Craniopharyngioma case series

Kim et al. retrospectively reviewed clinical features, surgical outcomes, and complications in children who underwent an endoscopic endonasal approach for intracranial and skull base tumors at the Seoul National University Hospital from July 2010 to October 2018.

A total of 82 patients underwent EESs for 77 intracranial and 5 skull base bony tumors. The mean age at diagnosis was 11.4 years (range 4-18 years), and the mean follow-up period was 46.8 months. The most common tumors were craniopharyngioma in the intracranial tumor and skull base chordoma. Gross total resection was the goal of surgery in 55 patients and achieved in 90.9%. The vision was improved in 76.1% of patients with visual impairments. Preoperatively, various endocrinological deficiencies were revealed in 73.7% of 76 patients with hypothalamus-pituitary lesions, and the hyposomatotropism was most common. Endocrinological status was improved only in 10. Aseptic meningitis or bacterial meningitis (7.3%) was the most common surgical complication, and the cerebrospinal fluid leakage rate was 2.4%.

The endoscopic endonasal approach provides favorable neurological outcomes with acceptable risk for children with pediatric intracranial tumors. The high incidence of endocrinological deficits in cases with hypothalamus-pituitary lesions emphasizes the importance of judicious pre- and postoperative evaluation.

Guo et al., retrospectively investigated 185 cases of children with craniopharyngioma who underwent neurosurgical treatment at the Beijing Tiantan Hospital from 2011 to 2016. The neuroendocrine function of patients was compared before and after tumor removal. Results Compared with the MEIS, the incidence of growth hormone insulin-like growth factor 1 axis dysfunction (47.03% vs. 57.30%), pituitary-thyroid axis dysfunction (20.00% vs. 50.27%), pituitary-adrenal axis dysfunction (18.38% vs. 43.78%) and diabetes insipidus (26.49% vs. 44.86%) was significantly increased in the ANS status. The incidence of hyperprolactinemia significantly decreased from 28.11% in the MEIS status to 20.54% in the ANS status. Compared with the MEIS group, changes in appetite, development of diabetes insipidus, body temperature dysregulation, sleeping disorders, personality abnormalities and cognitive abnormalities were more frequent after ANS, yet no statistically significant differences were found. Conclusions Endocrine dysfunction is common in children with craniopharyngioma. Both MEIS and ANS can be harmful to neuroendocrine function, and neurosurgical treatment may increase the level of neuroendocrine dysfunction.

Pascual et al. from the University Hospital La Princesa, Puerta de Hierro University Hospital, Ramón y Cajal University Hospital, Hospital del Sureste, Madrid, Spain. Department of Surgery and Experimental Medicine, University of Ferrara, Italy. Independent Medical Translator, Jenkintown, PA, US. School of Medicine, Technische Universität, Dresden, Germany. Statistics Department, Computing Center, C.S.I.C. Madrid, Spain. investigated a collection of 210 craniopharyngiomas published from 1823 to 2017 providing detailed clinical and pathological information about psychiatric disturbances, and compared the hypothalamic damage in this cohort with the present in a control cohort of 105 cases without psychiatric symptoms.

Psychiatric disorders occurred predominantly in patients with craniopharyngiomas developing primarily at the tuberoinfundibular region (45%) or entirely within the third ventricle (30%), mostly
affecting adult patients (61%, p<0.001). Most tumors without psychic symptoms developed beneath the third ventricle floor (53.5%, p<0.001), in young patients (57%, p<0.001). Psychiatric disturbances were classified in six major categories: i) Korsakoff-like memory deficits, 66%; ii) behavior/personality changes, 48.5%; iii) impaired emotional expression/control, 42%; iv) cognitive impairments, 40%; v) mood alterations, 32%; and vi) psychotic symptoms, 22%. None of these was associated with hydrocephalus. Severe memory deficits occurred with damage of the mammillary bodies (p<0.001). Mood disorders occurred with compression/invasion of the third ventricle floor and/or walls (p<0.012). Coexistence of other hypothalamic symptoms such as temperature/metabolic dysregulation or sleepiness favored the emergence of psychotic disorders (p<0.008). Postoperative psychiatric outcome was better in strictly intraventricular craniopharyngioma than in other topographies (p<0.001). A multivariate model including the hypothalamic structures involved, age, hydrocephalus and hypothalamic symptoms, predicts the appearance of psychiatric disorders in 81% of patients.

