Craniopharyngiomas

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ABSTRACT: A review has been carried out of those patients with a craniopharyngioma who were treated initially in our institution and had a microsurgical excision of their tumour during the period of time when newer and sophisticated modalities of investigation and treatment were available. Twenty-nine patients with craniopharyngioma were treated initially by microsurgical excision of their tumour at the Hospital for Sick Children from January 1976 to June 1985. In 21 a total removal of the tumour was performed, and there has been no clinical evidence of recurrence. No deaths were recorded in any of the 29 patients. The principal morbidity has resulted from endocrine deficits that these patients exhibit.

RÉSUMÉ: Les cranio-pharyngiomes Nous avons fait une révision de tous les cas de cranio-pharyngiome traités initialement dans notre institution et dont l'excision microchirurgicale s'est produite pendant une période permettant l'emploi des méthodes d'investigation et de traitement les plus récentes. Vingt-neuf patients furent ainsi opérés au Hospital for Sick Children du janvier 1976 au juin 1985. Chez 21 patients il y eut exérèse totale de la tumeur et aucune rechute. On observa aucun décès et les seules complications notées furent celles dues au déficit endocrinien inhérent à ces patients.

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Craniopharyngiomas are benign tumours of epithelial origin which commonly present in childhood. Their method of management continues to be a topic of controversy; some authors advocate aggressive surgical management and others plead for a more conservative approach utilizing radiotherapeutic techniques.[1,2,3,4] Even though the histologic appearance is benign, many patients with this tumour demonstrate a progressively deteriorating course despite treatment, and die of their disease.

In recent years, there has been a radical change in the surgeon's ability to deal with craniopharyngiomas. The surgeon now has available marvellous imaging tools such as CT and MRI scanners, as well as sophisticated tools in the operating room including the operating microscope, the ultrasonic aspirator and the laser beam. We therefore felt it worthwhile to review those patients with craniopharyngiomas who presented to our institution for initial treatment and had a microsurgical excision of their tumour during the period when these newer modalities of investigation and treatment were available. We consequently reviewed those patients treated initially at our institution by microsurgical removal of their tumour during the period January 1976 to June 1985.

Case Material

Twenty-nine children with craniopharyngiomas were treated initially by microsurgical excision of their tumour in our institution during the period January 1st 1976 to June 30th 1985. There were 18 boys and 11 girls. Ten patients were between 4 and 8 years of age, nine between 9 and 11 years, and ten between 12 and 17 years. The history was typically short: 23 patients had a history of less than two years, and in 17 the history was less than one year.

Visual problems were common. Thirteen patients had papilledema; in seven of these there were no other visual problems. Nine patients had significant loss of visual acuity, usually affecting one eye more than the other. Fourteen patients had a visual field defect which consisted of a homonymous hemianopia in six. Two of these homonymous field cuts involved only the superior quadrants. All but one of the homonymous defects were rightsided. One patient had a bitemporal field defect. Three patients had temporal field cuts in the left eye.

Twenty-two of the 29 patients had an endocrinopathy with three showing obesity; 14 were excessively short and 11 had diabetes insipidus at the time of initial examination. One patient presented with precocious puberty.

Radiological Findings

Calcification was seen on radiological study in 20 of the 29 tumours. A large sella turcia was present in 11 patients.

Fifteen of the tumours were cystic, eight were solid and six were a combination of cyst and solid tumour. Hydrocephalus was present in 14 patients, being severe in 11 and mild in three.

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Eight of the 11 patients with hydrocephalus had a shunt inserted prior to resection of their tumour.

Raimondi initially pointed out that the position of the anterior cerebral artery indicated the position of the craniopharyngioma in relation to the chiasm.[5] We reviewed the angiograms of our patients for this relationship and found that they could be angiographically divided into three groups: Group A — No significant deviation of either the A1 segment of the anterior cerebral artery or the basilar artery (Figure 1). Group B — The A1 segment of the anterior cerebral artery was elevated and there was no posterior displacement of the basilar artery (Figure 2). Group C — The A1 segment of the anterior cerebral artery was not elevated and there was posterior displacement of the basilar artery and stretching of the posterior communicating arteries (Figure 3).

