Radical excision of craniopharyngioma

Results in 20 patients

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A series is presented of 20 craniopharyngioma patients who were treated between 1977 and 1981. All 20 cases underwent radical tumor resection via a frontotemporal craniotomy, with a concomitant temporal tip resection. All operations were performed by the senior author. The operative mortality rate was 5%, and the major morbidity rate 22.2%. The average follow-up period was 3.1 years. The role of radical surgical extirpation in adults is emphasized: the results compare favorably with the current radiotherapeutic and more conservative surgical statistics.

KEY WORDS: craniopharyngioma • radical resection • temporal tip resection • radiation therapy

THE term "craniopharyngioma" was first used in 1931 by Frazier and Alpers and by Cushing in 1932. The first detailed autopsy account of such a tumor was given in 1857 by Zenker, when he described a cystic suprasellar lesion containing cholesterol crystals and squamous epithelium. Further histological evidence for the tumor which we today call "craniopharyngioma" was given 3 years later by Luschka in his description of squamous epithelial cells in the region of the infundibulum. Over the years, a number of other terms have been used to describe the same tumor, including "Rathke’s pouch tumor," "craniopharyngeal duct tumor," "craniopharyngeal fat tumor," "hemangioblastoma," "ameloblastoma," " adamantinoma," and "epidermoid tumor."

All of the above terms refer to the tumor described by Russell and Rubinstein as a suprasellar lesion, consisting of cells arranged in thin cords of uniform angulated columnar cells resting on a basement membrane. Of all tumors of the suprasellar region, few are as difficult to deal with as craniopharyngiomas or evoke such widely varying opinions among neurosurgeons. Most of the major surgical series collected over the last 20 years attest to the difficult operative management of these "benign" tumors and of their characteristic tendency to recur. They carry a poor prognosis. In early surgical experience without the aid of the surgical microscope, prolonged follow-up studies showed that these tumors usually recurred despite apparently complete resection. The encouraging results of radiotherapy published by Kramer, et al., led to the view that the optimum management consisted of minimal surgical intervention, aspiration of any associated cyst, and establishment of the diagnosis, followed by aggressive radiotherapy. Favorable results of surgery alone were described by Katz, who followed Matson’s patients, and by Sweet, who included a large number of adults in his series.

The series presented here includes adults almost exclusively; only four of the 20 patients were below the age of 20 years and only one was younger than 10 years old. The purpose of this presentation is to illustrate one operative approach to radical excision of these tumors.

Clinical Material and Methods

Patient Population

The 20 cases are not consecutive, but all were operated on by one of us (L.S.) between 1977 and 1981. The senior author has personally operated on over 60 tumors histologically verified as craniopharyngiomas, but radical microsurgical resection did not become the preferred treatment until 1977. Since then, over 90% of all procedures for craniopharyngiomas have been radical resections, the few exceptions being recurrent tumors operated on elsewhere, or massive cystic lesions...
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Extending into more than one cranial fossa and inaccessible by a single approach.

Most reports give no significant male or female predominance, but of the 20 cases in this series 14 were males and only six females. The age of patients at resection varied between 4 and 73 years, with 45% in the fourth and fifth decades (Fig. 1). The small number of pediatric cases reflects the referral pattern in London, with the associated Children's Hospital receiving the majority of the younger age group.

Symptomatology

Most of the patients (75%) presented with failure of vision and only 20% with raised intracranial pressure. The most prominent finding was bitemporal hemianopsia, which was present in 11 (55%) of the 20 cases. The only patient presenting with growth hormone deficiency and arrested growth was less than 10 years old. In patients over the age of 10 years, the most common symptoms were loss of libido secondary to decreased follicle-stimulating hormone (FSH) and luteinizing hormone (LH), and fatigue secondary to decreased thyroid-stimulating hormone (TSH), although detailed endocrine assessment was not invariably made prior to surgery.

