Benign cerebellar astrocytomas in children

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Object. Cerebellar astrocytomas are benign tumors of childhood known to be associated with excellent long-term survival in patients in whom complete surgical resection is possible. However, the roles of other factors—clinical, radiological, histological, and therapeutic—in the survival of the patient, tumor recurrence, and long-term patient outcome remain imprecise. The goal of this study was to examine these factors and their relationships.

Methods. To clarify these issues a retrospective review was conducted of 168 children who were surgically treated for a cerebellar astrocytoma at Hôpital Necker–Enfants Malades between 1955 and 1995. These patients' clinical files were examined, the histological characteristics of their tumors were reviewed, and their outcomes were assessed according to Bloom's scale and the Wechsler intelligence quotient test.

Of the 168 patients in the study, 91 were male and 77 were female with a mean age of 6.9 years and a mean follow-up lasting 7.7 years. Tumors were identified as being strictly located in the cerebellum in 76.2% of the patients and as involving the brainstem (referred to as the "transitional form") in 23.8% of the patients. Complete surgical excision was possible in 88.7% of cases. There was a total mortality rate of 4.2% and a tumor recurrence rate of 9.5%. Fifty-eight percent of the patients had no neurological sequelae at follow-up evaluation.

Pejorative factors that were discovered by multivariate analysis to be important included: a long preoperative duration of symptoms and the transitional form of tumor with respect to survival; incomplete tumor excision with respect to an increased risk of recurrence; and a long preoperative duration of symptoms, an early epoch during which surgery was performed (1955–1974), severe ventricular dilation, and the transitional form of tumor with respect to a poorer long-term patient outcome.

Conclusions. The presence of brainstem involvement (tumor in the transitional form) emerged as a significant negative prognostic factor and should be treated as a distinct nosological entity. The extent of surgical excision has a significant bearing on the risk of tumor recurrence.

Key Words • astrocytoma • cerebellum • prognostic factors • prognosis • surgery • children
the neurosurgical department of the Hôpital Necker–Enfants Malades between September 1955 and December 1995. In three (2%) of these children a malignant astrocytoma was diagnosed; these children were not included, leaving 168 patients in the study.

For the purposes of analyzing the patients’ prognoses, we divided the 40-year period into three epochs: 1955 to 1974 (17 patients [10.1%]), 1975 to 1984 (66 patients [39.3%]), and 1985 to 1995 (85 patients [50.6%]). The second and third of these epochs corresponded respectively with the availability of the computerized tomography (CT) scanner and other global factors including improvements in anesthesiology, earlier diagnosis, and the use of magnetic resonance (MR) imaging, the ultrasonic aspirator, and microsurgical techniques.

Radiological Studies

According to findings on CT scans or MR images with and without addition of a contrast agent when available, the tumors were divided into four groups (Fig. 1): 1) cystic tumor with a nonenhancing cyst wall (only the mural nodule was enhanced); 2) cystic tumor with an enhanced cyst wall (the mural nodule and the cyst wall were enhanced); 3) tumor appearing falsely cystic (the tumor was largely necrotic with no identifiable mural nodule); and 4) solid or mainly solid tumor (often with irregular contrast enhancement).

Anatomical Localization

The anatomical localization of the tumor was based on radiological images and/or operative findings. Tumors were classified as being located in the cerebellar hemisphere or within the vermis. They were further classified according to the presence of brainstem involvement. It can be difficult to differentiate between tumors that originate primarily within the brainstem and those that originate in the cerebellum when both involve intermediary structures (the middle cerebellar peduncle). We thus defined tumors as being “transitional” when they arose predominantly within the cerebellum and then extended toward the brainstem (Fig. 2).

