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Microsurgery for Giant Craniopharyngiomas in Children

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The authors report 20 cases of giant craniopharyngioma in children operated on during the 6-year period from 1978 through 1984. The tumor size (maximal diameter) ranged from 5 to 11 cm. These patients presented with increased intracranial pressure, severe visual loss, and neurological and endocrinological deficits. Tumor growth had occurred in several directions, causing displacement of the circle of Willis, the optic apparatus, and the hypothalamus, as well as encroachment into the interpeduncular and prepontine cisterns and into the frontal, temporal, and posterior fossae. The surgical technique for total removal of these giant tumors in the last 10 cases is described. Emphasis is placed on the use of microsurgical technique during the first operation, close endocrinological follow-up, and early reevaluation by computed tomographic scanning. (*Neurosurgery* 17:585–595, 1985)

Key words: Brain neoplasm, Computed tomography, Craniopharyngioma, Craniotomy, Microsurgery, Suprasellar tumor, Visual defect

> We might add that there are rare giant forms, both pre- and retrochiasmatic, which result in puzzling clinical and neuroradiological patterns, which offer limited operative possibilities and which frequently have a grave prognosis.
>
> J. Rougerie (61), 1979

This article describes 20 cases of these rare, giant forms of craniopharyngioma alluded to in the preceding quotation. Craniopharyngiomas histologically are benign tumors. It is widely believed that they arise from squamous cell nests of the hypophyseal-pharyngeal duct, which normally involutes during embryonic life (18, 22, 53, 66, 74). Nests of these cells are found in the infundibulohypophyseal region, especially in adults (10) and less frequently in children (22). Craniopharyngiomas make up approximately 2.5% of all brain neoplasms and constitute about 9% of pediatric brain neoplasms and are the most common nonglial tumor in children (14, 32, 47, 66). There are many features of craniopharyngioma in children that are different fom those in adults, requiring their separation into age groups (3, 21, 28, 31, 33). We address the problem of treatment of giant craniopharyngiomas only in the pediatric age group.

In 1932, Harvey Cushing considered that craniopharyngiomas "offer the most baffling problem which confronts the neurosurgeon" (15). The high mortality rate for surgical treatment in the era before the availability of steroid pharmaceuticals created a gloomy outlook that has hung over the heads of these unfortunate patients (23, 32, 46, 52, 66). Authors have pointed to the high surgical mortality rates, failure of total removal, recurrence after presumed total removal, impairment of intellectual and psychosocial functions, and neurological and endocrine disabilities secondary to hypothalamic and brain stem injury. Hence, controversy in the management of this tumor has continued, with conservative surgical approaches having strong proponents (4, 33, 35, 44, 49, 52, 56, 61, 65). This attitude has been further strengthened by the reported effectiveness of radiation therapy (from both external and interstitial sources) in retarding the growth of these tumors (2, 5–7, 11, 12, 28, 38–40, 44, 52, 55, 57, 60, 68, 70, 73).

On the other hand, some surgeons, most notably Matson in the 1960s and Hoffman in the 1970s, have reported gross total removal of craniopharyngioma with apparent cure in as many as three-quarters of their cases (9, 30, 31, 36, 47, 48, 50, 67). These results generally were associated with a respectable operative mortality and morbidity. Serious problems were encountered with attempted total removal at a second operation, suggesting that the removal should be completed during the first operation by neurosurgeons experienced with this problem (24, 34, 47, 59, 67, 69, 73). The availability of the new generation of computed tomographic (CT) scanners has presented a means to confirm the extent of surgical removal, thus either providing confirmation that a total removal in fact has been accomplished or demonstrating that there is evidence of residual or recurring neoplasm (9, 41, 50, 51, 62, 69). Many surgeons have concluded that total removal of large and giant tumors exceeding 3 cm in diameter is difficult, hazardous, hopeless, or impossible (14, 16, 23, 24, 33, 36, 40, 61, 62, 70). Hoffman et al. did not share this view (30).

CASE MATERIAL

Clincial findings

Twenty consecutive patients harboring giant craniopharyngiomas (5–11 cm in maximal diameter) were treated surgically from 1978 to 1984. Ages ranged from 2 to 17 years in 19 patients, with 1 23-year-old man included because of a 10-year history of severely retarded growth and failure of puberty. There were 10 females and 10 males. The presenting complaints in all cases were related to the presence of an expanding suprasellar mass of long duration. Five patients were totally blind in both eyes, 2 were totally blind in one eye with decreased visual acuity in the other eye, and 8 patients showed a severe decrease in visual acuity in both eyes. The visual symptoms of 2 infants and 3 comatose patients could not be assessed adequately. Visual fields could be measured satisfactorily in 9 patients: 6 demonstrated bitemporal defects and 1 had only central visual fields. Examination of the fundi revealed papilledema in 7, Foster-Kennedy syndromes in 3, and optic atrophy in 7.

Additional findings included the following: 3 patients had reduced levels of consciousness, 6 had significant gait disturbances, 3 had left hemiplegia, 5 had convulsive seizures, 2 were dyphasic, 15 had evidence of delayed growth, and 6 had evidence of diabetes insipidus. Ten patients had complete hormonal studies before and after operation. There was no consistency in the hormonal test responses among patients, but over 60% manifested a deficit in one or more of the hormones, reflecting pituitary axis dysfunction. Only 1 patient demonstrated a mild elevation of prolactin preoperatively.

Radiographic findings

The plain skull x-ray films demonstrated erosion and enlargement of the sella turcica in 12 patients and evidence of calcification in the tumor in 16 patients. CT scanning was obtained in all patients, nicely demonstrating the mass in each case (Figs. 1 and 2). In 19 patients, the tumor had cystic components; only 1 was completely solid. The cyst fluid was hyperdense in 11 patients and hypodense in 5 patients. In 4 patients, the cyst fluid showed loculations of mixed (increased and decreased) densities. High density consistent with calcification was seen on the CT scans in 19 of the 20 cases.

CT scanning showed evidence of moderate to severe hydrocephalus in 19 patients; the dilatation was severe in 14. The anterior horns of the lateral ventricles often were asymmetrical, depending on the direction of the tumor growth. Contrast infusion was performed during 18 of the 20 CT scans. Of these 18 patients, 7 showed no enhancement (these were some

