Surgical Experiences with Craniopharyngiomas

HENDRIK J. SVIEN, M.D.
Section of Neurologic Surgery, Mayo Clinic and Mayo Foundation, Rochester, Minnesota

During the 12-year period, 1951 through 1962, the author performed operations for craniopharyngiomas in 20 cases. These cases are reviewed here primarily from the standpoint of the various approaches to the tumor which were utilized; the symptoms, ocular findings, and endocrinologic aspects will not be considered in this paper.

In this series 12 of the patients were female and 8 male. Seven patients were 15 years of age or less, the youngest being 3 years of age; the oldest was 58 years old.

Direction of Growth

Carmichael demonstrated that the masses of epithelial cell rests from which craniopharyngiomas develop are present along the pituitary stalk and upper aspect of the pituitary gland (Fig. 1). As these tumors increase in size, growth may take a variety of directions, as demonstrated by findings at operation and from air studies in the cases in this series.

![Diagram](Image)

Fig. 1. Location of epithelial cell rests along pituitary stalk and upper surface of pituitary gland.

There were several cases in which the sella was enlarged.

1. The tumor may grow into the sella, enlarge it, and remain confined to the sella and the immediate suprasellar region. In 4 of the cases in this series the tumor was limited to the enlarged sella except for some growth upward against the optic apparatus immediately overlying the confines of the enlarged sella (Fig. 2A).

2. The tumor may grow into the sella, enlarge it, and also grow beneath and occasionally over the optic chiasm and behind the dorsum sellae, eroding it in some instances, and extending into the interpeduncular fossa elevating the floor of the third ventricle (Fig. 2B). There were 2 such instances in this series.

3. The tumor may grow into the sella, enlarge it and stay confined there. There was one such case in this series.

There were 13 cases in which the sella was normal in size.

4. The sella may be normal in size with the tumor confined entirely to the suprasellar space between the optic nerves. There were 3 such cases in this series (Fig. 3A).

5. The sella may be normal in size with the tumor presenting between the optic nerves and extending into the anterior fossa between the frontal lobes. In the one case of this type in this series the tumor presented essentially as a midline anterior fossa tumor (Fig. 3B).

6. The sella may be normal in size with the tumor growing up between the optic nerves, under and occasionally over the chiasm, and backward into the interpeduncular cistern, elevating the floor of the third ventricle (Fig. 4). In cases of this type, the dorsum of the sella is frequently eroded. There were 3 such cases of this type in this series.

7. The sella may be normal in size with the tumor lying entirely behind the optic chiasm in the interpeduncular fossa elevating the floor of the third ventricle (Fig. 5A). The dorsum of the sella may or may not be eroded.
Surgical Experiences with Craniopharyngiomas

Fig. 2. Enlarged sella. (a) Pneumoencephalogram showing enlarged sella with tumor projecting only slightly into the immediate suprasellar region. (b) Postmortem specimen in another case (no operation) showing tumor filling enlarged sella, extending into space between the two optic nerves, and also extending behind chiasm into interpeduncular space.

(Fig. 5B). There were 6 such cases in this series.

Another indication of the location and extent of the tumor is provided by calcification in the tumor which in some patients is evident in roentgenograms of the skull. This finding was present in 6 of these 20 cases, and in each instance indicated rather precisely the position of the tumor.

Thus in 7 instances the tumor was limited to the space between the optic nerves and chiasm (determined by the location of calcification when present, evidence from air studies and findings at operation). In all of these cases the tumor was cystic and contained from 5 to 35 cc. of characteristic fluid.

Avenues of Attack on the Tumor

In all but 1 of the 20 cases a unilateral transfrontal craniotomy was carried out. The tumor was removed through a subfrontal approach to the optic nerves and chiasm in

Fig. 3. Pneumoencephalograms showing normal-sized sella. (a) Tumor extending upward into the immediate suprasellar space. (b) Indentation of floor of anterior horn of lateral ventricle by a large tumor growing into the anterior fossa.
150
Hendrik J. Svien

Fig. 4. Pneumoencephalogram showing normal-sized sella with tumor extending into suprasellar space, projecting backward into interpeduncular cistern and almost occluding the third ventricle.

16 of these cases. A transventricular approach was used 5 times in 4 cases. In 1 other case, however, because no tumor was seen between the optic nerves or under or over the chiasm on this approach, the transventricular approach was used to remove the tumor. In 1 case the tumor did not present at the foramen of Monro and consequently the subfrontal exposure was used. Removal by the transventricular route was, however, carried out as a second procedure after recurrence had taken place in this case. A transantral-sphenoidal approach was employed in 1 instance in which the sella was excessively enlarged.