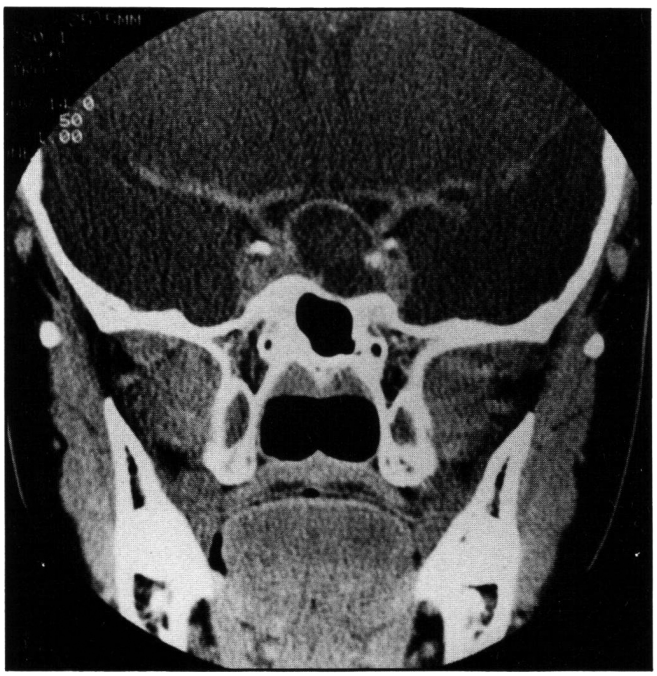


Figure 1 — CT scan showing small cystic craniopharyngioma arising out of sella and not elevating A1 segment of anterior cerebral arteries.

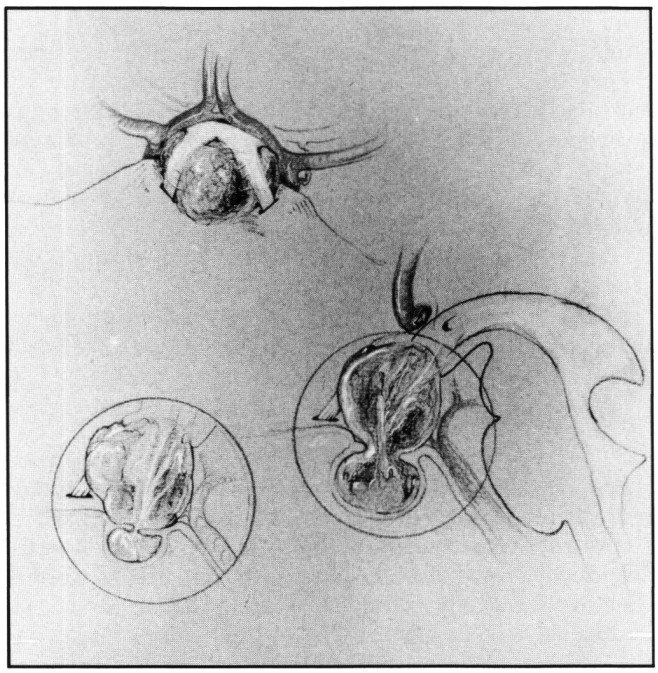
We found that this angiographic subdivision coincided with the positioning of the tumour. Tumours which were located within the sella and merely bulged a short distance above into the suprasellar area showed no deviation of either the A1 segment of the anterior cerebral artery or of the basilar artery, and thus had an A-type angiogram. Tumours which protruded anteriorly between the two optic nerves demonstrated a B-type angiogram. Tumours that protruded posteriorly, pushing the chiasm forward and producing the picture of a "prefixed" chiasm which abutted against the tuberculum sellae, had a C-type angiogram.

On the basis of CT scan we graded the tumours according to size. Those tumours which were 1-2 cm in diameter were called small. Tumours which were 2-3 cm in diameter were called moderate and those greater than 3 cm in diameter were called large.

Five tumours were small and all of those showed an A-type angiogram. Fifteen tumours were moderate in size and 11 of these were posteriorly placed with an angiographic picture described as C and four were anteriorly placed with an angiographic picture described as B. Nine tumours were large; four of these had an anteriorly placed tumour and a B-type angiogram and five had a posteriorly placed tumour with a C-type angiogram.

