

Endoscopic Approach in the Treatment of Craniopharyngiomas

Bouaita.K- L, Atroune, Benmamar.T-Shabhay.Z

EHS Cherchell Département of Neurosurgery

*Corresponding author: Bouaita.K- L, Atroune

Abstract:

Objective: Craniopharyngiomas are epithelial tumours developing from Rathke's pouch. It represents 1% to 4% of all primitive intra cranial tumours and 5% to 10% in children. The objective of our work is to show the utility of endoscopy in craniopharyngioma surgery and we will show detailed results of our series and a review of the literature.

Patients and methods: This is a retrospective study of 22 patients with craniopharyngiomas operated by transsphenoidal endoscopic endonasal approach, extending from 2010-2016. The mean age was 34 years. The sex ratio was 9 females/13 males. Clinical signs were dominated by opto-chiasmatic syndrome with visual deficits in 90% of patients. Endocrinal syndrome was found in 15 patients. All patients had an MRI preoperatively.

Results: maximum tumour resection was obtained in 68.2% of cases (15 cases) while sub total and partial resection was found in 31.8% of cases (07 patients).

A visual function improvement was noted in 64.7% of patients, remained unchanged in 24.5% and worsened in 13.8% of patients.

Diabetes insipidus was established in 36% of cases. CSF leak rate was estimated at 18% with a 9% meningitis rate; likely related to CSF leakage during the postoperative period. The mortality rate was zero. Tumor recurrence was marked in 36% of patients on a follow-up of 5 years.

Conclusion: Extended endoscopic endonasal approach in treating craniopharyngiomas is a safe and effective alternative to the transcranial approach in pre-selected patients.

Key Words: craniopharyngioma, endoscopic approach, skull base, total excision.

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I Introduction

Craniopharyngiomas are epithelial tumours that develop from the Rathke's pouch. They may extend to intra- and / or parasellar spaces. Until recently, only transcranial (sub-frontal, pteronal, transcallosal or combinations) approaches were used for the resection of these tumors, but these techniques necessitate some degree of manipulation of adjacent cerebrovascular structures. This led to the development of a new type of surgical procedure called extended endoscopic endonasal surgery (extended endoscopic endonasal approach.) We present in this paper our experience in endoscopic endonasal surgery of craniopharyngiomas.

II Materials And Methods:

This is a retrospective study of 22 patients with craniopharyngiomas operated by transsphenoidal endoscopic endonasal approach, extending from 2010-2016. The mean age was 34 years. The sex ratio was 9 females/13 males. The time to consultation was between 09 months and 2 years after onset of symptoms. All the patients were investigated in Neurosurgical and Endocrinological Departments. Visual acuity and visual field defects and cartographic fields were tested in Ophthalmology department pre and post-operatively during each visit. Hormonal investigations included T4, TSH, blood Cortisol, follicle-stimulating hormone (FSH), luteinizing hormone (LH), prolactin and Growth hormone (GH) for all patients pre and post-operatively. Clinical signs were dominated by opto-chiasmatic syndrome with visual deficits in 90% of patients. Diminution of visual acuity was the major symptom. Visual field deficits were observed in lesions compressing the optic chiasma with retro chiasmatic extension (classic bitemporal in 10 patients). Endocrinal syndrome was found in 15 patients. All patients had MRI preoperatively in precise sagittal, coronal and frontal cuts; the form, size of craniopharyngioma, the extent of the tumor and its close relationship with both supra-clinoid internal carotid arteries, the lateral extent of the lesion to the two cavernous sinuses and sub-frontal region. The magnetic resonance imaging in coronal cuts can show the retro chiasmatic extension of the tumor towards the foramen of Monro is responsible for a bi ventricular hydrocephalus. In addition, it shows the extension of the lesion in the sphenoid sinus to the clivus. The CT scan is essential especially in coronal and axial cuts. It retains a significant interest in the study of the sphenoid sinus morphology, the existence of any intra-sinus walls or pathology of the

mucosa (important in endoscopic endonasal approach). In the immediate postoperative period we performed axial cuts CT scan to eliminate a surgical cavity hematoma or cerebral edema, postoperative MRI 3-6 months would be repeated annually thereafter.