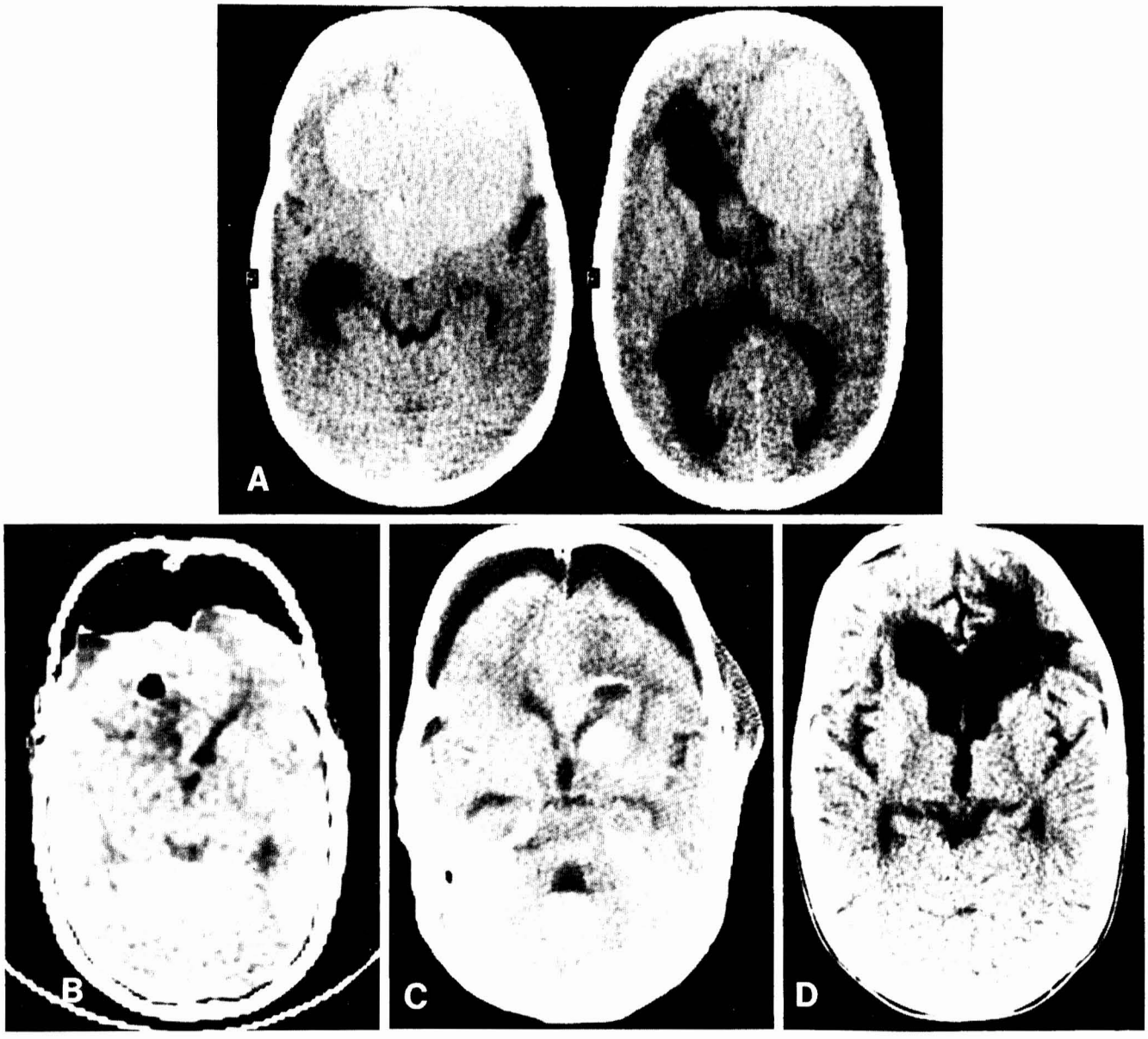

FIG. 1. Our largest craniopharyngioma (11-cm maximal diameter) imaged on CT scans. The right is on the observer's left. *A*: Preoperative unenhanced scan. Hyperdense tumor wraps around the anterior falx and has a locus of calcification (*left image*). At a higher cut, the tumor displaces the left frontal lobe and the 3rd ventricle (*right image*). Note hydrocephalus. *B*: Early postoperative CT scan shows air in the subdural space and right ventricle. *C*: Rapid recurrence of enhancing tumor 2 months later. Tumor recurrence is located in the left thalamus and the basal ganglia adjacent to the lateral ventricle. *D*: Scan after removal of the recurrent tumor and reconstruction of the frontal bones to obliterate the subdural space. Hydrocephalus and left frontal hypodensity are present.

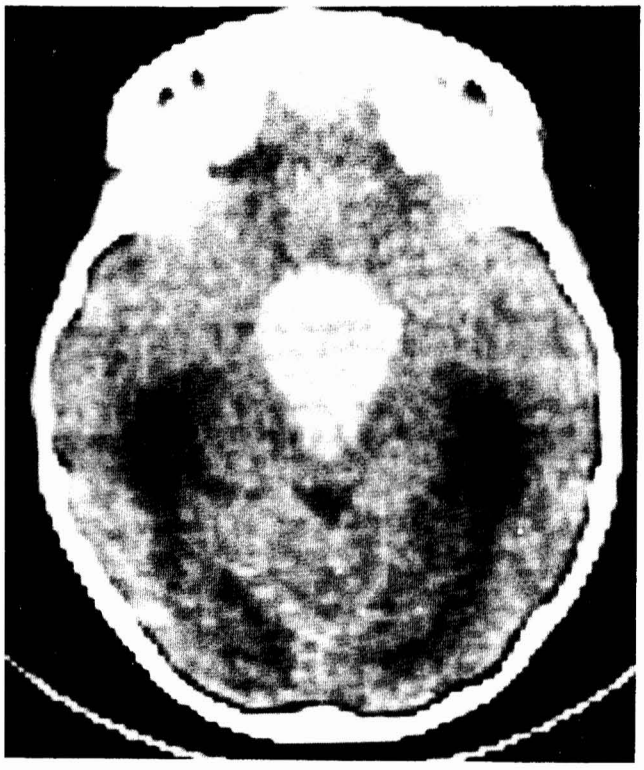

FIG. 2. Our smallest craniopharyngioma (5 cm in vertical diameter) imaged on an unenhanced CT scan. Note hydrocephalus, the hyperdense tumor, and two sites of calcification.

of the earliest cases in the series). In the 11 scans that showed tumor enhancement, 6 showed enhancement only in the tumor wall. The others showed some enhancement of the tumor wall and ill-defined enhancement of the tumor mass.

Cerebral angiography was performed on 19 patients. There was lateral displacement of one or both internal carotid arteries in 17 patients and elevation of the A-1 arteries in 15 patients. Twelve patients underwent vertebral angography; 11 of these showed displacement of the basilar artery posteriorly (Fig. 3). One-third of the cases demonstrated zones of narrowing of the intracranial segment of the internal carotid arteries or the anterior cerebral arteries; this suggested encirclement and compression of these arteries by tumor, which was proven at operation. In only 1 of the 19 patients was there evidence of a tumor blush. No neovascularity was identified.

SURGICAL TECHNIQUE

The earliest 10 patients (Group A) were operated on by four surgeons other than the authors. The technique described here is that used in the most recent 10 patients (Group B), operated on by one surgeon (O.A-M.). The surgical approach (i.e., either bilateral subfrontal or combined unilateral subfrontal and pterional) was selected after careful study of the neuroradiological images. The standard CT and angiographic studies were invaluable, and the coronal and sagittal reconstruction of the CT images provided significant information for proper orientation and planning.

A transsphenoidal approach was not utilized in any of our patients because of the large size and extent of the tumors. The tumors of 8 of our 10 Group B patients were approached bilaterally, and subfrontally. This approach and the microsurgical descriptions are detailed in our recent report on giant suprasellar meningiomas (1). In 2 patients, the right frontal and pterional approach was combined, based on the neuroradiological findings. In these 2 cases, the anterior cerebral arteries were not elevated, indicating the presence of prefixed chiasms (Fig. 4).

The Zeiss-Contraves operating microscope and various microinstruments were used to aid in the removal of tumor tissue. Microsurgical dissection was required to separate gently the adherent wall of the tumor from adjacent vital structures. The internal carotid arteries, posterior communicating arteries, anterior thalamic perforators, anterior choroidal arteries, and optic nerves were compressed and displaced laterally by the tumor. After aspiration of the cyst fluid, much of the cystic component of the tumor collapsed, permitting a safer dissection of tumor wall from these structures. We avoid spilling cystic contents into the spinal fluid areas to prevent chemical meningitis.

Further dissection of the tumor progressed to the region of the bifurcation of the internal carotid artery and continued along the middle cerebral artery and into the sylvian fissure. The anterior choroidal artery and nearby perforators were carefully preserved. Further dissection was carried along the anterior cerebral arteries and their perforators (Fig. 5). The critical blood supply to the hypothalamus, optic chiasm, and nerves was preserved (Fig. 6). The tumor was dissected free from the anterior communicating region and hypothalamus. No adverse traction was applied to the tumor to avoid injury to the compressed hypothalamus. The tumor wall then was dissected away from the infundibulum and pituitary stalk. In three of our patients, we were able to preserve the pituitary stalk. However, in the other patients, the tumor originated within the course of the pituitary stalk (Fig. 7), which was then sectioned as low as possible to avoid permanent diabetes insipidus and further injury to the hypothalamus. In one of our patients, the craniopharyngioma extended into the cavernous sinus, from which the tumor was extracted successfully (Fig. 8). An area that required particular attention was the region of the dorsum sellae. Here the tumor capsule and the stretched dura mater of the diaphragm adhered to each other (unlike our large suprasellar meningiomas (1)), and a careful dissection was required to obtain a cleavage plane between them. The tumor was followed into the interpeduncular and prepontine cisterns. It then was removed from the displaced basilar artery and its branches. There usually was a good plane for dissection here because of the intact and intervening arachnoid membrane of Liliequist (membrane of Key and Retzius) (Fig. 9). In two patients, the tumor extended into the cerebellopontine cistern, but it was not adherent to any nerve or vascular structure in this space.

In one patient with a prefixed optic chiasm (Fig. 4), the tumor was approached unilaterally between the drilled-down sphenoid wing and the opened sylvian fissure and downward between the internal carotid artery and the optic nerve. Part of this same tumor had to be removed through the lamina terminalis (Fig. 10).

