997): 343-351.
21. Page RB. "Craniopharyngioma: Indications for transsphenoidal surgery". *Current Therapy in Endocrinology and Metabolism* 5 (1994): 33-34.
22. Yasargil MG, et al. "Total removal of craniopharyngiomas. Approaches and long-term results in 144 patients". *Journal of Neurosurgery* 73.1 (1990): 3-11.

Volume 10 Issue 11 November 2018

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