Radiological Investigations

The standard radiological investigations for patients with a suspected suprasellar lesion at the time of most investigations in this series consisted of plain skull radiograms, computerized tomography (CT) scanning with and without contrast enhancement, four-vessel angiography under general anesthesia, and pneumoencephalography (PEG) with tomography of the anterior third ventricular region. Seventeen patients underwent preoperative PEG and, of these, 16 (94%) showed a mass in the suprasellar region (Fig. 2). The PEG was interpreted as positive in two cases when good quality CT scans were interpreted as normal (the CT scans were obtained on an older, third-generation, scanner without sagittal or coronal reconstructions, or metrizamide cisternography). In comparison with older CT technology, PEG was more sensitive in indicating masses in this region, but this is not necessarily the case for fourth-generation scanners with the addition of metrizamide contrast cisternography.

Angiography was deemed essential to delineate the vessels of the circle of Willis, and was used to plan the surgical attack. It did not, however, improve diagnostic accuracy beyond the CT or PEG studies (Fig. 3). Plain radiographs were interpreted as abnormal in the sellar region in eight of 20 cases (40%). Pathological calcification was present in 25% as in Banna's analysis. Sellar erosion or truncation of the dorsum sellae was present in an additional three cases. The CT scan was diagnostic of calcification in nine of the 20 cases with tumor in the suprasellar region, for a 45% incidence of calcification, whereas plain radiograms showed only a 25% incidence.

All patients have been followed at The National Hospital for lengths of time varying between 1.6 and 6.8 years. The average follow-up period was 3.1 years.

Surgical Approach

A wide variety of surgical approaches to suprasellar craniopharyngiomas have been recommended. This great variety of approaches reflects the differing goals, whether biopsy alone, biopsy with decompression of the optic apparatus, subtotal resection, or radical resection. The advent of newer CT methods, combined with PEG and high-quality angiography, enables a more precise surgical approach to be planned in advance rather than carrying out an exploratory procedure.
There are three generally accepted approaches: a transseptal transsphenoidal technique, a subfrontal technique, and a transtemporal technique. Extrapial, partially intra-arachnoid ("dumbbell") craniopharyngiomas and totally intrasellar extrapial, extra-arachnoid craniopharyngiomas lend themselves to transseptal transsphenoidal resection. These tumors seem to represent a more uncommon subgroup of craniopharyngiomas, and none of our cases were of this type. A second well recognized approach is the subfrontal approach. Tumors that lie well anteriorly, projecting forward without CT or PEG evidence of intrusion into the interpeduncular cisterns, lend themselves to this approach provided that the optic chiasm is placed posteriorly. If this approach is used for more posteriorly placed tumors (behind or above the chiasm), damage to the optic chiasm may occur. Vigorous retraction of the tumor capsule usually results in a dense bitemporal hemianopsia secondary to vascular damage. Nonetheless, in an elderly patient with a large cystic mass, a limited decompression with postoperative radiotherapy may well be possible from this exposure. Again, none of our 20 cases were approached in this fashion.

The majority of craniopharyngiomas in our adult patients were located in the region of the floor of the third ventricle, obliterating the suprasellar and interpeduncular cisterns, and intimately involving the branches of the posterior communicating and anterior choroidal arteries. The operative approach must allow adequate visualization of the optic apparatus and the region of the interpeduncular and suprasellar cisterns, and this is provided by a radical transtemporal approach with anterior temporal lobectomy. All 20 of our patients were explored in this fashion with the use of the surgical microscope.

The patient is positioned supine with a moderate (20%) foot-down tilt, and the head is slightly extended and rotated well to the left. A wing incision is then made with adequate posterior extension over the external auditory canal and frontal exposure reaching superiorly to just below the superior temporal line and anteriorly some 2 to 3 cm above the middle of the eyebrow (Fig. 4). A corresponding frontotemporal flap is cut across the sphenoid wing and extended across the anterior temporal fossa. The remaining anterior temporal fossa bone and the outer edges of the sphenoidal wing are removed with rongeurs. The dura is then opened with an asymmetrical Y incision across the Sylvian fissure. The anterior 2 cm of the temporal lobe is then removed, and the polar temporal veins are coagulated and divided to place the temporal region in continuity with the subfrontal plane. The double-arm Yasargil retractor is attached in the posterior temporal region, and the operative microscope is introduced. Under the microscope, the inner aspect of the Sylvian fissure is opened to define the carotid artery and optic nerve. This will allow dissection posterior to the carotid artery along the tentorial edge. The uncus can now be removed with suction coagulation to allow access to the interpeduncular fossa.