On the basis of the radiological studies, a craniopharyngioma appearing to lie inside the 3rd ventricle may actually be displacing the floor of the 3rd ventricle upward and lying outside the 3rd ventricle proper. When the image of a giant craniopharyngioma appeared to lie within the 3rd ventricle (one case) (Fig. 2), we utilized a combined unilateral subfrontal and pterional approach. The base of the brain was inspected first to confirm that the tumor was indeed inside the 3rd ventricle rather than displacing the floor of the 3rd ventricle upward. Were the tumor to lie outside the 3rd ventricle, the subfrontal and transsylvian routes would be available to the surgeon. But, with the tumor inside the 3rd

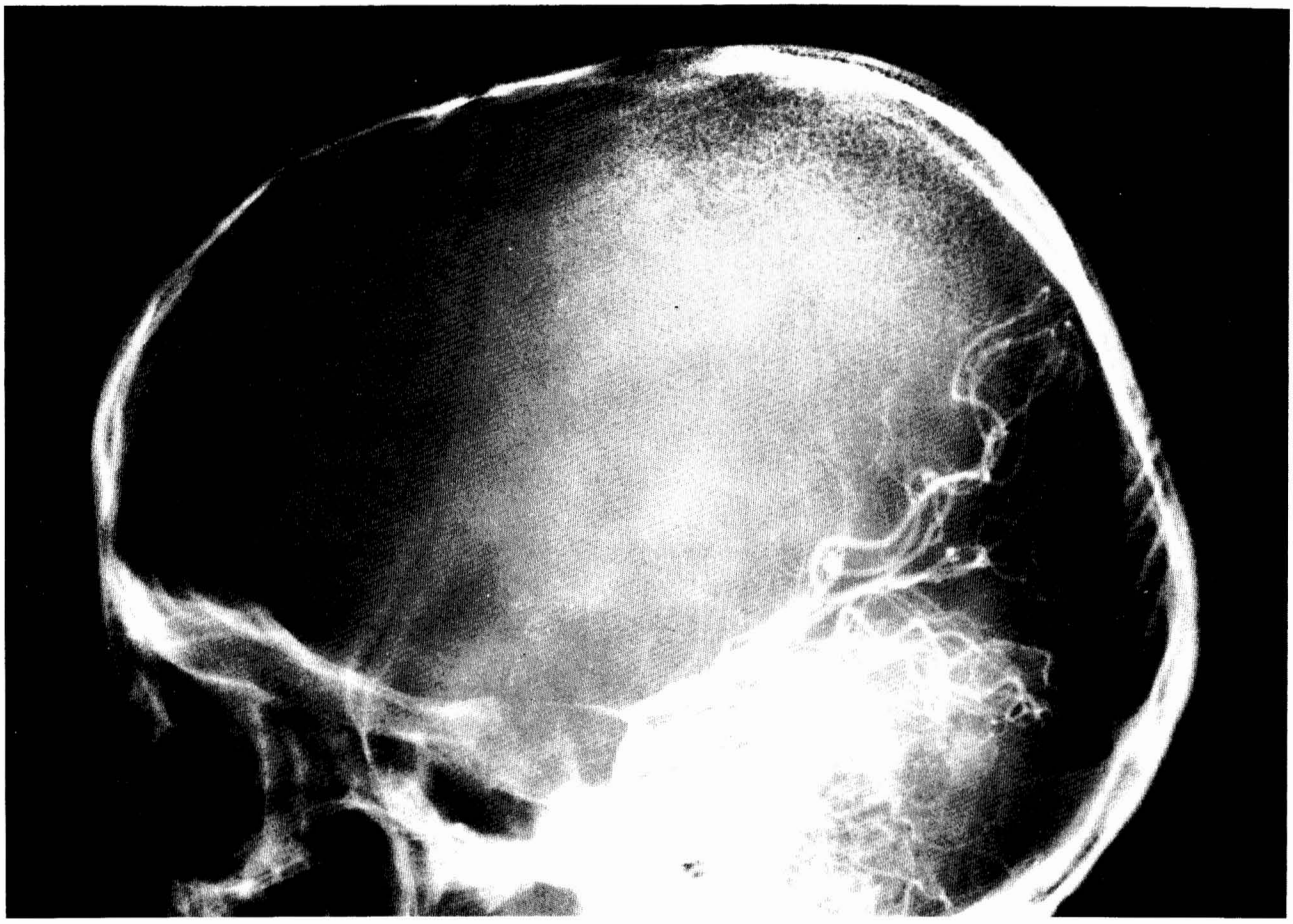

FIG. 3. Typical findings on vertebrobasilar angiography: slight suture separation, faint density (extensive calcification) in the suprasellar region, displacement of the termination of the basilar artery backward from the eroded dorsum sellae, stretching and posterior displacement of the posterior thalamic perforators and the posterior choroidal arteries, and stretching of a thinned-out posterior communicating artery.

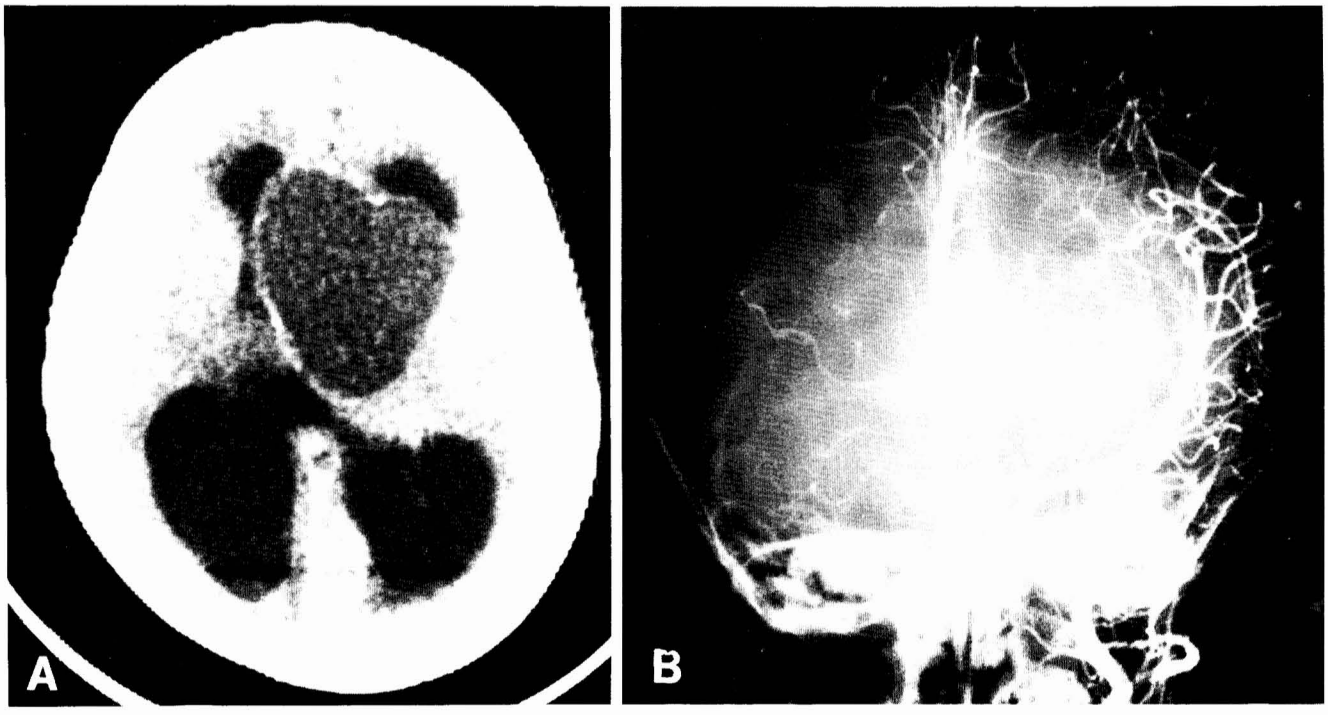

FIG. 4. Case of craniopharyngioma in the presence of a prefixed chiasm. *A*: Enhanced CT scan indicating hydrocephalus and cystic tumor with enhancing walls. Tumor pushed the floor of the 3rd ventricle upward from its retrochiasmatic position. *B*: Left carotid angiogram of the same patient. The A-1 arteries lie above a relatively immobile prefixed chiasm and so are not elevated by this large tumor.