The approach was preferred because it is a direct mid-line approach with a panoramic view (better visualization of optical pathways and internal carotid arteries), limiting as much as possible the manipulation of optical pathways (optic nerve and chiasma) and allowing relatively easy access and resection of the infra and retro-chiasmatic lesions as well as in the interturbinate cistern. The approach was binasine, with resection of the right middle turbinate, bilateral posterior ethmoidectomy, wide sphenoidotomy and ablation of the nasal septum behind. We have closed using the naso-septal pedicle flap technique (Kassam's method).

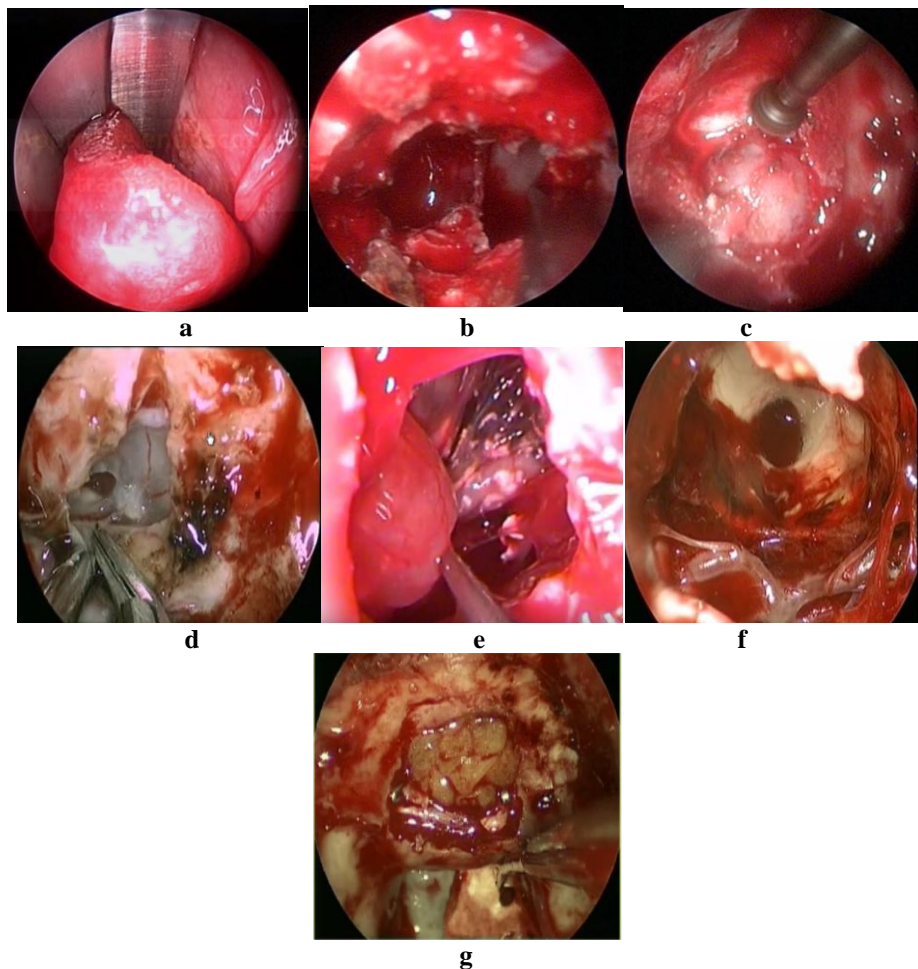


fig1 :stages or the surgical procedure:

a : resection of the right middle turbinate

b : extended anterior sphenoidotomy

c : drilling of the tuberculum sellae after opening the floor of the sella turcica

d : opening of the dura mater and tumor exposure.

e : tumor resection.

f : visualization of 3rd ventricular floor and foramen of Monro after complete tumor resection.

g : closure.