The posterior pole of the craniopharyngioma in the interpeduncular cistern is now apparent. Lateral to the tumor and invested in their own arachnoid lie the third nerve, and the posterior communicating artery and its thalamoperforating branches. Gentle dissection of the

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**Fig. 3.** Lateral carotid arteriogram showing downward bowing of the posterior communicating artery and stretching of the thalamoperforating vessels typical of a partially intrinsic craniopharyngioma.

**Fig. 4.** Incision line and burr hole placement for radical excision of craniopharyngioma by the transtemporal approach.
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Fig. 5. Artist's impression showing dissection of the posterior pole of a craniopharyngioma in the interpeduncular fossa (left) and dissection of the anterior subchiasmatic portion of a craniopharyngioma (right).

third nerve is now possible, and the nerve is covered with cottonoid to protect it from further trauma. (Although a transient partial third nerve palsy is a frequent postoperative finding, all instances have resolved completely.) The posterior cerebral artery and the basilar artery termination can now be defined, with their perforating branches also invested in the arachnoid of the interpeduncular fossa.

The posterior pole of the tumor is now visualized and available for mobilization and debulking of the tumor contents. If the tumor is small and has little posterior extension, it may be necessary to continue dissection in the triangular space between the carotid artery and the anterior choroidal artery anteriorly or between the posterior communicating artery and its anterior thalamoperforating branches inferiorly and posteriorly. Care must be taken not to divide either the posterior communicating artery, its thalamoperforating branches, or the anterior choroidal artery for fear of infarction in the basal ganglia. With careful bipolar coagulation, the small intrinsic branches supplying the tumor capsule can be separated from the branches of the posterior communicating artery and of the internal carotid artery itself.

The capsule of the tumor may now be incised, allowing access to the interior of the tumor, which will often be partially cystic. As one continues to decompress the interior of the mass, some recalcitrant portions of the mass may be mobilized with gentle controlled suction and monopolar coagulation. Blunt hooks and different dissectors, both sharp and blunt, may now be introduced progressively to mobilize the contents of the tumor.

Strong retraction on portions of the capsule adherent to the third ventricle or major vascular structures must be avoided. The entire posteriorinferior portion of the tumor capsule should now be free from the basilar artery and the posterior communicating and posterior cerebral arteries. The third ventricle is now entered anterior to the mamillary bodies.

Some blind dissection may be necessary to deliver the capsule from the third ventricle but, if adequately debulked, the capsule will come down with gentle traction and under direct microscopic vision. Even though the tumor is "invasive" in the walls of the third ventricle, its invasion is separated from the nuclear masses by a surrounding barrier of glial cells, which facilitates dissection. Histologically, this glial reaction has been noted for some time, but Sweet was the first to point out its significance in terms of surgical resection. The optic tract may be lightly adherent to the edge of the mass but can be retracted away under cottonoid, with the remaining portion of the temporal lobe. As long as no harm has come to any vascular structures supplying it, there will be no effect on vision from such gentle retraction of the optic tract itself.

With the roof of the capsule thus decompressed, vascular structures on the contralateral side will be visible. Again, the investing layer of arachnoid allows dissection of the capsule from the opposite internal carotid and posterior communicating arteries, and thalamoperforating branches. At this stage the tumor has been removed from all structures except the diaphragma sellae, the anterior aspect of the internal carotid artery, and the posterior chiasmatic region. The tumor may well be firmly adherent to the posterior aspect of the optic chiasm. Under microscopic vision it is possible, however, to free the tumor by sharp dissection from the posterior aspect of the chiasm. The entire ipsilateral optic nerve and tract are visualized, allowing accurate calculation of the line of the chiasm. Here sharp dissection minimizes dangerous retraction on the chiasm itself (Fig. 5 right).