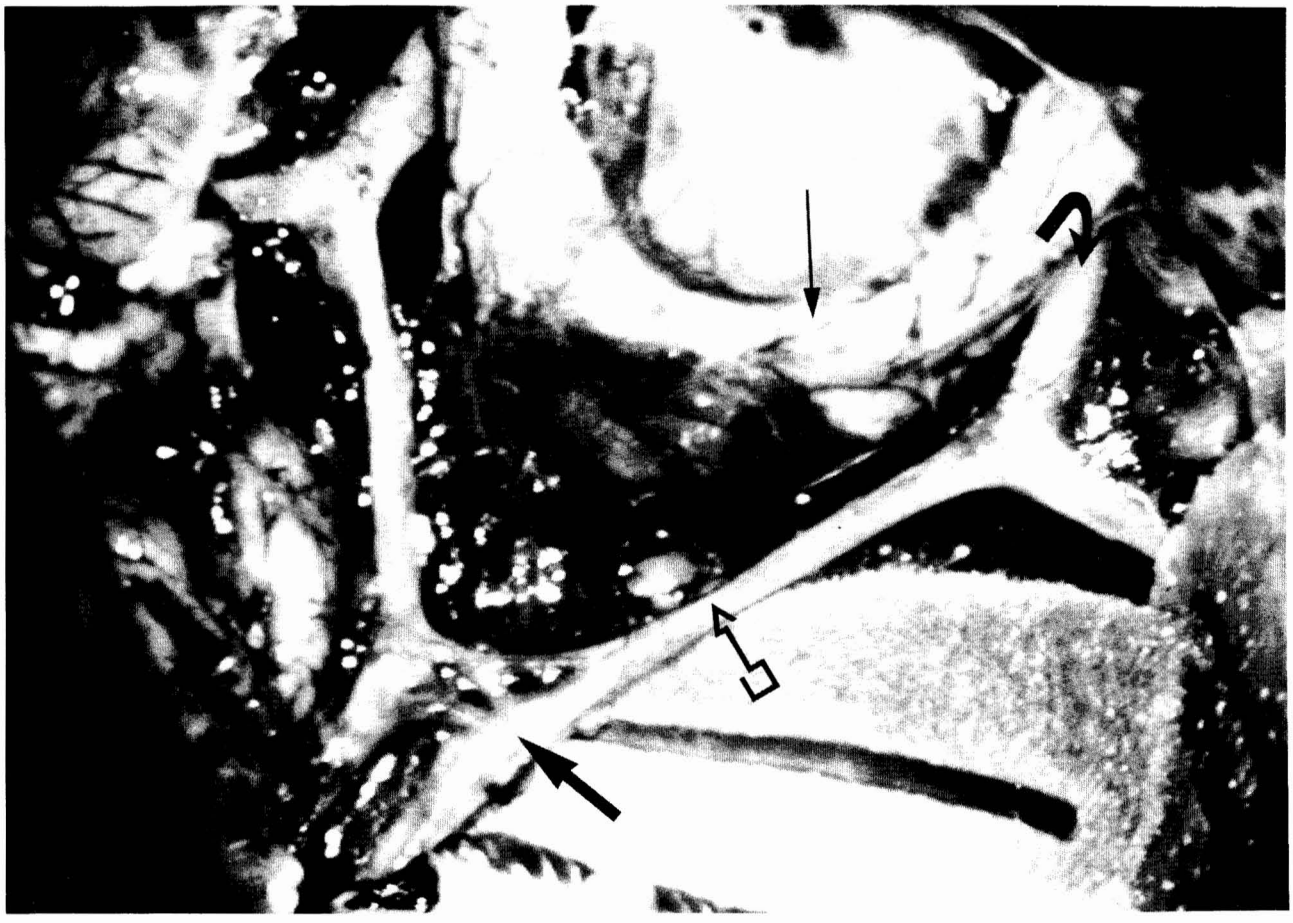

FIG. 5. Surgeon's view through the operating microscope after excision of the anterior half of the tumor. Patient's right is on the observer's right; patient's anterior direction is at *top*. *Small arrow* crosses the remainder of the tumor and points to the optic chiasm. *Curved arrow* crosses the right optic nerve below the anterior clinoid process and points to the carotid artery. *Open arrow* crosses cottonoid and points to the right A-1 artery. Both stretched A-1 arteries flank the lamina terminalis, which is covered with a film of blood. *Large arrow* indicates the anterior communicating complex displaced posterosuperiorly.

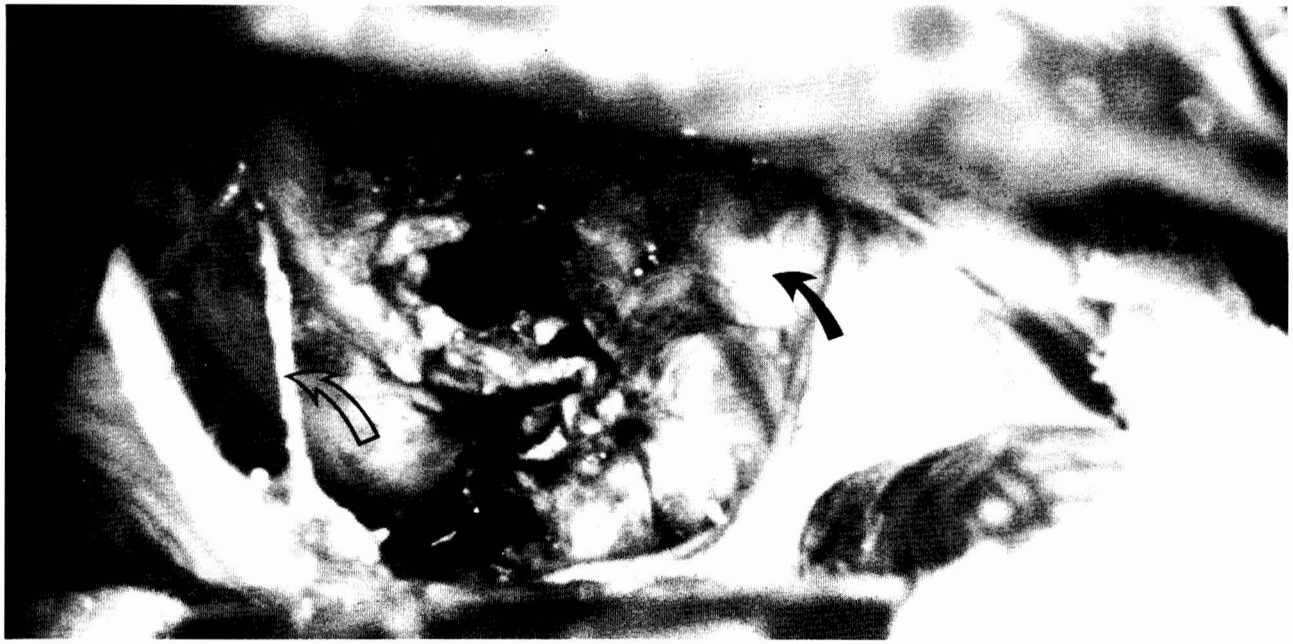

FIG. 6. Surgeon's view of region between the tuberculum sellae (*above*) and the optic chiasm hidden by the frontal lobe retractor (*below*). *Closed arrow* crosses the laterally displaced right optic nerve and points to the carotid artery below the more superficial anterior clinoid process (out of focus). *Open arrow* crosses the partially decompressed tumor capsule and points to one of two small arteries from the left carotid artery to the left optic nerve and chiasm.

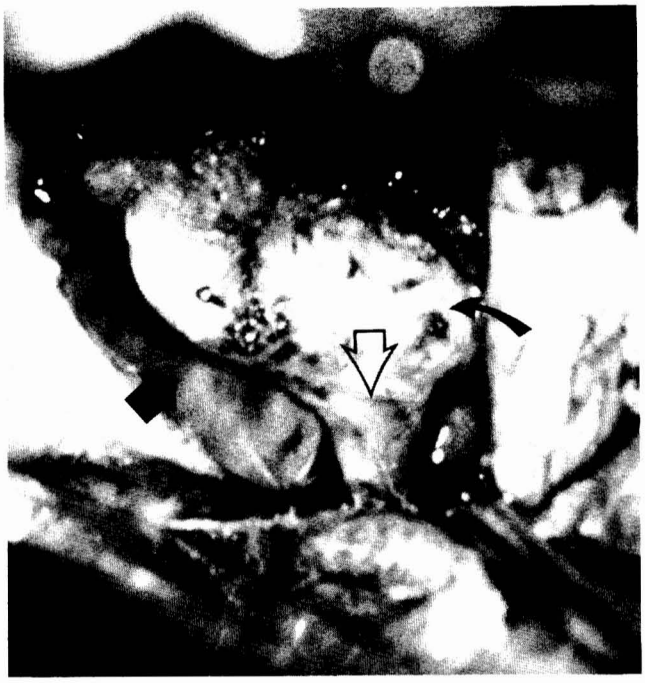

FIG. 7. Surgeon's view of the region inferior to the hypothalamus. Out of focus are the tuberculum sellae (*above*) and the frontal lobe retractor and probe (*below*). The bulk of the tumor has been removed. *Curved arrow* crosses the decompressed right optic nerve and points to deeper tumor tissue. *Straight solid arrow* crosses the left optic nerve and points to the deeper arachnoid membrane of Liliequist behind remaining tumor. *Open arrow* crosses the remaining origin of the craniopharyngioma and points to its junction with the pituitary stalk where the stalk was subsequently sectioned.

ventricle, a standard transventricular approach (either transcallosal or transcortical) was carried out through the dilated ventricle of the frontal horn and foramen of Monro (Fig. 11).

