

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

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Abstract:

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 supra sellar germinoma. Traditional transsphenoidal approach gives exposure to the pituitary fossa, whereas extended approach provides exposure to the optic nerve, chiasm, acon complex and basal frontal lobe, mammillary body, mid brain, 3rd nerve, basilar artery, and circle of Willis and laterally to the cavernous sinuses.

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 (C.S.F.) leak.

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. C.S.F. leak developed in 10 (25.00%) cases. All Patients required thyroxin and cortisol for replacement. Permanent diabetes insipidus (D.I.) developed in 10 cases (25.00%). No cases required traditional, open approaches following endoscopic resection. Three patients required permanent C.S.F 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 C.S.F leak compared with other tumor types in the same area.

Conclusion: Extended transsphenoidal 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 postoperative visual outcome for less manipulation and lower complication rate than craniotomy. However C.S.F. leak and D.I. are known common complications which have to be managed promptly and appropriately.

Bang. J Neurosurgery 2013; 3(1): 26-33

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 morbidity¹. As craniopharyngiomas grow, they can cause significant

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neurological complications, including visual loss, pituitary insufficiency, and hypothalamic damage, and recurrence. The first description 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 pouch³. 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. Distribution by age is bimodal, with the peak incidence in children at aged 5-14 years and in adults at age 50-75 years⁵. Craniopharyngioma accounts for 1%-3% of all brain tumors in adult, 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 structures⁷. 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}.

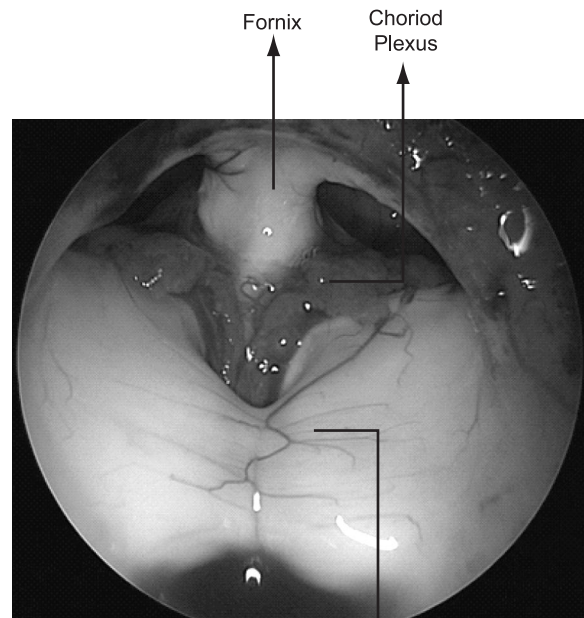


Fig.-1 (b)

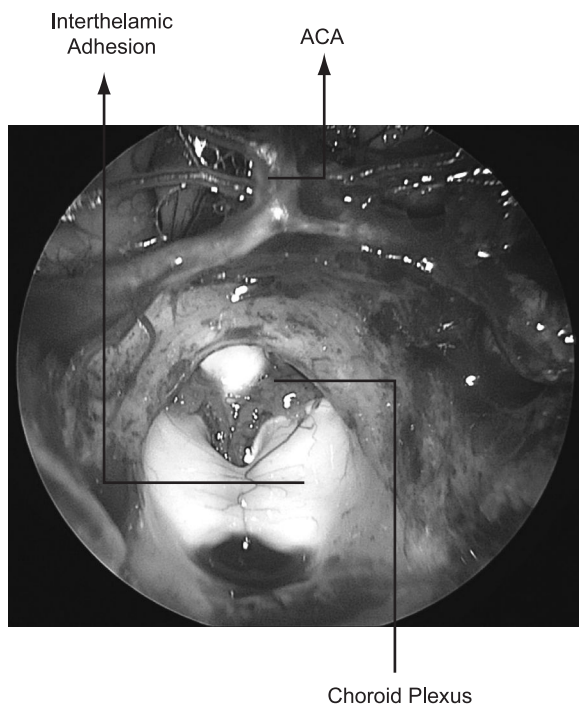


Fig.-1 (a)