Surgical Approach

The position of the craniopharyngioma influences the operative approach and can influence the degree of difficulty that the



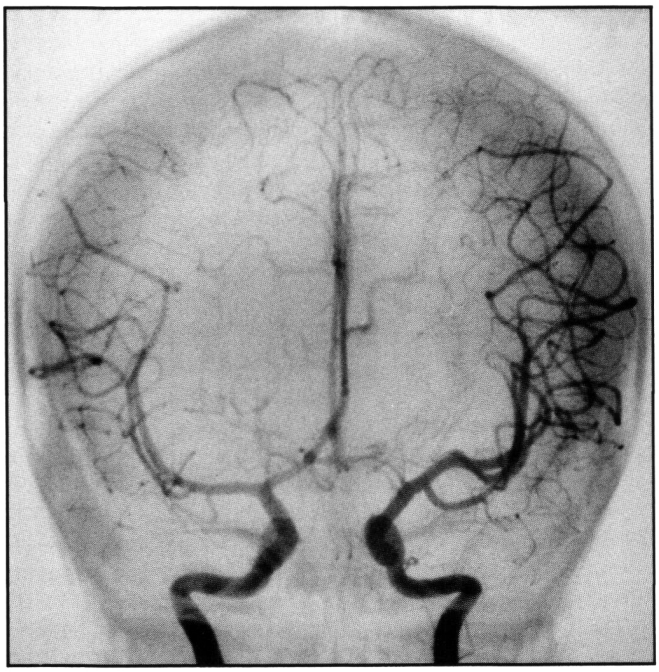


Figure 2 — A) - Drawing of Group-B craniopharyngioma showing tumour protruding between optic nerves and elevating A1 segment of anterior cerebral arteries. B) - Angiogram in Group-B craniopharyngioma showing elevation of A1 segment of anterior cerebral arteries.

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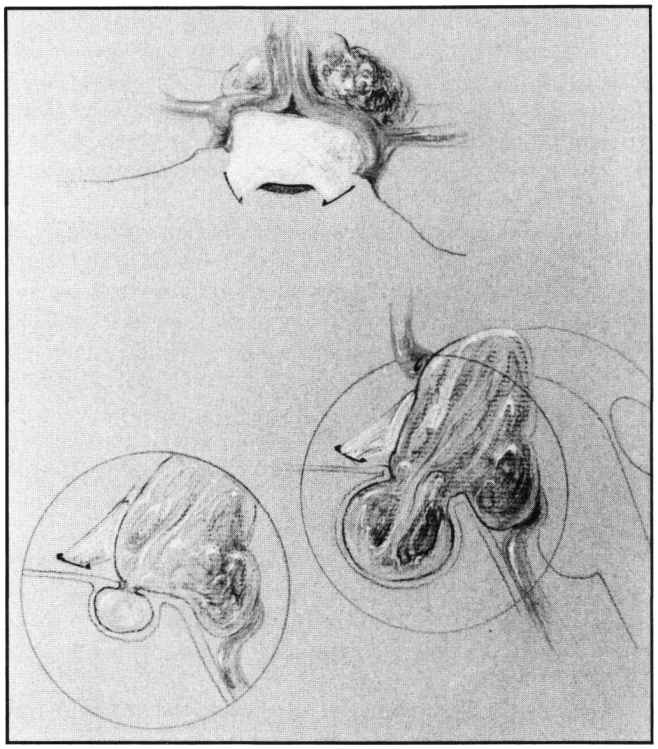

Figure 3 — A) - Drawing of Group-C craniopharyngioma which is protruding posteriorly and pushing thinned optic chiasm against clivus.

surgeon experiences in removing the tumour. Craniopharyngiomas can protrude forward between the two optic nerves and readily present when the frontal lobe is elevated. On the other hand, craniopharyngiomas which protrude backwards and push the chiasm up against the tuberculum sellae are hidden by a thinned optic chiasm so that there is no visible tumour when the frontal lobe is elevated.

We position our patients supine with the head in a pin fixation headrest, the neck extended and the nose pointing directly upwards. This position avoids distortion of anatomical relationships and facilitates the surgeon's orientation to optic nerves, internal carotid arteries and hypothalamus. A rightsided approach to the tumour is initially preferred, in that the right hemisphere is usually nondominant and consistency of approach facilitates anatomical orientation. In cases in which there is no suggestion of a posteriorly placed tumour pushing the chiasm forward, a small frontal bone flap is turned. If, however, the angiographic picture is that of C-type with the possibility of a prefixed chiasm, then the flap is extended inferiorly to allow for removal of the pterion.