This operative technique has allowed radical removal...
of the tumor in all 20 cases. In no case was it necessary to leave behind tumor or capsule that could be identified under the surgical microscope. Densely calcified tumor may be adherent to the medial aspect of the ipsilateral carotid artery, however. When it is not possible to create a plane between a calcified mass and the carotid wall, it is prudent to abandon the radical procedure, to amputate the remaining capsule, and to leave the calcified portion on the carotid wall in the hope that the more densely calcified material represents the least actively proliferating portion of the tumor.

**General Management**

Detailed endocrine assessment was not carried out in all of these cases preoperatively. Many patients presented with advancing visual failure, requiring prompt treatment which tended to preclude time spent in endocrine stress testing. A detailed postoperative endocrine assessment was invariably made in the Department of Endocrinology at St. Thomas's Hospital or elsewhere.

The poor results, and particularly the water and electrolyte disturbances associated with resection of these tumors have caused almost insuperable problems in the past. There is no doubt that the use of moderate-dose steroids (in this series dexamethasone, 4 mg/6 hrs during the radiological investigations and for the first 5 or 6 days postoperatively) has considerably eased management during the phase of postoperative edema in the hypothalamus and basal structures. Both hypothalamic hyperthermia were encountered after surgery. Four of our patients had considerably increased body temperature for 5 or 6 days postoperatively, treated by fanning and tepid sponging. One patient developed a persistently low body temperature, requiring fairly large doses of thyroid (thyroxine 0.4 mg/day) to restore a normal temperature. More common, however, were the disturbances of water and electrolyte balance. All these cases required the use of DDAVP (desamino-cis-1-8-D-arginine vasopressin) in the immediate postoperative period. In general, 0.1 mg DDAVP was given intravenously over the first 48 hours whenever the urinary output exceeded 1 liter in 4 hours. Thereafter, when the patient was sufficiently conscious to dispense with catheterization, the dose of DDAVP was chosen on the basis of plasma osmolality and electrolyte levels. Daily weighing of the patient was not found necessary in this series, and in most cases by 3 or 4 weeks postoperatively, DDAVP given once or twice a day as an endonasal spray was sufficient to control the urinary output. Less than half the cases still required DDAVP at the end of 2 years, suggesting that this unilateral transtemporal approach may well leave the supraoptic apparatus on the opposite side capable of return to normal function, avoiding the necessity for prolonged antidiuretic administration.

Gross weight gain was a problem in only two patients. In each instance it was associated with considerable increase in food intake which proved difficult to control. A discussion of the detailed management of these late endocrinological complications is not within the scope of this paper.

**Operative Results**

Of the 20 craniopharyngioma patients with radical excision, one died during the postoperative period (taken as the first 30 days after surgery). He died of pneumonia on Day 29, but had shown the fluctuating level of consciousness typical in cases of surgical damage to the hypothalamic region. Preoperatively, this patient had had raised intracranial pressure with drowsiness, papilledema, and urinary incontinence. Surgical mortality is thus 5%. There was one further death, not surgically related. That patient developed a Stevens-Johnson syndrome secondary to diphenylhydantoin which did not respond to aggressive medical management. The other 18 cases are analyzed for surgically related complications below.

Any patient requiring a second surgical procedure or developing a major neurological deficit (motor, sensory, or visual) was regarded as suffering "major morbidity." One patient (Case 5) developed a postoperative subdural hematoma requiring surgical drainage, but ultimately did well with no neurological sequelae. Two patients (Cases 6 and 8) developed secondary postoperative hydrocephalus and required ventriculoperitoneal shunts. Tragically, one of these (Case 6) developed shunt malfunction abroad and subsequently died from raised intracranial pressure; an autopsy was not performed. Another patient (Case 10) suffered hypothalamic damage with fluctuating level of consciousness which did not recover with time. This patient died of pneumonia 15 months postoperatively. Thus, of the 18 survivors, four (Cases 5, 6, 8, and 10) had major complications, an incidence of 22.2%. Combined with the one operative death (5%), this ultimately yields a combined major morbidity and mortality rate of 25% after a 3.1-year average follow-up period.