Surgical Findings

The surgical technique we used evolved during the period of this study in accordance with advances in technology. At present the standard treatment at our institution consists of the following: 1) placing the patient in the sitting or prone position, depending on the surgeon’s preference; 2) a suboccipital craniotomy in preference to a craniectomy, without removal of the arch of C-1 unless necessary; 3) preservation of the cisterna magna whenever possible; 4) horizontal incision of the cerebellum in those cases strictly located within the hemisphere; and 5) excision of the cyst wall according to the presence of enhancement on neuroimaging and the macroscopic appearance. In cases in which an abnormal enhancing cyst wall, viewed macroscopically, could not be removed, it was coagulated using a defocalized CO₂ laser, a contact laser, or bipolar coagulation.

A CT scan was obtained within 48 hours postoperative-
Cerebellar astrocytomas

Fig. 2. Magnetic resonance image displaying the transitional form of cerebellar astrocytoma.

Histological Data

Slides were examined twice by the same pathologist. Astrocytomas were classified as pilocytic or nonpilocytic according to the classification developed by the World Health Organization.

Follow-Up Review

Patients were examined at 3 months, 1 year, and every 3 years thereafter when possible, with CT or MR imaging and neuropsychological testing. A diagnosis of tumor recurrence was based on the evolution of radiological signs, with the appearance of, or increase in, contrast enhancement.

Neuropsychological tests (Wechsler Intelligence Scale for Children) were performed and adapted to the patient’s age, and, whenever possible, the patient’s intelligence quotient (IQ) or developmental quotient was determined (normal IQ or developmental quotient was considered to be > 80).

The patients were divided into four groups at the time of their last examination, according to Bloom’s scale: Group I, no deficit; Group II, mild deficit but no interference with the patient’s daily life; Group III, moderate deficit but the patient remains independent; and Group IV, severe deficit and the patient is dependent on others.

Study of Prognostic Factors

The following data were analyzed to assess their prognostic significance as related to patient survival, tumor recurrence, and patient outcome: gender, patient’s age at time of surgery, duration of clinical symptoms before diagnosis, period during which surgery was performed, hydrocephalus, extent of tumor excision (complete or otherwise), brainstem involvement (transitional form), and tumor location (cerebellar hemisphere or vermis), radiological appearance, and histological subtype (pilocytic or nonpilocytic).

In addition, to understand what factors are important in influencing postoperative shunt insertion at any stage in the natural history of the lesion, we analyzed the role of postoperative pseudomeningocele and meningitis.

Statistical Analysis

Curves for length of survival and life without recurrence were constructed according to the Kaplan–Meier method and compared using the log-rank test. To analyze survival and recurrence data, we used the multivariate Cox model. Cases were censored at the time of the patients’ last clinical examination. Potential prognostic factors were studied first in a univariate and second in a multivariate analysis, using a stepwise selection of covariates. For an analysis regarding “quality of life,” which consists of ordinal data, we used logistic regression after regrouping the four Bloom’s scale groups into good results (Groups I and II) and poor results (Groups III and IV). Regarding hydrocephalus, a chi-square test or Fischer’s exact test was used.

Results

Clinical Data

Of the 168 children there were 91 boys (54.2%) and 77 girls (45.8%). The mean age at diagnosis was 6.9 years (range 8 months–16 years). Thirty-three children (19.6%) were younger than 3 years of age. The duration of symptoms before diagnosis ranged from 1 day to 11 years with a mean of 7 months and a median of 4 months and was similar before and after the age of 3 years.

The majority of the patients’ clinical symptoms (Table 1) were related to intracranial hypertension, and their clin-
ical signs (Table 2) were related to the location of the tumor. In two patients the diagnosis was serendipitous. Two patients had neurofibromatosis.

**Radiological Studies**

One hundred and thirty-nine patients underwent preoperative CT scanning both before and after a contrast agent had been administered; 65 of these patients also underwent MR imaging. Twenty-nine patients did not undergo CT scanning before surgery; their tumors were diagnosed by ventriculography. Twelve of these patients were treated during the early CT scan era; thus, radiological classification of these 29 patients was not possible. The four radiological subtypes are presented in Fig. 3.

The solid form of the tumor was equally distributed between the vermis and the cerebellar hemisphere; however, proportionately it was found more frequently in tumors involving the brainstem; 50% of transitional-form tumors were solid and 28.8% of the nontransitional form were solid.