CPs primarily involving the hypothalamus represent a neurobiological model of psychiatric and behavioral disorders 3).

2017

Wijnen et al. from Rotterdam studied a single-centre cohort of 128 patients with craniopharyngioma treated from 1980 onwards (63 patients with childhood-onset disease). Median follow-up since presentation was 13 years (interquartile range 5-23 years). Initial craniopharyngioma treatment approaches included gross total resection (n=25), subtotal resection without radiotherapy (n=44), subtotal resection with radiotherapy (n=25), cyst aspiration without radiotherapy (n=8), and 90Yttrium brachytherapy (n=21).

Pituitary hormone deficiencies (98%), visual disturbances (75%), and obesity (56%) were the most common long-term health conditions observed. Different initial craniopharyngioma treatment approaches resulted in similar long-term health effects. Patients with childhood-onset craniopharyngioma experienced significantly more growth hormone deficiency, diabetes insipidus, panhypopituitarism, morbid obesity, epilepsy, and psychiatric conditions compared with patients with adult-onset disease. Recurrence-/progression-free survival was significantly lower after initial craniopharyngioma treatment with cyst aspiration compared with other therapeutic approaches. Survival was similar between patients with childhood- and adult-onset craniopharyngioma.

Long-term health conditions were comparable after different initial craniopharyngioma treatment approaches and were generally more frequent in patients with childhood- compared with adult-onset disease 4).

2016

A retrospective analysis of the clinical data of 92 children who underwent surgical treatment from May 2011 to January 2005. Long-term follow-up was performed from 12 months to 8 years. The pterional approach was used in 49 patients, the interhemispheric approach in 20 patients, the corpus callosum approach in 16 patients, and the butterfly approach in 7 patients. Pathological classification was performed by hematoxylin and eosin stain staining of the pathological tissues and evaluated according to the different surgical approaches, MRI calcification status, calcification type, pathological type, whether radiotherapy was performed, postoperative recurrence, and death.

For the pterion approach resection, there was near total resection in 46 patients (93.9%) with the lowest recurrence rate. The operative approach and postoperative recurrence rates were compared; the difference was statistically significant (P<0.05). For comparison of the operative approach and postoperative mortality, the difference was not statistically significant (P>0.05). There was not a
significant difference between the MRI classification and postoperative recurrence rate (P > 0.05). Comparing the degree of tumor calcification with the recurrence rate after operation and the mortality rate, the difference was statistically significant (P < 0.05). The recurrence rate and mortality rate of adamantimous craniopharyngioma and squamous papillary craniopharyngioma in 2 groups following operation were compared, and the differences were statistically significant (P < 0.05). Postoperative adjuvant radiotherapy was compared with the postoperative recurrence rate and mortality; the differences were statistically significant (P < 0.05).

The main effects on tumor recurrence include the choice of surgical approach and degree of calcification. The adamantimous craniopharyngioma relapse rate is higher, which could be because invasion of craniopharyngioma only occurs with adamantimous craniopharyngioma. Postoperative radiotherapy can significantly prolong the recurrence time and reduce the mortality rate of patients with craniopharyngioma.

Clinical data from 226 consecutive patients with primary CP were retrospectively reviewed. Tumor location and the relationship of the tumor to the third ventricle floor and the pituitary stalk were evaluated using preoperative MRI and intraoperative findings. A topographic classification scheme was proposed based on the site of tumor origin and tumor development. The clinical relevance of this classification on patient presentation and outcomes was also analyzed.