Postoperatively, close attention was paid to fluid and electrolyte balance, as well as hormonal therapy with the assistance of a pediatric endocrinologist. Postoperative CT scanning was done early to determine whether there were any tumor remnants. If such remnants are identified, early reoperation is advised before significant adhesions develop. Follow-up scans also are needed to detect recurrences from retained nests of tumor cells (Fig. 1*C*). On occasion, it may be advisable surgically to reconstruct the frontal fossa if excess space exists after tumor removal.

RESULTS

In the early group (Group A), tumors of 3 of the 10 patients were totally removed; 2 of these patients are alive and well without recurrence on follow-up CT scanning 3 and 6 years later, respectively. There were 5 postoperative deaths (one after a total tumor removal). One patient was lost to follow-up, 1 remained hemiplegic, and 1 remained in a vegetative state.

In the recent group (Group B), all 10 patients recovered from operation without any increase in their preperative neurological deficit. None of the patients had worsening of their visual fields, and 3 had improvement in both visual acuity and fields. There were two delayed deaths (during hospitalization); one from a cerebrospinal fluid shunt infection and septicemia and the other from gram-negative septicemia. All patients had postoperative CT scans (Fig. 12). In 9 of these 10 patients, the tumor was totally removed as seen through the operating microscope, but 1 of these 9 demonstrated rapid (in 2 months) tumor regrowth under the left lateral ventricle (Fig. 1). This additional growth was removed completely at a second operation. The 10th patient had a small piece of tumor adherent to the compressed hypothalamus, and no dissection plane could be established safely at operation. Both of these latter 2 patients received postoperative radiation therapy. To date, there has been no evidence of additional tumor recurrence in any of the Group B survivors over a 4-month to 3-year follow-up period. None of the survivors had mental deficits or behavioral problems; psychometric testing was not performed.

In our Group B, all patients had transient diabetes insipidus during the immediate postoperative period. This was managed by close control of the fluid and electrolyte balance and by the intranasal administration of DDAVP (desmopressin acetate; Armour Pharmaceutical Co., Kankakee, Illinois) to

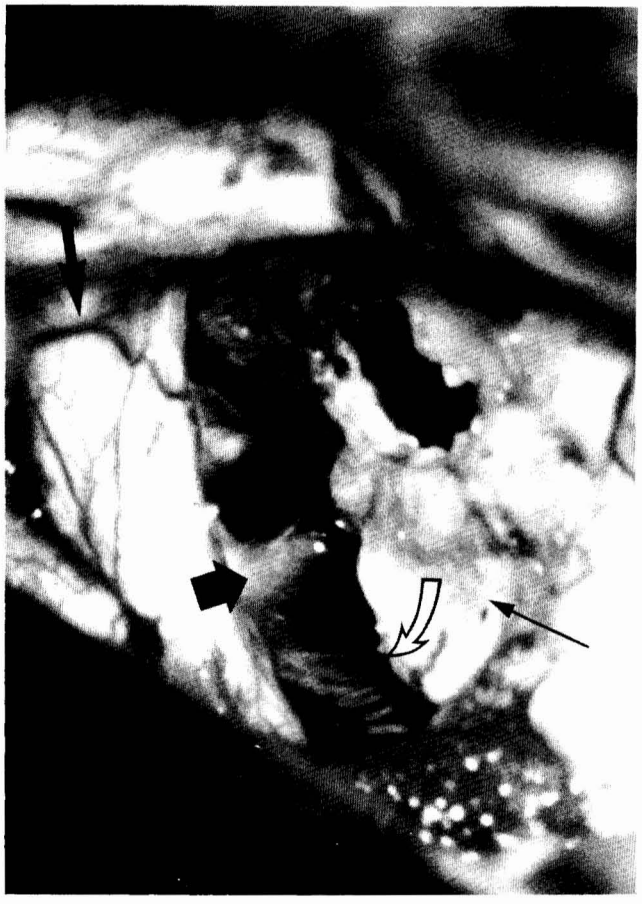

FIG. 8. Surgeon's view after the remains of the tumor (*thin arrow*) had been dissected out of the left cavernous sinus. The view is through the microscope, tilted and looking leftward under the left optic nerve. *Long thick arrow* crosses the left anterior clinoid process and points to the left optic nerve. *Short thick arrow* crosses the left optic nerve and points to the deeper cavernous sinus. *Open arrow* crosses the tumor capsule and points to the deeper cavernous sinus. At operation, this appeared as a blue plexus of venous walls, which had to be carefully separated from the craniopharyngioma. *Thin arrow* crosses the cottonoid and the right optic nerve and points to the decompressed remaining tumor capsule. Below is gelatin sponge and the frontal lobe retractor.

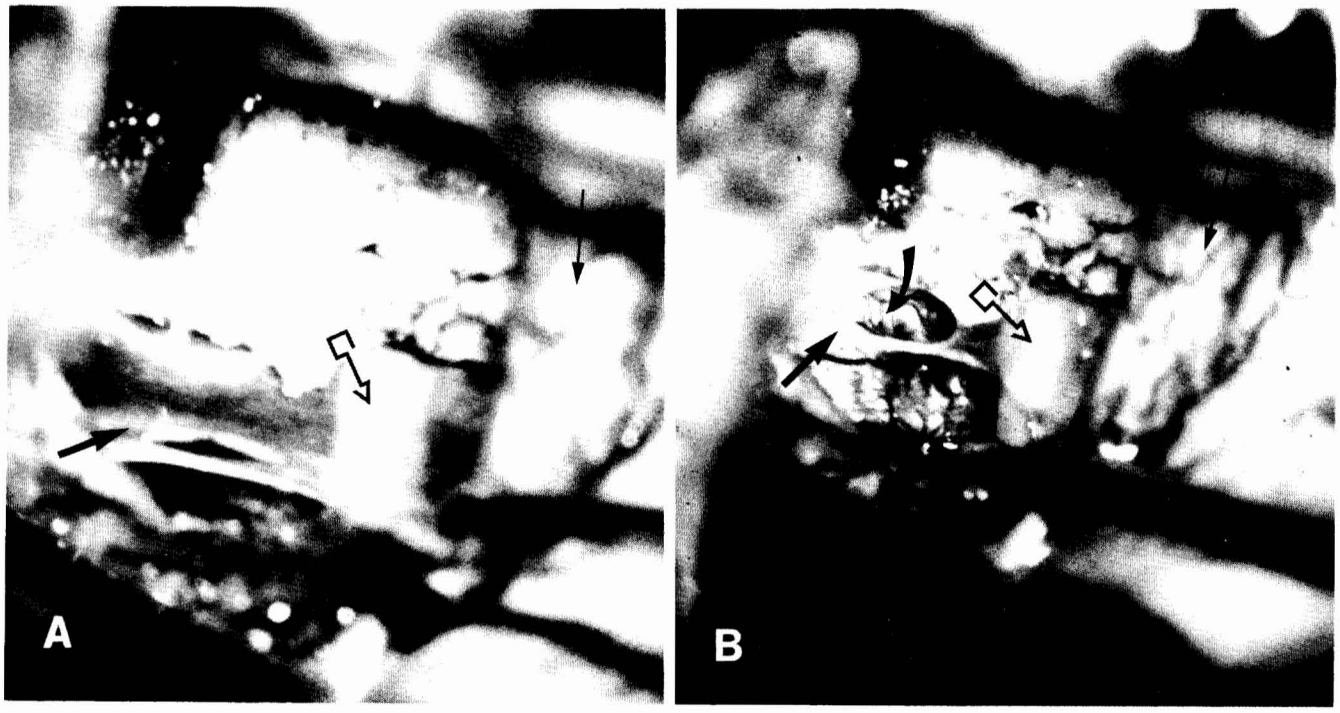

FIG. 9. Surgeon's view of tumor dissection in front of the arachnoid membrane of Liliequist (membrane of Key and Retzius). *Large straight arrow* crosses the edge of the left optic nerve (out of focus) and points to the deeper membrane of Liliequist. *Open arrow* crosses the tumor and points to the deeper basilar artery behind the arachnoid membrane of Liliequist. *Thin arrow* crosses the right anterior clinoid process and points to the right optic nerve (not in focus). *B, curved arrow* crosses the tumor and points to the deeper pons. *Below* are the frontal lobe retractor and the bipolar forceps (not in focus). Photographs were taken before (*A*) and after (*B*) interruption of the membrane of Liliequist.

