Craniopharyngiomas in children and adults

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The records of 51 patients with craniopharyngiomas were reviewed to compare the clinical features of the tumor in children and adults, to weigh the results of radical removal of the neoplasm, and to evaluate the benefit of postoperative radiation therapy. Signs and symptoms in the two age groups differed modestly. Endocrine abnormalities and visual field deficits were recognized more often in adults, whereas skull films were more likely to be abnormal in children. Six patients underwent radical removal of tumor. All have returned to normal life and remain free of recurrence. When tumor removal was incomplete, recurrence was common, appearing earlier in the younger patients. The administration of postoperative radiotherapy following partial removal of tumor appeared to delay recurrence in both age groups.

KEY WORDS craniopharyngioma parasellar tumor recurrence radiation therapy radical removal brain tumor

Craniopharyngiomas have challenged the skills of neurosurgeons since Erdheim described the tumor in 1903 and Lewis, 7 years later, reported the first attempt to remove it. For many years, the operative approach remained difficult, excision was rarely complete, and the mortality rate was prohibitive. Excessive morbidity commonly followed surgical treatment, the major disabilities being decreased vision and endocrine insufficiency. Then, as now, the fear of recurrence added to the difficulties of long-term management.

Earlier, therapy was restricted to the evacuation of cysts and resection of easily accessible tumor tissue. When the threat of adrenal insufficiency was removed, a gratifying fall in the immediate mortality from operation resulted, and interest in radical removal of the neoplasm was rekindled. Then the risk of prolonged disability from hypothalamic damage after radical excision became more clearly defined. Consequently, total extirpation has been reserved primarily for children since they are thought to be more susceptible to recurrence.

Most surgeons currently believe that a craniopharyngioma should be excised when possible in children and should at least be resected in adults. Some surgeons advocate postoperative radiotherapy, others deny its value, and many remain uncommitted. The purposes of this report are: 1) to compare the natural history of the craniopharyngiomas in children and adults who have been treated surgically at the New York Hospital-Cornell Medical Center; 2) to weigh the results of radical removal; and 3) to evaluate the benefits of postoperative radiation therapy.

Clinical Material

Of 61 patients with craniopharyngiomas treated from 1932 through 1967, 51 who underwent operation form the basis of this report. Sixteen were children between the ages of 1 week and 13 years (six boys, 10 girls). Six were between 14 and 21 years
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Fig. 1. Graph showing symptoms of patients with craniopharyngioma upon admission to the hospital. "Children" include those patients under 14 years of age (16), "adults" those over 14 years (35).

Symptoms

The duration of symptoms before diagnosis ranged from several months to several years, averaging 8 months. The most frequent complaint was failing vision: the adults frequently noted defects in their visual fields whereas the children often did not appreciate visual loss until total blindness approached. Symptoms suggesting intracranial hypertension frequently prompted the patient's first visit to a physician. Headache, vomiting, and behavioral changes were prominent symptoms in 63% of the children and 59% of the adults. There was a history suggesting endocrine deficiency in only one-quarter of the children; more than half of the older patients complained of symptoms of hypogonadism such as delayed pubescence, amenorrhea, or loss of sexual potency. About one-quarter of the patients in each age group complained of disorders that might be attributed to hypothalamic dysfunction, particularly somnolence and polyuria (Fig. 1).

Physical Signs

Visual field deficits were present in 86% of the adults: bitemporal hemianopia in 21 of 35, and homonymous defects in 10. A satisfactory eye examination was possible in 12 of the 16 children: 58% had marked visual loss. Papilledema was uncommon (19% of children, 14% of adults), but optic atrophy was frequent (56% of children, 37% of adults). Signs of pituitary insufficiency were present in 31% of the children and 49% of the adults. Cranial-nerve palsies gave evidence that the tumor had compromised paraseptal structures in one-quarter of the children and one-third of the adults (Fig. 2).

Roentgenography

Plain skull films were often diagnostic of craniopharyngioma. Calcification was present in or above the sella turcica in 81% of the children, but in only 40% of the adults. An enlarged or eroded sella turcica suggested that a tumor was present in 31% of the children and in 51% of the adults. Interestingly, skull roentgenograms were entirely normal in one child and in seven adults. Contrast studies were used infrequently and rarely aided diagnosis, although on occasion they aided the surgeon's approach to the tumor.