III Results:

▲ Quality of resection: postoperative imaging (CT or MRI) was used to assess the quality of excision or extent of surgical resection, the majority of patients underwent immediate postoperative brain CT and remote MRI to determine the quality of surgical resection.

The maximum resection of the tumor was obtained in 68.2% of the cases (15 cases) while the total and partial resection was found in 31.8% of the cases (07 patients).

▲ **Visual Results:** The visual function analysis was first performed in the immediate postoperative period looking for a visual improvement or worsening by the neurosurgeon himself, followed by the ophthalmologist in the short and long term.

A visual function improvement was noted in 64.7% of patients, remained unchanged in 24.5% and worsened in 13.8% of patients. The visual field defect was improved after surgery in ten patients.

▲ **Endocrine Results:** The endocrinological evaluation in the immediate postoperative period found diabetes insipidus in eight patients, pan-hypopituitarism in 03 patients who needed replacement therapy postoperatively. 03 patients are still under treatment with Minirin for permanent diabetes insipidus.

All patients are monitored regularly by the Department of Endocrinology clinically, radiologically and by regular hormonal analysis.

▲ **Morbidity:** the postoperative period was marked by the following complications:

Diabetes insipidus was found in 36% of cases. The CSF leak rate was estimated to be 18% with a meningitis rate at 9% of cases probably related to CSF leakage during the postoperative period. The mortality rate was zero. Tumor recurrence was marked in 36% of patients on a follow-up of 5 years.

▲ **Postoperative Follow up:** All patients were regularly followed up clinically and radiologically by the neurosurgeon, endocrinologist and ophthalmologist.

The follow-up period ranged from a 1 to 5 years. All patients with residual tumor underwent radiological examinations especially brain MRI to assess the evolutionary potential of the lesion and to act accordingly.

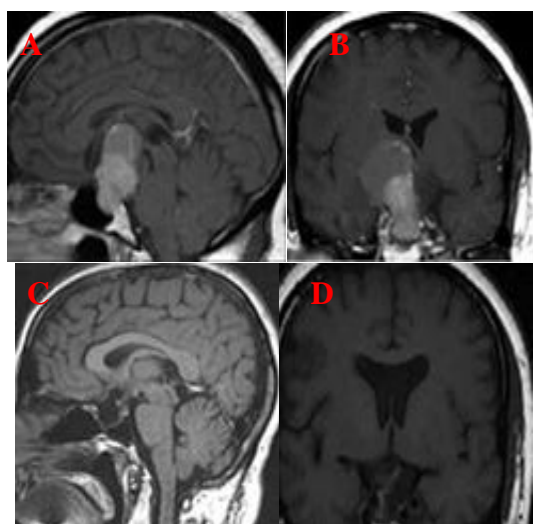


Fig2 : A-B : MRI images in sagittal and coronal sections show preoperative intrasellar and suprasellar craniopharyngioma.

C-D : MRI images in sagittal and coronal postoperative sections show the quality of satisfactory resection.

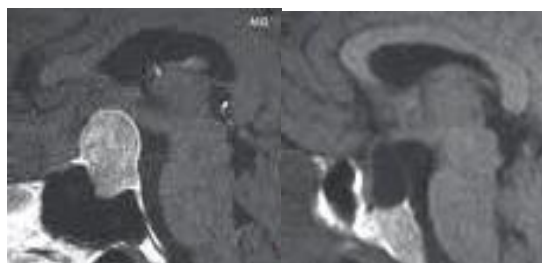


Fig 3: A-B : MRI image in sagittal section shows preoperative intrasellar and suprasellar craniopharyngioma.