FIG. 10. Surgeon's view of the opened lamina terminalis (same patient as Fig. 4). *Short arrow* crosses the right anterior clinoid process and points to the deeper carotid artery. *Open arrow* crosses the right optic tract and points to the opened lamina terminalis, deep to which is dissected tumor tissue. *Solid curved arrow* crosses the posterior origin of the left optic nerve and points to the prefixed optic chiasm. *Lower left*, a suction tube goes inside the tumor cavity. *Below* is the frontal lobe retractor.

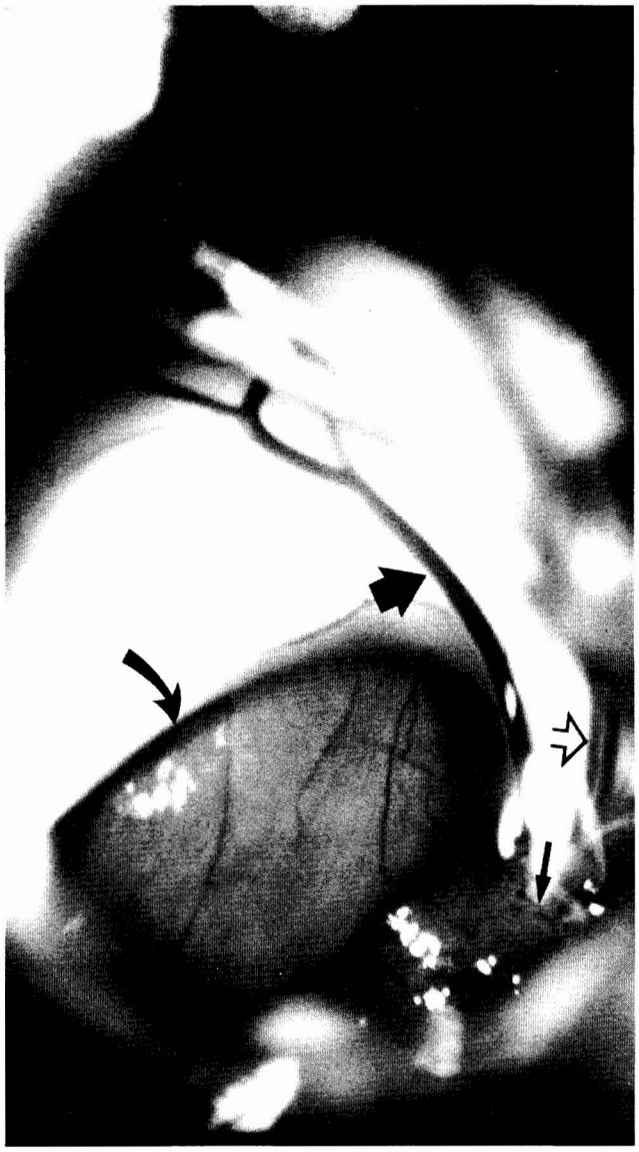

FIG. 11. Surgeon's view of the right foramen of Monro as seen through a transcortical incision and the hydrocephalic lateral ventricle. Medial direction is to observer's left. *Curved arrow* crosses the right fornix and points to the edge of the foramen of Monro and to the outer wall of the craniopharyngioma inside the 3rd ventricle. *Heavy straight arrow* crosses the fornix and points to the septal vein. *Open arrow* crosses the fornix and points to the anterior caudate vein. *Small straight arrow* indicates choroid plexus at the posterior rim of the foramen of Monro. (Same patient as Fig. 2.)

control the diabetes insipidus. Among the surviving eight patients, four required hormonal replacement (two on DDAVP, four on corticosteroids, and two on thyroid replacement). All patients resumed normal growth patterns without the need for the administration of growth hormone. The growth of the 23-year-old patient with delayed puberty and retarded growth was accelerated after operation. Prolactin levels remained normal in all patients. Four patients required cerebrospinal fluid shunting for hydrocephalus postoperatively.

DISCUSSION

The results in these cases must be viewed in terms of their circumstances. Most of the patients traveled long distances, and their medical problems often were far advanced before they reached our hospital. In addition to the neurological and visual deficits, many of the patients had other medical and nutritional problems. The neurological findings in our patients, although qualitatively similar to those of other centers (3, 23, 28, 30, 32, 33, 66, 72), were quite advanced because of the great size of these lesions. In our patients, the sex distribution was equal, as noted in some other reports (4, 29); other communications have shown a male predominance (66, 68).

Modern imaging procedures, such as CT scanning and magnetic resonance imaging, are now indispensable in the work-up of these patients. The craniopharyngioma is identified on CT scanning based on location, cystic components, calcifications, and enhancement after intravenous contrast injection. Not only are these studies important for the identification of these tumors, but extensions and encroachments of the tumor are visualized, which assists in the planning of the surgical approach. Our CT examinations detected calcification in 95% of our cases, a detection rate higher than that in other reported series (8, 19, 54). A unique feature in our scans was the high incidence of hyperdense cystic fluid and the frequency of multiple cysts. These findings most likely were due to the long duration of tumor growth, with degeneration and calcification. The long histories also accounted for the higher incidence of hydrocephalus in our series. The incidence of tumor enhancement on our CT studies was similar to (8) or higher than (54) that reported elsewhere. Modern dynamic CT scanning techniques very likely will demonstrate enhancement in the walls of nearly all of these tumors. Because of the size of these tumors, we have not used contrast-enhanced cisternography with CT scanning; it may be of value in cases of smaller craniopharyngiomas (50).

Cerebral angiography also is necessary for planning the surgical approach and showing the relationship of the tumor to the vasculature. As shown in Figure 4, the absence of elevated A-1 arteries suggests a prefixed chiasm with retrochiasmatic growth of the tumor (50), which requires a combined unilateral subfrontal and pterional approach rather than a bilateral subfrontal approach to the tumor. We agree that vertebral angiography is helpful in identifying the posterior extension of the tumor (33, 37, 50). Venous digital subtraction angiography may provide sufficient anatomical information in these children.

The various techniques, indications, limitations, and complications of each approach for removal of craniopharyngiomas are well described in the litreature (25–27, 33, 43, 45, 47, 50, 61, 63–65, 67, 71). The need to select the approach most suitable for individual cases is emphasized (13, 50, 61, 65, 67). The tremendous stretching of the optic nerves and internal carotid arteries makes the anterior extension of the tumor difficult to remove. This phenomenon, however, does provide better access to the interpeduncular cistern and posterior fossa after the anterior extension of the tumor is excised by dissecting it away from the anterior circle of Willis and hypothalamus. To date, we have not had to remove any sphenoid bone at the tuberculum sellae to gain access to the retrochiasmatic tumors, as described by Patterson and Danylevich (58).

In cases of large retrochiasmatic tumors with extension into the 3rd ventricle, the surgeon should be prepared to take other