With the frontal lobe elevated and an A or B-type angiogram, the craniopharyngioma can be readily visualized between the optic nerves. In patients with a B-type angiogram, the tumour protrudes below the optic nerves and frequently impinges on the internal carotid arteries. When the chiasm is pushed forward by a posteriorly placed tumour, the tumour cannot be visualized by the subfrontal route because the chiasm, optic nerves and optic tracts form a sheet which completely cover it. In such cases, the tumour is removed by incising the lamina terminalis, removing the tuberculum sellae, or approaching the tumour through a transpterional approach, separating the right optic nerve from internal carotid artery and extracting the

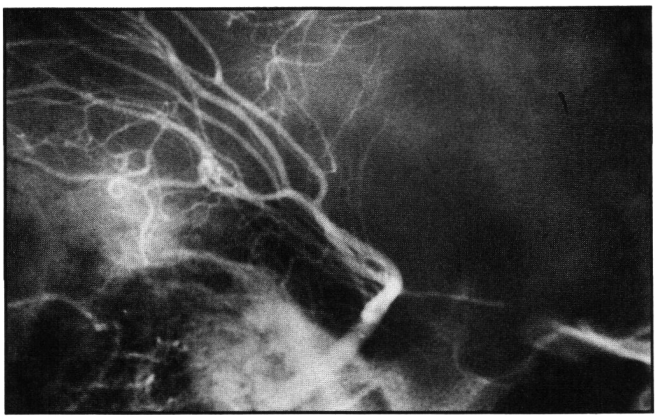

Figure 3 — B) - Angiogram of Group C craniopharyngioma showing basilar artery pushed back by retrochiasmatic tumour.

tumour between these two structures. Frequently a combination of these maneuvers is necessary, allowing tumour to be removed through lamina terminalis as well as between the optic nerve and internal carotid artery.

Once the craniopharnygioma is visualized, the capsule is incised and the fluid contents are aspirated. In cases of solid tumours, the tumour contents were removed with the ultrasonic aspirator. The craniopharyngioma capsule collapses and traction can then be applied to the capsule and the filmy adhesions between tumour and adjacent structures released. The tumour typically is easily separated from the optic nerves and chiasm and from the internal carotid artery and posterior communicating vessels. The membrane of Lilliquist is always intact at the time of initial removal of the craniopharyngioma. It thus provides a barrier between tumour and basilar artery and posterior cerebral vessels. Occasionally, the tumour is adherent to the internal carotid artery and posterior communicating vessels with particular tenacity, and it is in such cases that some tumour may have to be left behind. Care must be taken of the third cranial nerve, because the tumour can adhere to this structure which may be damaged during removal of the tumour.

Once the tumour has been dissected from the major vessels and optic nerves, the capsule can be forcibly pulled from its attachment to the tuber cinereum. The glial reaction within the tuber cinereum to invaginating tumour prevents damage to surrounding hypothalamus, although the tuber cinereum itself will sustain injury as the tumour is removed, thus accounting for the high incidence of postoperative diabetes insipidus in our series of patients.

Inferiorly, the tumour can be adherent to the diaphragma sellae and on occasion the diaphragma sellae can be deficient with tumour contents freely invading pituitary gland.

Results

Postoperatively, our patients were assessed clinically, neuroophthalmologically, endocrinologically, psychologically and by CT scan. The postoperative CT scan was assessed in an arbitrary fashion using the following scheme: Grade 1 — Normal CT scan without any evidence of residual tumour. Grade 2 — Postoperative CT scan which shows a tiny calcific fleck without evidence of enhancement and without any evidence of mass. Grade 3 — Postoperative CT scan which shows a small calcific chunk without any evidence of enhancement or mass

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effect. Grade 4 — Postoperative CT scan which shows evidence of a small contrast enhancing lesion, without mass effect. Grade 5 — Postoperative CT scan which shows evidence of a contrast enhancing lesion which does have a mass effect.

Twenty-five patients were felt to have had a total excision of their tumour at their initial operation. In four patients, visible tumour was left at the time of initial surgery because of adherence of tumour to important surrounding structures. In all four of these patients, the postoperative CT scan showed residual tumour (Grade 5), and clinical symptomatic recurrence of tumour occurred within a matter of months. All four patients had a subsequent operative approach to their tumour followed by a course of radiotherapy. They all remain alive, between three and nine years after treatment. All showed decrease of tumour size on CT scan after radiotherapy, with a final CT scan showing a Grade 2 picture in one patient, a Grade 3 picture in two patients and a Grade 4 picture in one patient. Two of these four patients continue to have a normal I.Q., one is mildly retarded (I.Q. 73) and one is significantly retarded with a full scale I.Q. of 62.