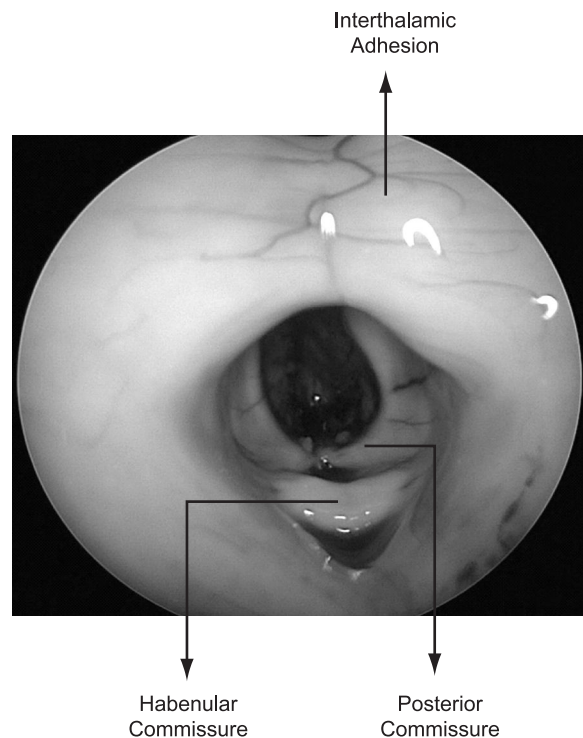


Fig.-1 (c)

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. Eighty-eight percent of men experience decreased sex drive, while 82% of women have amenorrhea. Other endocrine dysfunction may lead to precocious puberty and obesity⁸.

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 atrophy⁹.

Craniopharyngioma is surgically divided into 3 groups: seller, prechiasmatic, 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 transinfundibular (extending into the stalk); Type III is retroinfundibular, 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}. (Fig-2)

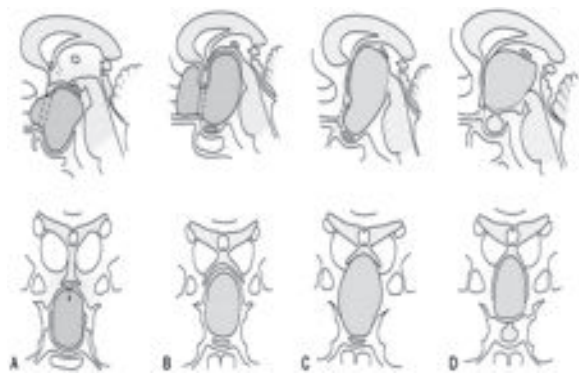


Fig.-2

Grading In relative to hypothalamus: In relation to hypothalamus, craniopharyngioma is divided in 3 grades. Grade 0 means no invention. Grade 1 means Abutting/displacing the hypothalamus. Grade 2 means Involving/Infiltrating the hypothalamus –marked by absence of hypothalamus on imaging. (Figure 3,a,b,c)

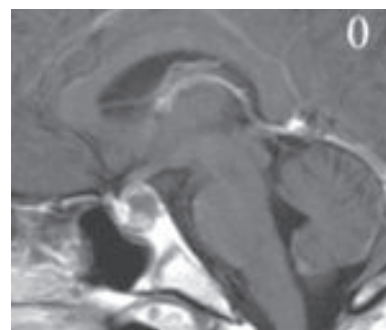


Fig.-3 (a): Grade 0 = No invention



Fig.-3 (b): Grade 1 = Abutting/displacing the hypothalamus.

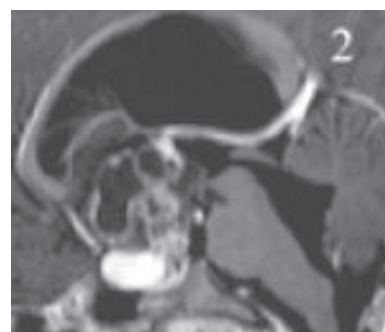


Fig.-3 (c): Grade 2 = Involving/Infiltrating the hypothalamus –marked by absence of hypothalamus on imaging

The arterial supply is usually from the anterior cerebral and anterior communicating arteries or from the internal carotid and posterior communicating arteries¹².

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 parenchyma¹³.

When predominantly in the sella, these tumors erode the bony floor and enlarge the sella¹³.

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 Monro¹⁴, interthalamic adhesion, habenular commissure, posterior commissure, aqueductal sylvius (Figure 1a, 1b and 1c).

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 supra sellar craniopharyngioma. Patient's history, clinical findings, preoperative & 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 C.S.F. 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 scan and MRI 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 prechiasmatic in 3 patient (7.5%), pre and retrochiasmatic in 4 patients (10.00%) (Table-I). In all the cases surgery was performed with the help of endoscopic extended endonasal approach.