**Anatomical Location**

The astrocytomas involved the vermis in 119 cases (70.8%) and were strictly located in the cerebellar hemisphere in 49 cases (29.2%). One hundred twenty-eight astrocytomas (76.2%) were purely cerebellar, whereas 40 (23.8%) were of the transitional form.

**Surgical Data**

In the 139 cases in which data were available, tonsillar herniation was present in 107 cases (77%) and tumoral herniation in seven cases (5%). No attempt was made to remove the cyst wall in patients who harbored cystic astrocytomas with a nonenhanced cyst wall (64 cases), although 10 of these patients underwent cyst wall biopsy. Of the 29 patients who harbored cystic astrocytomas with an enhanced cyst wall (29 cases), 10 underwent cyst wall biopsy only. In the other cases the cyst wall that appeared abnormal macroscopically was either surgically removed or was cauterized using a defocalized laser if complete removal was not possible.

One hundred forty-nine patients (88.7%) underwent a complete excision, whereas in 19 patients (11.3%) the excision was deemed incomplete. Of those latter patients all but four had tumor that involved the brainstem and were surgically treated either before or during the early CT scan era. Surgical resection was deemed incomplete because the degree of residual brainstem involvement could not be determined operatively or radiologically. Thus, complete excision was possible in 124 (96.9%) of the 128 cases of the pure cerebellar form compared with 25 (62.5%) of the 40 cases of the transitional form of astrocytoma.

**Postoperative Period**

Two patients (1.2%) died during the early postoperative period, one from an epidural posterior fossa hematoma and the other from hematemesis. Ten patients suffered from postoperative mutism, which resolved within 8 to 90 days postoperatively (mean 37 days). Eight of these 10 patients harbored a tumor of the transitional form. In all cases, the cerebellar incision was made via the vermis.

**Radiation Therapy**

Postoperative radiotherapy was used in five patients (3%), all of whom had tumors of the transitional form that were incompletely excised during the first surgical epoch. Radiotherapy was administered at a dose of 40 to 55 Gy to the posterior fossa over 5 weeks. One child underwent three sessions of cobalt therapy at another institution before being referred to our department.

**Presence of Hydrocephalus**

Hydrocephalus was present on preoperative imaging in 157 cases (93.4%). Ventricular dilation was considered to be mild in 40.6% and severe in 51.4%. Transependymal resorption was present in 48.3% of the cases. Seven patients received shunts prior to arrival at our institution. Hydrocephalus was treated preoperatively in 14 patients. In 12 of these patients treatment was predicated on the patient’s clinical state and was accomplished by ventricular puncture in one patient, external ventricular drainage

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**TABLE 2**

*Clinical signs at admission in 168 patients with benign cerebellar astrocytomas*

<table>
<thead>
<tr>
<th>Physical Sign</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>papilledema</td>
<td>89 (68.5)*</td>
</tr>
<tr>
<td>cerebellar signs</td>
<td>78 (46.4)</td>
</tr>
<tr>
<td>abducent nerve palsy</td>
<td>38 (22.6)</td>
</tr>
<tr>
<td>macrocephaly</td>
<td>28 (16.7)</td>
</tr>
<tr>
<td>pyramidal signs</td>
<td>23 (13.6)</td>
</tr>
<tr>
<td>nystagmus</td>
<td>15 (8.9)</td>
</tr>
<tr>
<td>cranial nerve palsies other</td>
<td>11 (6.5)</td>
</tr>
<tr>
<td>Parinaud’s syndrome</td>
<td>4 (2.3)</td>
</tr>
<tr>
<td>hemiparesis</td>
<td>3 (1.8)</td>
</tr>
<tr>
<td>hyperthermia</td>
<td>3 (1.8)</td>
</tr>
<tr>
<td>occipital bone deformation</td>
<td>1 (0.6)</td>
</tr>
<tr>
<td>scoliosis</td>
<td>1 (0.6)</td>
</tr>
<tr>
<td>exophthalmos</td>
<td>1 (0.6)</td>
</tr>
</tbody>
</table>