Twenty-five patients had what was interpreted at the time of operation as total removal of their tumour. In one of these patients the postoperative CT scan showed a considerable amount of tumour within the third ventricle with a Grade 5 CT picture. This patient was reoperated ten days later through a transcallosal route and the residual tumour removed.

The postoperative CT scans on the remaining 24 patients who had total removal of their tumour ranged between Grades 1 and 5. In ten patients the CT scans were Grade 1, and in none has the tumour recurred during a mean follow-up of 2.6 years. Five patients had a Grade 2 postoperative CT scan and none of these patients have shown recurrence. Two patients had a Grade 3 CT picture and have shown no evidence of recurrence. Six patients had a Grade 4 CT scan and 2 of these patients have had symptomatic recurrence, one at 3 years after initial surgery and one at 8 months. One patient had a Grade 5 CT scan and showed evidence of clinical recurrence 2 years following initial surgery.

All three patients with clinical recurrence were treated with a course of radiotherapy following reoperation for recurrent tumour. I.Q. testing of these 3 patients showed that they are functioning within the normal range with a full scale I.Q. of 86, 90 and 91. The final CT scan in these 3 patients following radiotherapy continues to show residual pathology and all have a Grade 4 picture.

Thus, 21 of the 29 patients have had no further treatment following their initial surgery. Eleven of these 21 patients now have a perfectly normal CT scan (Grade 1), four have a small fleck of calcium (Grade 2), two have a small chunk of calcium (Grade 3) and four have a small contrast enhancing lesion which may be residual tumour (Grade 4). These 4 patients remain at risk. Follow-up of these latter patients ranges between 1.5 and 6.5 years. They all remain well with no evidence of clinical recurrence.

The 21 patients with total resection and no recurrence to date have I.Q.'s ranging between 84 and 128.

Diabetes insipidus is common in our patients and 28 of the 29 patients are on treatment for this condition. The only one patient who is free of this disorder had a small tumour which was totally resected. This same patient is not on replacement Cortisone replacement has been necessary in all except 2 patients, both of whom had total removal of their tumour without recurrence. Twelve of our 29 patients receive growth hormone replacement. Only 7 of the 29 patients with a total removal and no recurrence are receiving growth hormone. Two of the total resections with recurrence are taking growth hormone and three of the partial resections take growth hormone. Sex hormone replacement is in current use in 8 of our 29 patients. This therapy only becomes necessary after the age of puberty. Excessive weight gain and obesity has been relatively common. It affects 9 of our 29 patients; only 5 of the 21 with total resection showed weight gain, whereas 4 of the 8 patients who received radiotherapy for recurrence show a tendency towards excessive weight gain.

Conclusions

Craniopharyngiomas are resectable tumours. They can be removed safely provided one uses the surgical microscope and is familiar with the anatomy of the sellar and parasellar regions. The operating microscope and surgical tools such as the ultrasonic aspirator and the laser beam, provide a safe, reliable method of excising these tumours. In our experience over 65% of craniopharyngiomas in childhood can be successfully, totally resected without serious morbidity and mortality, with the technological aids now available. The size, consistency and location of the tumour should not deter the surgeon's quest to totally remove it.

Anteriorly protruding tumours push the chiasm back, elevate the anterior cerebral arteries, distort the optic nerves and produce visual loss. These tumours are best approached via a subfrontal approach. Posteriorly protruding tumours push the chiasm forward, do not distort the anterior cerebral arteries, push the basilar artery backwards, fill the third ventricle and produce hydrocephalus. These tumours require a transpterional approach in addition to the subfrontal approach. Small tumours do not distort the neighbouring blood vessels. They have no effect on the optic nerves and chiasm, and so visual problems are not present. They frequently enlarge the sella turcica, impair endocrine function, and the children typically present with headaches and endocrine problems. These tumours are also best managed by a subfrontal approach.

If, after total excision, the postoperative high resolution CT scan shows no tumour, or only a fleck or chunk of calcium, there appears to be no risk of tumour recurrence. On the other hand, if after total excision, some contrast enhancing tissue is still present, there is a significant risk (30%) of clinical recurrence.

If after what was felt to be a total excision, there is obvious tumour left behind on CT scan, then reoperation is indicated to remove residual tumour.

If only a subtotal excision can be performed because of the condition of the tumour, and its relation to surrounding structures, then postoperative radiotherapy is indicated. Without such therapy, symptomatic recurrence becomes evident in 6 to 12 months.

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