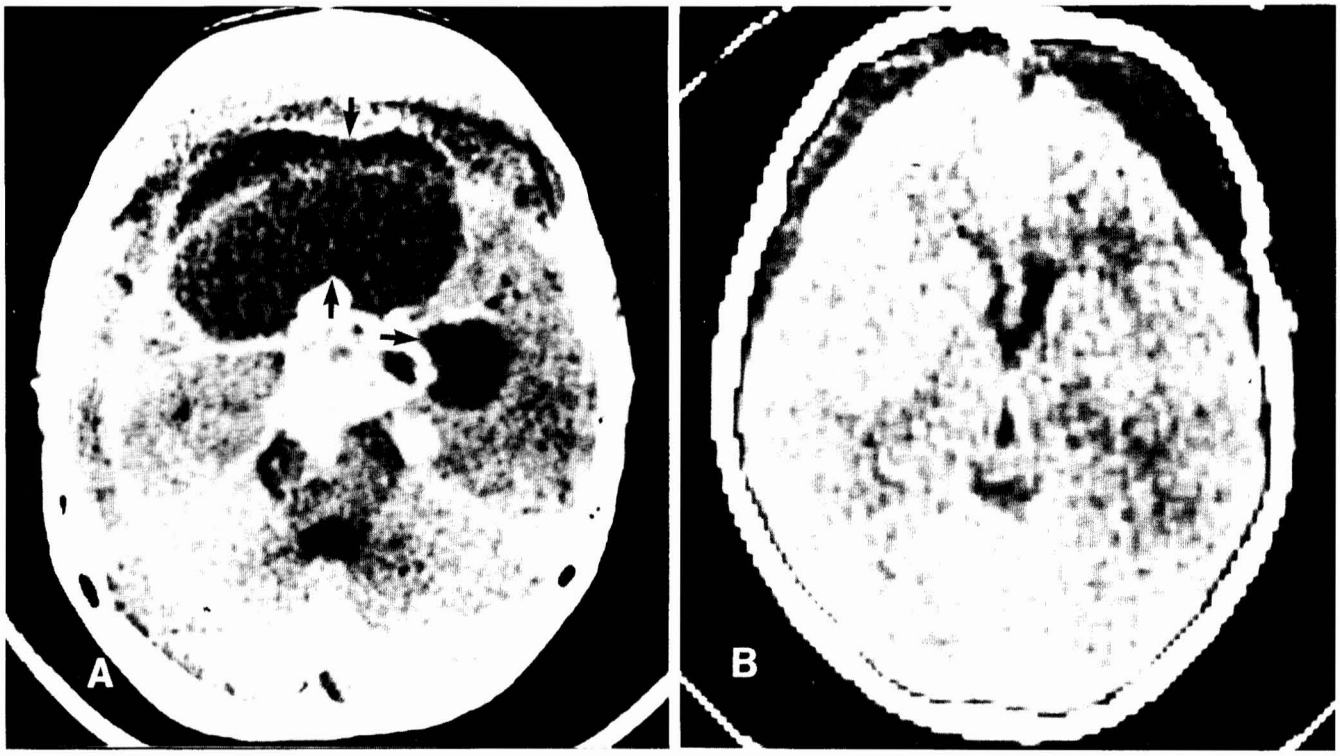

FIG. 12. Example of preoperative (*A*) and postoperative (*B*) scans illustrating gross total removal of a giant craniopharyngioma. *A*: Enhancing tumor with large multilocular cystic components (*arrows*). The hyperdense outer ring is an artifact. *B*: Postoperative CT scan illustrates collapse of the brain, leaving bifrontal subdural fluid collections to take up the space.

avenues, including a more lateral approach, a transventricular approach, or an approach through the lamina terminalis. In these situations, it may be advisable to plan a frontal-pterional bone flap rather than to do two or more operations through separate openings. It is important to identify those tumors that are actually inside the 3rd ventricle because some only appear to be so, but actually push the floor of the 3rd ventricle upward. Attempts to remove tumors in the latter situation by an approach through the foramen of Monro may result in devastating damage to the floor of the hypothalamus (17, 61, 71).

Microsurgical technique is indispensable to preserve the perforating blood vessels and the blood supply to the optic apparatus and hypothalamus (20, 30, 67). A gliotic layer usually exists between the functioning wall of the hypothalamus and the tumor (42, 67, 71), thus providing a plane for microsurgical dissection. When possible, the pituitary stalk should be saved if the tumor can be dissected free from that structure (36, 61, 67). The fact that only two of our Group B survivors required permanent DDAVP administration postoperatively attests to this.

The relatively high recurrence rate of craniopharyngioma after presumed total removal in some series may be due to residual tumor that was not detected by intraoperative inspection. Present may microsurgical techniques are improving the incidence of successful total removal. CT scanning is valuable in assessing evidence of residual tumor after operation (9, 41, 50, 51, 62, 69). Caution must be exercised to differentiate postoperative residual tumor from postoperative changes such as hematoma or contusion, which resolve over a period of time (51). Despite total removal, four of our patients required shunting for persistent hydrocephalus postoperatively. This raises the question of whether the hydrocephalus is due entirely to ventricular outlet obstruction or is also related to other factors, such as inadequate absorption pathways in the subarachnoid space or the persistence or development of adhesions secondary to the operation.

Shapiro et al. pointed out that success with radical excision of such tumors is inversely proportional to the size of the tumor, with only 20% of mixed (i.e., solid and cystic) tumors over 3 cm in diameter being radically excised (62). Other surgeons also suggested that large size and adverse location often prevented radical surgical excision (14, 23, 24, 33, 40, 61, 70). Guidetti and Fraioli in 1979 stated, "The postoperative mortality and morbidity rates were high in those tumours classified as giant, especially when total excision was performed" (24). Djordjević et al. in the same year noted, "Radical surgery cannot be performed on sellar-suprasellar giant craniopharyngiomas with fibrous capsules and small cysts with a hollow sella and retrodorsal extension, with or without the use of a microscope" (16). Konovalov in 1983 concluded, "Very serious difficulties can arise in the case of huge giant tumours growing in various directions" (36). Our patients belong to the category suggested by these descriptions. Only long term follow-up will confirm whether our patients are indeed cured. However, we now suggest that, with modern neuroradiology, experience in microsurgical technique, and proper endocrinological support, the majority of these neoplasms can be removed safely. This is best done during the first operation on the craniopharyngioma.