Table-I
Types of Craniopharyngioma (N=40)

| Name of disease | | Number of Cases | Percentage |
|-------------------|--------------------------|-----------------|------------|
| Craniopharyngioma | Retrochiasmatic | 25 | 62.5% |
| | Subchiasmatic | 8 | 20.00% |
| | Prechiasmatic | 3 | 7.5% |
| | Pre and Retro-chiasmatic | 4 | 10.0% |
| Total | | 40 | 100% |

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 40 patients underwent gross-total resection (Table-II). Ten cases (25.00%) of the patients underwent sub-total. All the twelve patients were followed-up with early postoperative CT scan (Figure 4a, 4b, 4c and 4d) and neurological evaluation.

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. Six cases mortality in 40 cases caused by C.F.S leak, meningitis, meningoencabathalitis and septicemia. This patient was treated by antibiotic therapy and lumbar drain for C.S.F. leak.

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 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-II).

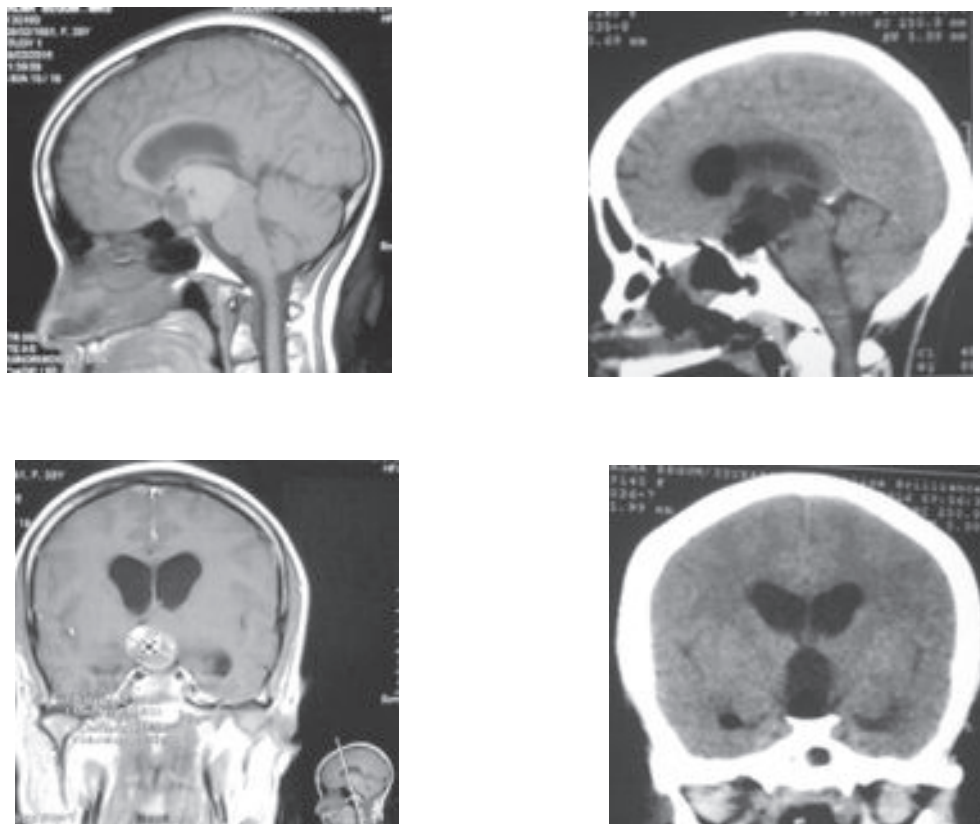


Fig.-4: (a) suprasellar craniopharyngioma(pre-op) Sagittal view, (b): suprasellar craniopharyngioma (post op), (c): suprasellarcraniopharyngioma (pre op)coronal view, (d): suprasellarcraniopharyngioma(post op)

Table-II

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

| Size of Tumor(cm) | No. of Cases | Percent | Extent of Tumor Removal | No. ofCases | 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 % |
| Total | 40 | 100% | Total | 40 | 100% |

Post-op visual outcome: Visual improvement was satisfactory. Postoperative visual acuity and visual field improved in 25 cases (62.5 %) (Table-III) (Figure 3a, 3b and Figure 4a, 4b). 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.