The growth of CPs can be broadly divided into 3 groups based on the site of tumor origin and on tumor-meningeal relationships: Group I, infrasellar/infradiaphragmatic CPs (Id-CPs), which mainly occurred in children;

Group II, suprasellar subarachnoid extraventricular CPs (Sa-CPs), which were mainly observed in adults and rarely occurred in children; and Group III, suprasellar subpial ventricular CPs (Sp-CPs), which commonly occurred in both adults and children. Tumors in each group may develop complex growth patterns during vertical expansion along the pituitary stalk. Tumor growth patterns were closely related to both clinical presentation and outcomes. Patients with Sp-CPs had more prevalent weight gain than patients with Id-CPs or Sa-CPs; the rates of significant weight gain were 41.7% for children and 16.7% for adults with Sp-CPs, 2.2% and 7.1% for those with Id-CPs, and 12.5% and 2.6% for those with Sa-CPs (p < 0.001). Moreover, patients with Sp-CPs had increased hypothalamic dysfunction after radical removal; 39% of patients with Sp-CPs, 14.5% with Id-CPs, and 17.4% with Sa-CPs had high-grade hypothalamic dysfunction in the first 2 postoperative years (p < 0.001).

The classification of CPs based on growth pattern may elucidate the best course of treatment for this formidable tumor. More tailored, individualized surgical strategies based on tumor growth patterns are mandatory to provide long-term tumor control and to minimize damage to hypothalamic structures. Differences in the distribution of growth patterns between children and adults imply that hierarchical comparison is necessary when investigating outcomes and survival across treatment paradigms in patients with CP.

2014

103 patients underwent the endoscopic endonasal approach at two institutions (Division of Neurosurgery of the Università degli Studi di Napoli Federico II, Naples, Italy, and Division of Neurosurgery of the Bellaria Hospital, Bologna, Italy), between January 1997 and December 2012, for the removal of infra- and/or supradiaphragmatic craniopharyngiomas. Twenty-nine patients (28.2%) had previously been surgically treated.
The authors achieved overall gross-total removal in 68.9% of the cases: 78.9% in purely infradiaphragmatic lesions and 66.3% in lesions involving the supradiaphragmatic space. Among lesions previously treated surgically, the gross-total removal rate was 62.1%. The overall improvement rate in visual disturbances was 74.7%, whereas worsening occurred in 2.5%. No new postoperative defect was noted. Worsening of the anterior pituitary function was reported in 46.2% of patients overall, and there were 38 new cases (48.1% of 79) of postoperative diabetes insipidus. The most common complication was postoperative CSF leakage; the overall rate was 14.6%, and it diminished to 4% in the last 25 procedures, thanks to improvement in reconstruction techniques. The mortality rate was 1.9%, with a mean follow-up duration of 48 months (range 3-246 months).

The endoscopic endonasal approach has become a valid surgical technique for the management of craniopharyngiomas. It provides an excellent corridor to infra- and supradiaphragmatic midline craniopharyngiomas, including the management of lesions extending into the third ventricle chamber. Even though indications for this approach are rigorously lesion based, the data in this study confirm its effectiveness in a large patient series.

2010

From 1990-2008, 90 patients (64 adults and 26 children) underwent standard transsphenoidal surgery (TSS) for craniopharyngioma (34 subdiaphragmatic and 56 supradiaphragmatic). TSS was performed as the initial surgery in 62 patients and as the second procedure in 28 patients.

Total tumor removal was achieved in 70 (77.8%) patients, subtotal removal was achieved in 17 (18.9%), and partial removal was achieved in 3 (3.3%). Total removal was more often accomplished in initial surgery (56 of 62 [90.3%]) than second surgery (14 of 28 [50.0%]). Postoperative deterioration of anterior pituitary hormones developed in 31 of 47 (66.0%) patients with preoperative normal function or partial anterior pituitary loss. New-onset postoperative diabetes insipidus (DI) developed in 35 of 67 (52.2%) patients. Of 61 patients with preoperative visual loss, 55 (90.2%) noted some degree of visual improvement after surgery. The early postoperative mortality rate was 2.2% (2 of 90 patients). Cerebrospinal fluid (CSF) leakage occurred in 11 patients (12.2%), and 5 patients required surgical repair of the leak. Tumor recurrence was observed in seven (7.8%) patients during a mean follow-up period of 4.6 years.