There were several minor operative complications. Four patients (Cases 6, 9, 14, and 18) developed partial ipsilateral third nerve palsy which resolved spontaneously, and a fifth (Case 11) had a complete third nerve palsy which resolved within 6 months. One patient (Case 3) developed temporal lobe seizures (5.5%), which responded well to an anticonvulsant regimen without further problem.

Only one patient received postoperative radiotherapy, in that instance because of doubts about the completeness of clearance of an intrasellar extension. All patients underwent routine postoperative CT scanning at regular intervals as well as neurological examinations at The National Hospital or by referring physicians if sent from a foreign country. This follow-up regimen has revealed one postoperative recurrence (Case 16). This patient developed a recurrent cystic collection at 8 months with increased intracranial pressure. The cyst was drained and a course of radiotherapy was then...
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Discussion

Nowhere in this paper has the word "cure" been used. It would indeed be presumptuous to use such terminology in a series with an average follow-up length of 3.1 years. Periods of 15 to 25 years are necessary before one might consider whether a "cure" has been achieved.\textsuperscript{9,10,26,27} Completeness of removal was evaluated by postoperative PEG (Fig. 6) or, more recently, by iohexol CT cisternography (Fig. 7) and plain and enhanced CT, using fourth-generation scanners. The existence of small tumor residua is best assessed by deformation of the anterior third ventricle on iohexol cisternography.

Maximum control of tumor recurrence by removal of all tumor accessible and visible to the surgical microscope must be balanced against the possibility of irreversible damage from aggressive surgical manipulation of the optic apparatus and anterior third ventricular structures. Newer diagnostic methods, however, have improved the accuracy of preoperative assessment of the extent of involvement of structures in the suprasellar region,\textsuperscript{19} and have allowed greater precision in planning the surgical attack. The surgical microscope facilitates a more precise determination of planes of dissection between tumor capsule and surrounding neural tissues; however, neither the microscope nor the CT scanner can predict the adherence of the capsule to vascular structures, nor can they detect microscopic tongues of tumor invading the glial tissue.\textsuperscript{26}

The data in 20 patients with radical tumor excision collected here by one surgeon over a 5-year period shows an operative mortality rate of 5.0%, a major morbidity rate of 22.2%, and a minor morbidity rate of 33.3%. One patient required surgical drainage of a recurrent cyst and a course of postoperative radiotherapy. None of the other patients were treated pre- or postoperatively with radiotherapy. All patients received postoperative CT scanning with neurological examination at 6- to 12-month intervals. No patients who were operated on elsewhere were included in this series. Previous reviews of the total case experience from The National Hospital included patients with reoperation,\textsuperscript{27} who form a radically different group with a higher mortality rate and increased morbidity.\textsuperscript{26} Recurrent tumors seem to be more tightly adherent to vascular and neural structures, making total excision impractical and more dangerous.

The results of these 20 cases must be compared with those of other surgical series involving radical excision of suprasellar craniopharyngiomas. In their pioneer work, Matson and Crigler\textsuperscript{16} reported no surgical mortality in 40 pediatric cases in which 25 tumors were considered to be radically resected. They reported late death in 12% of their patients, but a long-term follow-up review of that series, reported in 1975 by Katz,\textsuperscript{10} showed that the percentage of patients doing well at that time, without recurrence or neurological impairment, had fallen to 55%. The follow-up period for that series, however, is far longer than the 3.1-year average of the current report. Shapiro, et al.,\textsuperscript{23} again reporting on children, collected 22 cases with an average follow-up period of 2.4 years.\textsuperscript{23} They reported an 8% surgical mortality rate, and a success rate of 69%. In his classic report, Sweet\textsuperscript{25,26} described primary radical resection of craniopharyngiomas in 37 patients, including both adults and children. He reported an 8% surgical mortality rate, and a success rate of 69%. In a later follow-up review, he again reported an operative mortality rate

\textbf{Fig. 6.} Lateral pneumoencephalogram showing the completeness of excision (same case as Fig. 2). The restoration of the chiasmatic recess is the key observation.