Subfrontal Approach. In 6 of the 7 cases in which the tumor was limited to the space between the optic nerves and chiasm, the subfrontal approach was utilized. In two cases the cystic tumor was completely removed, while in the remaining cases radical subtotal removal was accomplished, leaving only bits of capsule attached intimately to the optic apparatus.

The subfrontal approach was also used in 10 cases in which the tumor extended beyond the area bounded by the optic nerves and chiasm. In 7 of these cases the tumor was cystic. One of these cystic tumors projected forward into the anterior fossa between the frontal lobes. It contained about 25 cc. of fluid and was radically but subtotally removed. The patient died 7 years later of myeloma. At postmortem examination a cystic mass 1 cm. in diameter was present over the sella.

In 4 cases the portion of the tumor lying behind the chiasm was partially removed by entering the lamina terminalis. In one of

Fig. 5. (a) Postmortem specimen (no operation) showing tumor present behind chiasm. (b) Pneumoencephalogram showing tumor similar in size and location to that shown in (a).
these cases only cystic fluid was removed as
no solid tissue was seen. Three months later
the cyst had refilled, and was aspirated
through a burr hole. In another case, some
of the tumor could not be seen and, there-
fore, was not removed; it was an interesting
fact that much of the calcium deposit be-
hind the dorsum sellae was still demonstrable
in postoperative roentgenograms. In another
case in which this approach was employed,
removal of more than a portion of the tumor
was deemed unsafe. This patient had signs
of recurrence 15 months later and reoperation
was performed elsewhere. In the fourth case,
cystic fluid and solid tumor aggregating 2.5
cm. in diameter was removed; however,
signs of recurrence appeared after 2 years,
and at reoperation a large tumor was re-
moved by the transventricular route. This
case will also be mentioned in discussion of
the transventricular approach.

Lateral Infrachiasmatic-Optic Tract Ap-
proach. This approach, working between
the laterally retracted carotid artery and the
medially retracted chiasm and optic tract,
was employed in 4 of the 16 cases. In 2 cases
the tumor was located between the optic
nerves as well as behind the chiasm, whereas
in the other 2 cases it was limited to the ret-
rochiasmal region. In 3 cases total removal
of the tumor was accomplished with its cap-
sule intact by this approach. In the fourth
case, the tumor was massive and abutted on
the pons; although an aggregate of tissue
amounting to 3.5 cm. in diameter was re-
moved, an equal amount remained on the
opposite side. After removal of all that could
be feasibly taken, the cerebral peduncles
were visible. The patient died 6 days later
from thrombosis of the longitudinal sinus
leading to extensive cerebral infarction.

Transantral-Sphenoidal Approach. In 1
case, in which the sella was excessively en-
larged and projected deeply into the sphen-
oid sinus, a cystic tumor was evacuated and
subtotally removed by a transantral-sphen-
odial approach (Fig. 6).

Transventricular Approach. This method
was employed 5 times in 4 cases. In all in-
stances the tumor was located entirely be-

Fig. 6. Roentgenogram of skull showing enlarged
sella dipping deeply into sphenoid sinus.
procedure in this case, and ventricular drainage was established for several days of preoperative preparation. While the head was being shaved preparatory to operation, the patient stopped breathing but was revived. The tumor, which practically obliterated the third ventricle, was approached transventricularly and was removed radically. A total of 35 cc. of cystic fluid was removed, together with an estimated two-thirds of the capsule. The patient never regained consciousness.

Another death followed partial removal of a solid tumor which occupied the suprasellar and interpeduncular space. Partial removal was carried out through the lateral infrachiasmatic-optic tract route. At autopsy examination it was clear that only about one half of the tumor had been removed.

The 3rd death occurred in a demented patient with optic atrophy 9 days after satisfactory total infrachiasmatic removal of a large tumor, obliterating the third ventricle and also presenting under the right optic nerve. The tumor, partially cystic, appeared on air study to be about 4 by 3 cm. in size. Electrolyte imbalance with hypernatremia developed after operation and the patient died. At postmortem examination no tumor tissue remained but chronic necrosis and compression of the hypothalamus were present.