C-D : MRI image in sagittal section postoperatively shows the quality of satisfactory resection.

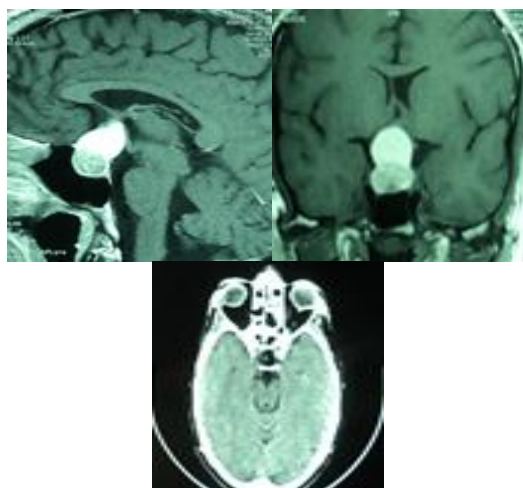


Fig 4: A-B: MRI image in sagittal section show a preoperative intrasellar and suprasellar craniopharyngioma. C-D: CT image in axial section postoperatively shows the quality of satisfactory resection.

IV Discussion :

Craniopharyngiomas are epithelial tumors which are derived from Rathke's pouch and develop in the course of the craniopharyngeal canal. They account for 1% to 4% of all primary intracranial tumors, and 5% to 10% in children. According to the literature, there is a bimodal age distribution with two peaks in children aged 0 to 19 years and in adults aged 40 to 79 years with a uniform distribution between the two sexes(2, 3, 4). In our series, the average age is 34 years with a slight male predominance.

There are two main histological types of craniopharyngiomas, the adamantine type is more common in children and the papillary type is found almost exclusively in adults. The craniopharyngioma is considered a grade I tumor according to WHO 2016(5). Their diameter varies from 2 to 4 cm, but can be significantly greater. They often have irregular contours with a cystic component (46% to 64%). The adamantine type is frequently calcified. Craniopharyngiomas from the pituitary stalk tend to extend mainly to the sellar region and then develop towards the parasellar region. On the other hand, tumors from tuberumereum extend mainly upwards towards the hypothalamus and back into the third ventricle.

Kassam et al proposed a new classification taking into account the relationship between the infundibulum and the tumor, thus identifying 4 categories: type I, pre-infundibular; Type II, transinfundibular; Type III, retro-infundibular; And type IV, intraventricular(6).

In our series, type I is the most frequent in 11 cases (50%), retro-infundibular in 07 cases (31.8%) with a large cystic portion extending to the interpeduncular cistern and 04 intraventricular cases extending in the third ventricle through the foramen of monro.

The clinical presentation is dominated initially by endocrine disorders (endocrine pallor, diabetes insipidus, hormonal imbalance), then visual disorders by compression of chiasma and optic nerves, memory disorders, disorders of behavior by hypothalamic attack, and finally symptoms related to intracranial hypertension due to the extension of the tumor in the third ventricle. Albert E. Halsted was credited with the first successful trans-sphenoidal resection of a craniopharyngioma performed in 1909(7). Gardner mentioned that Cushing had abandoned this approach for security reasons, viewing limitations. Hardy stressed the importance of the approach and mentioned that the subdiphagmatic type could be totally resected transphenoidally(8). The Pittsburgh group with Carrau, Kassam, and the Napoli group with Cappabianca and De Divitiis, as well as the Bologna group with Frank and Pasquini, all favored the endoscopic endonasal approach at the beginning of the 21st century(9,10, 11, 12). This approach provides direct access to the base of the skull, the suprasellar and parasellar regions, the interpeduncular cistern and the third ventricle, allowing direct visualization of the optic pathways and the pituitary stem while avoiding the complications associated with brain retraction maneuvers.