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REFERENCES

1. Al-Mefty O, Holoubi A, Rifai A, Fox JL: Microsurgical removal of suprasellar meningiomas. Neurosurgery 16:364–372, 1985.
2. Backlund E-O: Studies on craniopharyngiomas: III. Stereotaxic treatment with intracystic yttrium-90. Acta Chir Scand 139:237–247, 1973.
3. Banna M, Hoare RD, Stanley P, Till K: Craniopharyngioma in children. J Pediatr 83:781–785, 1973.
4. Bartlett JR: Craniopharyngiomas—a summary of 85 cases. J Neurol Neurosurg Psychiatry 34:37–41, 1971.
5. Bloom HJG: Combined modality therapy for intracranial tumors. Cancer 35:111–120, 1975.
6. Bloom HJG, Harmer CL: Craniopharyngiomas. Br Med J 2:288–289, 1972 (letter).
7. Cabezudo JM, Vaquero J, Areitio E, Martinez R, Garcia-de-Sola R, Bravo G: Craniopharyngiomas: A critical approach to treatment. J Neurosurg 55:371–375, 1981.
8. Cabezudo JM, Vaquero J, Garcia-de-Sola R, Leunda G, Nombela L, Bravo G: Computed tomography with craniopharyngiomas: A review. Surg Neurol 15:422–427, 1981.
9. Carmel PW, Antunes JL, Chang CH: Craniopharyngiomas in children. Neurosurgery 11:382–389, 1982.
10. Carmichael HT: Squamous epithelial nests in the hypophysis cerebri. Arch Neurol Psychiatry 26:966–975, 1931.
11. Cavazzuti V, Fischer EG, Welch K, Belli JA, Winston KR: Neurological and psychophysiological sequelae following different treatments of craniopharyngioma in children. J Neurosurg 59:409–417, 1983.
12. Chin HW, Maruyama Y, Young B: The role of radiation treatment in craniopharyngioma. Strahlentherapie 159:741–744, 1983.
13. Choux M, Lena G: Bases of surgical management of craniopharyngioma in children. Acta Neurochir (Wien) [suppl]28:348, 1979.
14. Cobb CA, Youmans JR: Brain tumors of disordered embryogenesis in adults. in Youmans JR (ed): *Neurological Surgery*. Philadelphia, WB Saunders Co. 1982, pp 2899–2935.
15. Cushing HW: *Intracranial Tumors: Notes upon a Series of Two Thousand Verified Cases with Surgical-mortality Percentages Pertaining Thereto*. Springfield IL, Charles C Thomas, 1932, p 97.
16. Djordjević M, Djordjević Z, Janićijević M, Nestorović B, Stefanović B, Ivkov M: Surgical treatment of craniopharyngiomas in children. Acta Neurochir (Wien) [suppl]28:344–347, 1979.
17. Ehni G: Craniopharyngioma surgery in children. J Neurosurg 52:439, 1980 (letter).
18. Erdheim J: Über Hypophysenganggeschwülste und Hirncholesteatome. SB Acad Wiss Wien 113:537–726, 1904.
19. Fitz CR, Wortzman G, Harwood-Nash DC, Holgate RC, Barry JF, Boldt DW: Computed tomography in craniopharyngiomas. Radiology 127:687–691, 1978.
20. Fox JL: *Intracranial Aneurysms*. New York, Springer-Verlag, 1983, vol II, pp 877–909.
21. Garcia-Uria J: Surgical experience with craniopharyngioma in adults. Surg Neurol 9:11–14, 1978.
22. Goldberg GM, Eshbaugh DE: Squamous cell nests of the pituitary gland as related to the origin of craniopharyngiomas: A study of their presence in the newborn and infants up to age four. Arch Pathol Lab Med 70:293–299, 1960.
23. Gordy PD, Peet MM, Kahn EA: The surgery of the craniopharyngiomas. J Neurosurg 6:503–517, 1949.
24. Guidetti B, Fraioli B: Craniopharyngiomas: Results of surgical treatment. Acta Neurochir (Wien) [suppl]28:349–351, 1979.
25. Hamberger C-A, Hammer G, Norlén G, Sjögren B: Surgical treatment of craniopharyngioma: Radical removal by the transantrosphenoidal approach. Acta Otolaryngol (Stockh) 52:285–292, 1960.
26. Hamer J: Removal of craniopharyngioma by subnasal-transsphenoidal operation. Neuropädiatrie 9:312–319, 1978.
27. Hardy J, Wigser SM: Trans-sphenoidal surgery of pituitary fossa tumors with televised radiofluoroscopic control. J Neurosurg 23:612–619, 1965.
28. Hoff JT, Patterson RH Jr: Craniopharyngiomas in children and adults. J Neurosurg 36:299–302, 1972.
29. Hoffman HJ: Supratentorial brain tumors in children. in Youmans JR (ed): *Neurological Surgery*. Philadelphia, WB Saunders, 1982, pp 2702–2732.
30. Hoffman HJ, Hendrick EB, Humphreys RP, Buncic JR, Armstrong DL, Jenkin RDT: Management of craniopharyngioma in children. J Neurosurg 47:218–227, 1977.
31. Humphreys RP, Hoffman HJ, Hendrick EB: A long-term postoperative follow-up in craniopharyngioma. Childs Brain 5:530–539, 1979.
32. Ingraham FD, Scott HW Jr: Craniopharyngiomas in children. J Pediatr 29:95–116, 1946.
33. Kahn EA, Gosch HH, Seeger JF, Hicks SP: Forty-five years of experience with the craniopharyngiomas. Surg Neurol 1:5–12, 1973.
34. Katz EL: Late results of radical excision of craniopharyngiomas in children. J Neurosurg 42:86–90, 1975.
35. Kempe LG: *Operative Neurosurgery*. Berlin, Springer-Verlag, 1968, vol 1, pp 90–93.
36. Konovalov AN: Microsurgery of tumours of diencephalic region. Neurosurg Rev 6:37–41, 1983.
37. Kramer RA, Poole GJ, Moody DM, Newton TH: Angiography in craniopharyngiomas. Radiology 109:99–103, 1973.
38. Kramer S: The value of radiation therapy for pituitary and parapituitary tumours. Can Med Assoc J 99:1120–1127, 1968.
39. Kramer S, McKissock W, Concannon JP: Craniopharyngiomas: Treatment by combined surgery and radiation therapy. J Neurosurg 18:217–226, 1961.
40. Kramer S, Southard M, Mansfield CM: Radiotherapy in the management of craniopharyngiomas: Further experiences and late results. AJR 103:44–52, 1968.
41. Kubota T, Ito H, Aizumi S, Yamamoto S: Treatment of craniopharyngioma estimated by follow-up CT. No Shinkei Geka 9:1495–1501, 1981.
42. Landolt AM: Die Ultrastruktur des Kraniopharyngeoms. Schweiz Arch Neurol Neurochir Psychiatr 111:313–329, 1972.
43. Laws ER Jr: Transsphenoidal microsurgery in the management of craniopharyngioma. J Neurosurg 52:661–666, 1980.
44. Leksell L, Backlund E-O, Johansson L: Treatment of craniopharyngiomas. Acta Chir Scand 133:345–350,1967.
45. Long DM, Chou SN: Transcallosal removal of craniopharyngiomas within the third ventricle. J Neurosurg 39:563–567, 1973.
46. Love JG, Marshall TM: Craniopharyngiomas (pituitary adamantinomas). Surg Gynecol Obstet 90:591–601, 1950.
47. Matson DD, Crigler JF Jr: Management of craniopharyngioma in childhood. J Neurosurg 30:377–390, 1969.
48. Matson DD, Crigler JF Jr: Radical treatment of craniopharyngioma. Ann Surg 152:699–703, 1960.
49. McKissock W, Ford RK: Results of treatment of the craniopharyngiomas. J Neurol Neurosurg Psychiatry 29:475, 1966 (abstr).
50. McLone DG, Raimondi AJ, Naidich TP: Craniopharyngiomas. Childs Brain 9:188–200, 1982.
51. Metzger J, Nachanakian A, Gardeur D, Sichez JP: CT follow-up studies in postoperative craniopharyngiomas. Acta Neurochir (Wien) [suppl] 28:357–361, 1979.
52. Mori K, Handa H, Murata T, Takeuchi J, Miwa S, Osaka K: Results of treatment of craniopharyngioma. Childs Brain 6:303–312, 1980.
53. Mott FW, Barratt JOW: Three cases of tumour of the third ventricle. Arch Neurol Psychiatry 1:417–439, 1899.
54. Naidich TP, Pinto RS, Kushner MJ, Lin JP, Kricheff II, Leeds NE, Chase NE: Evaluation of sellar and parasellar masses by computed tomography. Radiology 120:91–99,1976.
55. Nakayama T, Kodama T, Matsukado Y: Treatment of inoperable craniopharyngioma with radioactive gold. No To Shinkei 23:509–513, 1971.

56. Northfield DWC: Rathke-pouch tumours. Brain 80:293–312, 1957.
57. Onoyama Y, Ono K, Yabumoto E, Takeuchi J: Radiation therapy of craniopharyngioma. Radiology 125:799–803, 1977.
58. Patterson RH Jr, Danylevich A: Surgical removal of craniopharyngiomas by a transcranial approach through the lamina terminalis and sphenoid sinus. Neurosurgery 7:111–117, 1980.
59. Ray BS: Surgery of recurrent intracranial tumors. Clin Neurosurg 10:1–30, 1964.
60. Richmond IL, Wara WM, Wilson CB: Role of radiation therapy in the management of craniopharyngiomas in children. Neurosurgery 6:513–517, 1980.
61. Rougerie J: What can be expected from the surgical treatment of craniopharyngiomas in children: Report of 92 cases. Childs Brain 5:433–449, 1979.
62. Shapiro K, Till K, Grant DN: Craniopharyngiomas in childhood: A rational approach to treatment. J Neurosurg 50:617–623, 1979.
63. Shillito J Jr: Craniopharyngiomas: The subfrontal approach, or none at all? Clin Neurosurg 27:188–205, 1980.
64. Shucart WA, Stein BM: Transcallosal approach to the anterior ventricular system. Neurosurgery 3:339–343, 1978.
65. Svien HJ: Surgical experiences with craniopharyngiomas. J Neurosurg 23:148–155, 1965.
66. Svolos DG: Craniopharyngiomas: A study based on 108 verified cases. Acta Chir Scand [suppl] 403:1–44, 1969.
67. Sweet WH: Radical surgical treatment of craniopharyngiomas. Clin Neurosurg 23:52–79, 1976.
68. Thomsett MJ, Conte FA, Kaplan SL, Grumbach MM: Endocrine and neurologic outcome in childhood craniopharyngioma: Review of effect of treatment in 42 patients. J Pediatr 97:728–735, 1980.
69. Till K: Craniopharyngioma. Childs Brain 9:179–187, 1982.
70. Trippi AC, Garner JT, Kassabian JT, Shelden CH: A new approach to inoperable craniopharyngioma. Am J Surg 118:307–310, 1969.
71. Van den Bergh R, Brucher JM: L'abord transventriculaire dans les crânio-pharyngiomes du troisième ventricule: Aspects neurochirurgicaux et neuro-pathologiques. Neurochirurgie 16:51–65, 1970.
72. Vladyková J: Kraniofaryngeom z hlediska oftalmologa. Cesk Oftalmol 29:109–116, 1973.
73. Vyramuthu N, Benton TF: The management of craniopharyngioma. Clin Radiol 34:629–632, 1983.
74. Weber FP, Worster-Drought C, Dickson WEC: Cholesterol tumour (craniopharyngioma) of the pituitary body. J Neurol Psychopathol 15:39–45, 1934.

COMMENT

The authors have collected a remarkable series of very large craniopharyngiomas in children. They have shown that, with the tools of the modern neurosurgeon, these tumors can be safely removed. The authors have pointed out the value of the preoperative neuroradiological work-up in planning the surgical approach to the tumor. I agree with the authors that, if the A-1 segment of the anterior cerebral artery is not elevated, the chiasm has been pushed forward by the tumor, and an approach along the pterion and through the lamina terminalis becomes necessary.

The immediate postoperative follow-up computed tomographic scan is of extreme value in confirming that the tumor has been totally resected. Certainly, if this postoperative computed tomographic scan does show residual tumor, then reoperation and removal of the missed tumor should be undertaken during the postoperative period.

The authors point out that craniopharyngiomas usually invaginate the floor of the 3rd ventricle when they fill the 3rd ventricle, but they do state that they had one patient in whom the tumor actually lay free within the 3rd ventricle. All of the craniopharyngiomas that I have treated have invaginated the floor of the 3rd ventricle and filled the 3rd ventricle from the suprasellar area rather than arising primarily within the 3rd ventricle.

Although the size of a craniopharyngioma should never deter the neurosurgeon from attempting total removal, it is certainly much easier to remove a small craniopharyngioma than a very large craniopharyngioma. The authors are therefore to be congratulated on their success rate in totally removing these very large tumors in this group of children.

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