Treatment

Excision of the tumor by a transcranial approach was accomplished at the first operation in only two adults and two children. In two adult patients who had undergone resection initially, the tumor was totally removed at a second operation. The remaining patients underwent transcranial resection of the tumor with removal of as much as appeared safe to remove and drainage of cysts when present.

Following partial removal of the tumor, 24 patients received radiation therapy, administered by either a low voltage or a cobalt-60 source. The calculated tumor dose ranged from 2750 to 4500 rads and was greater than 3000 rads in all but one patient. Operation for recurrent tumor was done
in 19 patients. These procedures were similar to the first, and postoperative radiation therapy was given if it had not been given before.

**Results of Therapy**

**Vision and Endocrine Status**

Surgical therapy with or without irradiation improved vision in 54% of the adults and 33% of the children. Vision was not altered in 29% of the adults and in 50% of the children. Although no patient became blind as the result of operation, the visual-field defects increased postoperatively in one child and in one adult. In addition, one adult lost all vision in one eye, but had improved vision in the other eye.

Most patients required corticosteroid and thyroid supplements postoperatively. An occasional patient needed posterior pituitary extract as well. Persistent diabetes insipidus was unusual.

**Recurrence and Radiotherapy**

The propensity of the tumor to recur, particularly in children, is amply demonstrated by Fig. 3 and by the observation that 42% of the patients required a second operation. Complete excision appears to provide the only safeguard against recurrence, since the six patients so treated have remained well for 7 to 26 years. Radiation therapy retarded, but was unlikely to prevent, recurrence among the children. Adults, on the other hand, often had long-lasting benefit from combined surgical and radiation therapy (Fig. 4, Table 1). Apparently the tumor grows at a slower rate in the older age group.

The overall mortality from the initial transcranial operation in 51 patients was 8%. Three of the four operative deaths occurred prior to 1950; the fourth death followed removal of a 100-gm tumor from an 8-day-old infant. In that child, autopsy revealed hypoplastic lungs and a massive intra-abdominal hepatoma in addition to the intracranial tumor. Since no deaths followed craniotomy for recurrent tumor, the overall operative mortality rate for 74 operations was 5%. From 1950 through 1971, 63 operations have been performed, with one death, for an operative mortality of 1.6%.

**Pathology**

Forty-nine of the original tumors were partially cystic. The other two, one in a child and one in an adult, were solid. Three cyst-containing tumors recurred as solid tumors, but no solid tumors recurred as cystic tumors.

**Comment**

In this series, the clinical features of craniopharyngioma in children differed modestly from those observed in adults. Endocrine abnormalities and visual-field deficits were recognized more frequently in adults, whereas roentgenograms of the skull were
TABLE 1

Recurrence of craniopharyngioma after the first course of therapy

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Follow-up Intervals</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>2 yrs R/N*</td>
</tr>
<tr>
<td>resection</td>
<td>12/18</td>
</tr>
<tr>
<td>resection &amp; radiotherapy</td>
<td>7/23</td>
</tr>
<tr>
<td>excision</td>
<td>0/4</td>
</tr>
</tbody>
</table>

* R/N = Number of recurrences of craniopharyngioma in the number of patients surviving.

more likely to be abnormal in children. Surgical decompression of the optic nerves and chiasm was beneficial to vision in both groups, since vision either remained stable or improved after operation in 83% of patients.

Partial removal of the tumor was of temporary value in most adults and of less benefit in children, since recurrences seem to appear earlier in the latter group. Postoperative radiation therapy delayed recurrence in both children and adults, but failed to prevent eventual recurrence, particularly in the children. Long-lasting relief did, however, occur in many of the older patients.

This study has confirmed the observations of others on the response of craniopharyngioma to surgical management. Excision, when it can be accomplished safely, is the best form of therapy. Use of the operating microscope may permit better dissection of the tumor from adjacent structures and thereby allow surgeons to accomplish radical removal more often. When the tumor cannot be excised, it should be resected and the visual pathways decompressed. Radiation therapy is advisable following incomplete removal of tumor since it appears to delay recurrence.

References


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