One of the most important considerations is the position of the optic chiasma with respect to the tumor. If the tumor is retrochiasmatic, pushing the chiasma anteriorly (prefixed chiasma), the endoscopic endonasal approach offers better access to the lesion by avoiding the excessive manipulation of the chiasma, while knowing that in most craniopharyngiomas there is a prefixed chiasma. However, if the tumor has a significant lateral extension (more than 1 cm laterally with respect to the internal carotid artery), the endoscopic endonasal pathway seems to be less indicated. Komotar et al. conducted a review of the literature including 88 studies, involving 3470 patients, extending from 1995 to 2010(13). Endonasal approaches had significantly higher total tumor excision rates than after transcranial surgery (66.9% vs 48.3%), an improvement in visual results (56.2% vs 33.1%) and fewer recurrences. There was a higher rate of CSF leakage after endoscopic surgery (18.4% vs.

2.6%), but CSF leakage rates of 0% to 4% were reported using the new technique for reconstructing the base of the skull(14, 15).Koutourousiou et al in a retrospective study extending from

1999 to 2011[16]showed that the rate of total excision after endoscopy was only 37,5%. Visual improvement was 86.4% with 0% permanent visual deterioration.

18-67% of patients showed panhypopituitarism and 8-48% had permanent diabetes insipidus. The obesity rate at diagnosis was 43.8%, while only 11.1% of the patients were obese in the postoperative period. The rate of CSF leakage decreased to 23.4% after introduction of nasoseptal pedicle flap(17).

Postoperative hydrocephalus requiring treatment was 12.7% (VP shunt). The perioperative mortality rate was 0%. The recurrence rate was 34.4%. Zacharia et al demonstrated a transcranial approach, patients undergoing endoscopic resection recorded higher rates of total tumor excision (66.9% vs 48.3%) (18).This is also valid for the improvement of the visual function (56.2% vs. 33.1%). Permanent diabetes insipidus developed in 42% of patients and panhypopituitarism in 38%. These rates are compatible with those of the endoscopic and transcranial literature. The rate of CSF leakage was higher in the endoscopic group than in the transcranial group (18.4% vs 2.6%).

Thanks to the new skull base reconstruction technique, we achieved a CSF leakage rate of 3.8% (19). 69% of patients have entered their profession and their level of education, which corresponds to the literature reports. Wannemuehler et al carried out a retrospective study from 2005 and 2015. Only 25% of patients after transcranial surgery had visual improvement and 88.9% after endoscopic surgery. Visual deterioration occurred only in 25% of patients after transcranial surgery. Permanent diabetes insipidus affected half of the patients without significant difference between the two groups (55.5%). Panhypopituitarism was 16.7% after transcranial surgery and 33.3% after endoscopic surgery. CSF leakage occurred only after endoscopic surgery (22.2%). None of the patients had postoperative hydrocephaly. Tumor recurrence occurred in only one patient in each group (20)

Fomichevet al in a series of 136 patients showed that complete excision had been completed in 72% of the cases. 20% of patients had a tumor recurrence, 22 of them had undergone radiotherapy under stereotactic conditions. Improvement of vision or absence of visual deterioration was observed in 89% of cases. 52% of patients with preoperative visual field deficits improved. The deterioration of vision in this group was 5%. Failure of the hypothalamic-pituitary axis (previous failure or diabetes insipidus) was observed in 42.6% of cases. 8.8% of patients had CSF leaks. Mental disorders were observed transiently in 11% of cases, and permanently in 1.5% of cases. Mortality was 5.8%.

In our study, the maximum resection of the tumor was obtained in 68.2% of the cases (15 cases), while the total and partial resection was found in 31.8% of the cases (07 patients).

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V Conclusion

Extended endoscopic endonasal approach to treating craniopharyngiomas is a safe and effective alternative to the transcranial approach in previously selected patients. Although this technique is associated with more than satisfactory tumor excision and improved visual results, CSF leakage and endocrine dysfunction will remain a major challenge.

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