* Of 130 patients who underwent funduscopys.
Cerebellar astrocytomas

**TABLE 3**

*Neurological sequelae at the time of the last examination in 168 patients with benign cerebellar astrocytomas*

<table>
<thead>
<tr>
<th>Neurological Sequela</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>no deficit</td>
<td>98 (58.3)</td>
</tr>
<tr>
<td>mild cerebellar dysfunction</td>
<td>26 (15.5)</td>
</tr>
<tr>
<td>severe cerebellar dysfunction</td>
<td>4 (2.4)</td>
</tr>
<tr>
<td>psychiatric disturbance</td>
<td>10 (6.0)</td>
</tr>
<tr>
<td>facial palsy</td>
<td>6 (3.6)</td>
</tr>
<tr>
<td>oculomotor signs</td>
<td>6 (3.6)*</td>
</tr>
<tr>
<td>quadraparesis or paraparesis</td>
<td>5 (3.0)</td>
</tr>
<tr>
<td>hemiparesis</td>
<td>4 (2.4)</td>
</tr>
<tr>
<td>decreased visual acuity</td>
<td>4 (2.4)</td>
</tr>
<tr>
<td>blindness</td>
<td>2 (1.2)</td>
</tr>
<tr>
<td>postanoxia vegetative state</td>
<td>2 (1.2)</td>
</tr>
<tr>
<td>kyphosis</td>
<td>2 (1.2)</td>
</tr>
<tr>
<td>deafness</td>
<td>2 (1.2)‡</td>
</tr>
<tr>
<td>seizures</td>
<td>1 (0.6)</td>
</tr>
</tbody>
</table>

* One patient had Parinaud's syndrome.
‡ One patient had speech delay.

in five patients, and endoscopic third ventriculostomy in six patients. The last method has become the procedure of choice at our institution. In two patients prophylactic ventriculoperitoneal shunts were placed to avoid the rapid brain collapse seen during surgery in patients with severe ventricular dilation who are treated in the sitting position. There were no cases of upward herniation after shunt placement.

Sixteen patients received intraoperative management of their hydrocephalus (as was the surgical practice at that time) via placement of an external ventricular drain that was removed during the postoperative period.

Postoperative shunt placement was required in six patients (3.6%) within 1 to 234 days (mean 75 days, median 18 days). Four of these six patients harbored tumors of the transitional form. Four had had an incomplete excision.

**Histological Findings**

Pilocytic astrocytomas comprised 93.5% of the tumors. Of the 10 astrocytomas with a nonenhanced cyst wall found at biopsy, tumor was found in only one case. Of those cases in which the cyst wall was enhanced by addition of contrast agent, the tumor was not found in three instances. These findings included a cyst wall bordered by vascular arcades, glial tissue with many Rosenthal fibers, or no tumor.

**Tumor Recurrence**

Overall, there were tumor recurrences in 16 patients (9.5%). Of the 149 patients whose tumors were reported to be completely excised at the initial operation, eight (5.4%) suffered recurrences. Nineteen patients had an incomplete excision; eight (42.1%) of these had recurrences. Recurrences were proportionately higher in patients whose tumors were the transitional form: eight (20%) of 40 patients compared with eight (6.3%) of 128 patients with purely cerebellar tumors. The delay until the time of tumor recurrence ranged from 3 months to 9.3 years (mean 3.6 years). Of the five patients who underwent radiotherapy three had tumor recurrence.

Recurrences were managed by repeated surgery in 13 patients. Malignant transformation was seen in one patient who had undergone postoperative radiotherapy 9 years earlier. There were no postoperative deaths in these patients; complications included pseudomeningocele in three cases, meningitis in two cases, paraplegia in one case, and right facial palsy in one case. Two ventriculoperitoneal shunts were inserted after the second operation, in one case at 1 month and in the other at 5 months. No patients in this group underwent radiotherapy. The three patients with tumor recurrences who did not undergo a second operation were treated by radiotherapy.