Other Deaths. Three patients died at intervals of 2½ months, 5 months and 7 years after operation. The first two of these deaths were probably related to unsuccessful treatment of acute infections. In one a solid tumor had been subtotally removed by the subfrontal route and vision had improved considerably during her hospital stay. The other patient had a cystic tumor limited to the area confined by the optic nerves and chiasm; all of the tumor except a small portion of the capsule was removed. Both patients did very well after operation and were dismissed to the care of doctors in their home communities. These operations were done in 1952, however, and in those days none of us was as knowledgeable concerning proper supportive measures as we hope we are today.

The 3rd patient in this group died of myeloma 7 years after operation. He was demented when first seen and was thought to be suffering from cerebral degenerative disease. A huge tumor projecting between the frontal lobes was radically removed. He remained abnormal mentally, but otherwise was intact until his death.

Condition of Survivors. Of the 13 patients who are known to be living at present, 1 is being studied because of signs of recurrence 1½ years after operation. This patient had a solid tumor over and behind the sella; it could be only partially removed. One patient is a pituitary dwarf whose symptoms appeared, and who was operated on, before puberty. He is mentally retarded and lives in an institution. His tumor was entirely behind the chiasm and was removed by the transventricular route following signs of recurrence 2 years after incomplete removal by the subfrontal approach. Another patient, operated on during puberty, has signs of pituitary deficiency. His tumor was limited to an enlarged sella and the immediate suprasellar region; complete removal was effected. He is also congenitally deaf and has psychiatric difficulties. Had rehabilitation facilities been available where he lives, he could have learned a simple skill.

Two patients are working at jobs less exacting than those they held previously. One, whose symptoms suggested cerebral degenerative disease, was an executive before his symptoms began but now has some difficulty in learning new methods and skills. His tumor was limited to the region of the third ventricle and was removed through a transventricular approach. He currently works satisfactorily as an accountant. The other patient was admitted with memory and visual loss. His tumor was entirely behind a prefixed chiasm and was removed by the lateral infrachiasmatic-optic tract avenue. Prior to operation he was an unskilled laborer. Because of his visual loss, he cannot drive a car. This limits his capacity for employment.

Eight patients have returned to their former employment; the ministry in 1 case,
teaching in high school in another, secretarial work in another, and the role of housewife in the remaining 5. All of these patients are entirely normal. In 4 of them the tumors were limited to the space confined by the optic nerves and chiasm. In 1 patient the tumor was under and behind the chiasm. In the remaining 3 patients in this group the tumor was entirely retrochiasmal. The tumor of 1 patient was partially removed by the transventricular route; in another, total removal was effected through the lateral infrachiasmatic-optic tract approach. In the 3rd case aspiration followed by roentgen therapy was used on 2 occasions.

*Surgical Treatment Alone.* Roentgen therapy was not given to 7 patients. Four of these 7 patients are still alive, 11, 5, 21/2 and 11/2 years after operation. Another patient was alive and well 3 years after operation, but was then lost to follow-up. Two patients died 21/2 and 5 months after satisfactory surgical convalescence, from what appeared to be inappropriate substitution therapy during acute infections. All of the surviving patients had radical removal of the tumor.

*Roentgen Therapy.* Roentgen therapy was given to 10 patients in the postoperative period. Nine have survived: 2 for 10 years, 1 for 7 years, 1 for 6 years, 1 for 3 years, 1 for 2 years, 2 for 11/2 years, and 1 for 1/2 year. One patient died of myeloma 7 years after surgical and roentgen therapy. In 2 of the patients, of whom 1 has survived for 10 years and 1 for 7 years, radical removal was accomplished, and in 1, who has survived for 2 years, it was thought that a total removal was done.

The evidence of benefit from roentgen therapy is noteworthy in 4 cases. One patient, who had a recurrence of the tumor 2 years following drainage of a cyst and incomplete removal of the tumor via the lamina terminalis approach, had subtotal removal of the tumor by the transventricular route followed by a course of roentgen therapy. The patient is alive without signs of recurrence 8 years after the 2nd operation. Another patient is alive 6 years after subtotal removal via the transventricular route and postoperative roentgen treatment. One patient, who had had subtotal removal of a large cystic tumor presenting between the frontal lobes followed by roentgen therapy, died 7 years later from myeloma. At postmortem examination only a cystic mass 1 cm. in diameter remained. The most striking example of benefit from roentgen therapy is that of a patient who had only drainage of a large cystic tumor presenting behind the chiasm. Three months after operation aspiration of the tumor was carried out again and roentgen therapy was given. At that time he was markedly confused and had required custodial care for 4 months. He then improved progressively and is currently back in his former position as a high school teacher. He has maintained his improved state for 11/2 years.