Most craniopharyngiomas including the supradiaphragmatic type can be removed safely by TSS with a good outcome, although endocrine function frequently worsens after surgery. Dural fascia graft is a very effective technique to prevent CSF leaks, especially after eTSS.

14 patients underwent a purely endoscopic, endonasal approach for resection of newly diagnosed craniopharyngiomas. These procedures represent index surgeries; no patient had undergone previous tumor resection. A retrospective review of endocrinological and ophthalmological outcomes, extent of resection, and complication prevalence was completed. Additionally, a review of the English literature was performed to evaluate outcomes of similar endoscopic techniques for resection of craniopharyngiomas.

Four patients (28.6%) underwent gross-total resection; near total resection or better was achieved in 9 patients (64.3%). All patients presented with some form of visual field or acuity deficit. Postoperatively, 12 patients (85.7%) experienced visual improvement, with 6 patients (42.9%) having complete visual recovery. One patient experienced worsening of her visual deficit. Visual acuity improved in 8 patients (57.1%), while visual field defects improved in 11 (78.6%). The pituitary stalk
was preserved in all cases. Eight (57.1%) of 14 patients experienced some form of anterior pituitary dysfunction postoperatively. Although 9 patients (64.3%) were documented to have either transient or permanent new diabetes insipidus immediately after surgery, at 1-month follow-up only 1 patient met clinical criteria. Five patients (35.7%) developed CSF leaks that were successfully treated by subsequent endoscopic revision. All CSF leaks occurred early in the series. Two patients (14.2%) were treated for presumed meningitis postoperatively.

The endoscopic endonasal approach is a minimally invasive alternative to open transcranial approaches for select craniopharyngiomas. Similar to previous transcranial series, rates of endocrinopathy and gross-total resection were dependent upon the adherence of the tumor capsule to the hypothalamus, pituitary stalk, and associated vasculature. A review of the literature suggests that the results of the current series are similar to other published series on this topic.

2000

Eight patients who were surgically treated for craniopharyngiomas located exclusively within the third ventricle were considered. The initial symptoms were acute hydrocephalus in two cases, psychological disturbances in two, amenorrhea in two, headaches in one, and hypopituitarism in one. The diagnoses were established, in all cases except one, with magnetic resonance imaging. In all cases, the tumor completely filled the third ventricle.

Total removal of the lesion was achieved in seven cases. One patient underwent partial removal. In the immediate postoperative period, no major complications were observed. Five patients required replacement hormonal therapy. All patients returned to a normal life. Many months after surgery, two patients exhibited psychological disturbances and died, the first because of voluntary withdrawal of replacement therapy (12 mo after surgery) and the second because of a severe imbalance in body fluids and electrolytes, with a subsequent hyperosmolar coma (27 mo after surgery). Only one patient who underwent initial total removal experienced a small recurrence of the lesion (30 mo after surgery); after 3 years, the lesion exhibited unchanged size.

The translamina terminalis approach is a valid choice for the removal of purely intraventricular craniopharyngiomas. These tumors can be removed without significant sequelae related to the surgical approach. The proximity to the hypothalamus requires accurate neuroendocrine and electrolyte control in the postoperative period, in some cases even years after surgery.

An examination of the Charles Harrison Frazier papers at the College of Physicians of Philadelphia allowed the authors to identify 54 CPs that he had treated during his career. In the early 1910s, Frazier developed the subfrontal approach, which would become the primary surgical route to access these lesions, providing better control of the adjacent vital neurovascular structures than the transsphenoidal route hitherto used. Nevertheless, strong adhesions between CPs and the third ventricle floor, the major reason underlying Frazier's disappointing results, moved him to advocate incomplete tumor removal followed by radiotherapy to reduce both the risk of hypothalamic injury and CP recurrence. This conservative strategy remains a judicious treatment for CPs to this day.