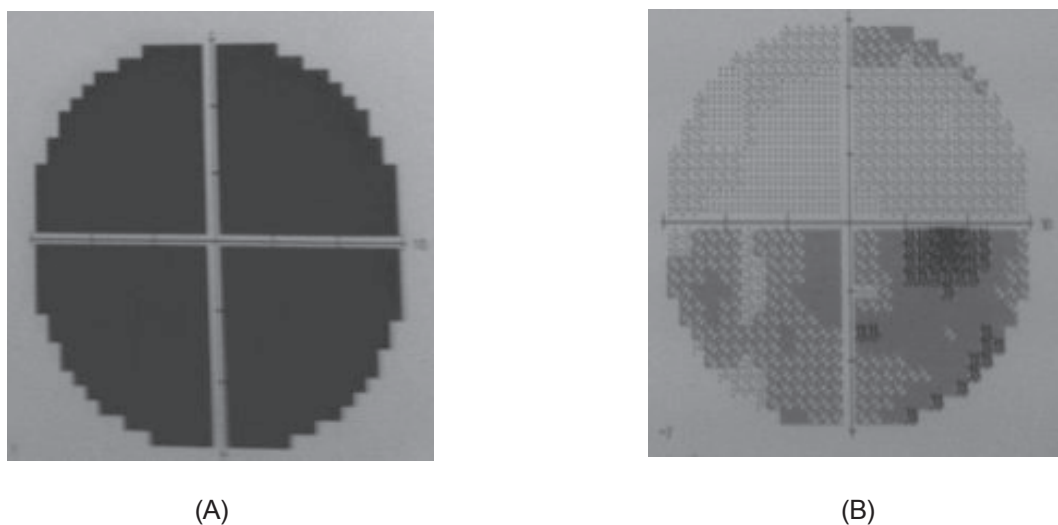


Fig.-5: Pre op (A) and Post op (B) picture of visual field of right eye showing significant improvement.

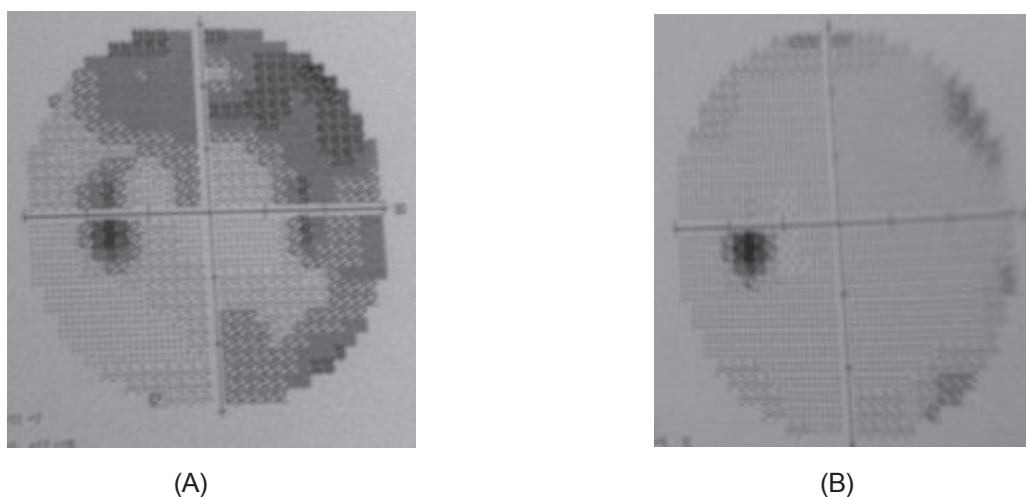


Fig.-6: Pre op (A) and Post op (B) picture of visual field of left eye showing significant improvement.

Table-III
Preoperative & Postoperative Visual Status (N=40)

| 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 % |
| Total | 40 | 100% | Total | 40 | 100% |

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

Table-IV
Table of Complication (N=40)

| 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 % |
| Total | 40 | 100 % |

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 V

Table-V
Post Operative Outcome (N=40)

| 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 % |
| Total | 40 | 100% |

Endocrine outcome:

Pre-op endocrine presentation: Typical endocrinological findings were hypocortisolism, hypothyroidism and hypogonadism were found in most of the cases.

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 injury¹⁴.

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 approach¹⁵.

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.

In an international study by Amin B. Kassam et al, shows in 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 shunt¹⁶.

In an international publication of endocrinological results of 30 patients of craniopharyngioma (Honegger et al. 1999) treated transcranially developed postoperative diabetes insipidus in 60% cases, adrenal failure in 53.3% and hypothyroidism in 36.7% cases¹⁷.

Microscope-based removal of purely suprasellar craniopharyngiomas and meningiomas has been associated with a 20 to 33% rate of C.S.F. 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 C.S.F. leak of 10 cases (25.00 %).

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 C.S.F. 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 glue²².

Conclusion: Extended transsphenoidal 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 C.S.F. leaks and D.I. are known common complications which have to be managed promptly and appropriately.

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