The results in these 4 cases have impressed me with the efficacy of postoperative roentgen therapy and now use it in all of my cases. This experience supports the observations of Love and Marshall,6 of Leddy and Marshall,3 and of Kramer *et al.*,3 that roentgen therapy is beneficial in some of these cases.

**Selection of an Operative Approach**

Pneumoencephalography, with special attention to filling the subarachnoid cisterns, is a valuable aid in the selection of the surgical approach to the tumor. From it we can determine both the extent of obliteration of the third ventricle and the degree of invasion of the interpeduncular cistern can be described. In Fig. 3a the interpeduncular cistern is not invaded, while in Fig. 4 the tumor occupies the interpeduncular cistern and elevates the floor of the third ventricle so that it almost occludes the foramen of Monro. These points of assessment of the line of growth taken by the tumor, plus the determination of whether or not the lateral ventricles are enlarged, are essential to the selection of the best approach to the tumor.

If the tumor is confined in the suprasellar region in the area outlined by the optic nerves and chiasm, the approach is obvious and straightforward. If the tumor is situated in part or entirely behind the chiasm, there
are several choices.

Approach through the lamina terminalis just behind the chiasm has not proved generally effective. Usually not enough of the tumor wall can be seen and uncovered to permit mobilization and removal of the solid portion of the tumor even though the cystic portion of the tumor can be evacuated. In addition, bleeding from within the tumor after partial intracapsular removal is difficult to control because of the inaccessibility of the greater portion of the tumor.

The transventricular approach to a tumor which elevates the floor of the third ventricle sufficiently to occlude one or both foramina of Monro and produce hydrocephalus proved to be reasonably feasible in 4 cases. In these cases the tumor projected into one or both foramina of Monro and usually enlarged this opening. The top of the tumor can be exposed by transecting the portion of the fornix that lies anteriorly; a good portion of the body of the tumor can be exposed and manipulated by carefully wiping away adjacent tissue. Thus, a generous, though subtotal, removal can be effected if the tumor presents primarily upward into the region of the third ventricle. If the tumor does not obliterate the anterior portion of the third ventricle and does not present into or at the foramen of Monro, this approach is considerably more difficult and hazardous, because the surgeon has to reach down into the third ventricle to grasp the uppermost portion of the tumor. Also, he must be willing to settle for subtotal removal. Krayenbuhl, Kahn, and Northfield, and others have condemned this approach because of the forbidding mortality.

The approach which I currently favor for the tumors presenting under and behind the chiasm, particularly if the growth extends into the interpeduncular fossa, is the lateral infrachiasmatic-optic tract avenue. In these cases, and in those in which the tumor grows directly upward from the sella, the optic apparatus is elevated by the tumor, so that in the space between the laterally displaced carotid artery and the elevated chiasm and optic tracts a relatively large tumor can be safely manipulated. Further exposure of the tumor can be obtained (Fig. 7) by gentle medial retraction of the lateral chiasm-optic tract complex. When this approach is used, the portion of the tumor which has grown back into the interpeduncular cistern can be seen and dissected away from the tissue adjacent to it. Fig. 8 shows a case in which the chiasm and optic tracts were at the top of the tumor. The chiasm in this case was prefixed. Thus, a generous portion of the tumor could be attacked under direct vision from the space between the elevated optic apparatus and the laterally retracted carotid artery. The portion of the tumor which extended

![Fig. 7. Diagram of large tumor elevating optic apparatus and projecting backward into interpeduncular fossa. The tumor is more accessible to removal from a lateral infraoptic-chiasmatic approach.](image_url)

![Fig. 8. Pneumoencephalogram showing tumor extending upward and elevating markedly the optic nerves, chiasm, and tracts, which are pointed out by the arrows.](image_url)
Surgical Experiences with Craniopharyngiomas

into the interpeduncular space could be seen well. The fact that this tumor was cystic facilitated total removal of the remaining tumor (2.5 by 2.5 cm.). Fig. 9 shows the tumor removed in this case.

Summary

Study of the operative approach, procedures and results in 20 cases of craniopharyngioma permit the following conclusions: For craniopharyngiomas which elevate the chiasm, the lateral infrachiasmatic-optic tract approach is preferred. For tumors limited to the sella and immediate suprasellar region, in which the sella is enlarged and projects deeply into the sphenoid sinus, the transantral-sphenoidal approach is preferable. There is obvious merit in removing the tumor radically or completely but life should not be unduly jeopardized by this procedure since evidence strongly suggests that roentgen therapy after radical removal is beneficial and effective.

References