\textbf{Fig. 7.} Iohexol cisternography scans with a GE 9800 scanner after radical excision of a craniopharyngioma. \textit{Upper:} Coronal cut showing a large opacified area of the temporal resection and the absence of deformity in the base of the third ventricle. \textit{Lower:} Sagittal scan in the same patient outlining the chiasm and the infundibular recess, and indicating the communication between the third ventricle and the interpeduncular cistern.
of 8% (all in the earlier years of surgical extirpation), with one late death. Thus, at 3 years postoperatively, he reported a success rate of 81%; Sweet’s data extend to 20 years, but an evaluation at 3 years most closely approximates the average length of follow-up of our cases. Our series included a 5% operative mortality rate, one late death, and a satisfactory outcome of 88% at 3.1 years of follow-up (Table 1).

The question of radiotherapy remains controversial. Some surgeons have advocated cyst drainage and diagnostic biopsy only, with postoperative radiotherapy. \(^{9,12,13}\) Between 1950 and 1969, Bloom and Harmer\(^4\) collected 99 cases from The National Hospital, Atkinson Morley’s Hospital, and The Hospital for Sick Children. Of these cases, 95 had “less than total resections.” Their regimen of “radical radiotherapy” consisted of a tumor dose of 5000 rads delivered to three fixed fields over a 6- to 7-week period. Follow-up evaluation was performed on 74 patients at 3 years, at which point 81% were alive and well. The majority of these cases were children (42% were under 15 years of age).

Another large series with radiotherapy was collected by Kramer\(^4\) from Philadelphia. Of his 43 patients, 25 were adults, followed from 1 to 20 years. He delivered 6500 rads to his adult patients over a period of 6½ weeks, which produced radiation death in 12%. He reported that 70% of his patients were alive following treatment. No mention is made of the quality of survival. Finally, a different form of radiotherapy has been advocated by Backlund.\(^1\) He injected colloidal beta-emitting yttrium-90 into the cystic portion of the tumor, using the Leksell stereotaxic frame. To this source he added radiation from cobalt-60 delivered via multiple ports to the solid portions of the tumor. Follow-up CT clearly demonstrated tumor shrinkage, with only two radiation-related deaths in the first 34 cases (5.5%).

Two independent theories have been advanced by Russell and Rubinstein\(^2\) regarding the origin of these tumors. Whether the pediatric group represents disorders of embryogenesis with cell rests of squamous epithelium left behind from the primitive gut endodermal elements and whether the adult tumor arises from metaplasia of cells in this region cannot be definitively settled. The possibility of such difference in pathogenesis must be considered when comparing reports of pediatric and adult cases, as Northfield\(^20\) stressed although for different reasons. The use of the surgical microscope with the radical subtemporal approach has allowed 88% of the patients to be doing well at follow-up review an average of 3.1 years postoperatively. The current surgical series seems to compare favorably with the majority of surgical reports and with radiotherapy. Whether or not this approach will result in a long-term “cure” will require prolonged evaluation, but it is clear that an initial surgical approach has the best chance of success. This initial surgery should be undertaken recognizing all the dangers attached to the initial procedure and remembering that these dangers are multiplied when such tumors are reexplored, even by the most experienced surgeons.

### Addendum

Since this paper was prepared, a further five cases have been operated on by the radical technique described, without mortality. Three cases have been uncomplicated. At no more than 3 months postoperatively, two patients have continuing but diminishing loss of recent memory.

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