A third operation was performed in four cases, in two of these cases to complete an incomplete excision, in one to achieve a more aggressive excision of the tumor shown to be an astrocytoma Grade II/III, and in one case to treat a true recurrence with significant brainstem involvement, which was subsequently treated with postoperative radiotherapy.

In an additional case, the second regrowth of residual tumor left after a subtotal excision of an astrocytoma of the cerebellopontine angle was not treated surgically but was treated with radiation by using a gamma knife.

**Follow Up and Outcome**

The mean duration of follow-up review was 7.7 years (range 10 days–40 years, median 59 months). Seven patients (4.2%) died, with an overall survival rate of 95.8%. Except for the two postoperative deaths, death was related to tumor in four cases and hydrocephalus in one case. Six children were lost to follow-up review (their follow-up periods were < 6 months).

The patients’ neurological sequelae are summarized in Table 3. One hundred fifty-five children underwent neuropsychological assessment. Of these, 19 (12.3%) had mild mental delay and seven (4.5%) had severe mental delay. These patients’ quality of life at the time of their last examination ranged as follows: Group I, 98 cases (58.3%); Group II, 36 cases (21.4%); Group III, 21 cases (12.5%); and Group IV, 13 cases (7.7%)

**Analysis of Prognostic Factors**

With respect to survival (Table 4), multivariate analysis showed that the pejorative factors were long preoperative duration of symptoms and the transitional form of tumor. Figure 4 depicts the rates of survival according to brainstem involvement. With respect to tumor recurrence
(Table 5), multivariate analysis showed that the pejorative factor was incomplete excision. Figure 5 depicts the rates of tumor recurrence according to the extent of surgery. Regarding quality of life (Table 6), multivariate analysis showed that the pejorative factors included long preoperative duration of symptoms, severe ventricular dilation, the surgical epoch (1955–1974), and the transitional form of tumor. Regarding the risk of shunt insertion (Table 7), an adjusted chi-square test showed that the risk factors were incomplete excision, recurrence with a second surgical procedure, and postoperative meningitis.

Discussion

Since the publication of early treatises on cerebellar astrocytomas,7,14,15 numerous authors have searched for factors that have a bearing on survival duration and tumor recurrence in patients harboring these tumors. Indisputably, one of the major prognostic factors is that of complete surgical resection.1,17,23,44,53,54,56,58,61 However, controversy surrounds the roles of other factors such as the tumor’s histological subtype, brainstem involvement, and solid structure and, secondarily and consequently, the role of adjuvant therapy. In this regard our results were largely additive to a host of others and aid only to fuel the controversy.

Surprisingly, what did become rapidly apparent during the evolution of this study was the primary role played by the presence of brainstem involvement. It emerged as the sole unifying factor that had both a direct and indirect bearing on prognosis with respect to survival, recurrence, and long-term outcome. In this regard we concluded that its presence should denote a distinct subgroup of astrocytomas, the transitional form.

The term “transitional form” was applied because it was believed that, perhaps, these tumors represented part of a spectrum of posterior fossa astrocytomas ranging from the very benign, purely cerebellar astrocytoma, which carries a 10-year patient survival rate of 90 to 100%,22 to the dramatically less favorable, diffuse brainstem astrocytoma, which has a 3-year patient survival rate of 10%.20,43,49,50 The dorsally exophytic brainstem tumors (with minimal deep brainstem involvement) are, similarly, a part of this spectrum and are described as having a favorable prognosis after surgical resection.32 Intrinsic cerebellar peduncle lesions are also accessible to surgical resection without postoperative deficit.80 In distinction to the latter group, the term “transitional form” defines those tumors that have a predominantly cerebellar origin but spread to involve the brainstem. In this group we found a 10-year survival rate of 80% (Fig. 4).

Other authors have reported on the negative prognosis that brainstem involvement incurs,24,27,53,54 but have not defined it as the single determinant. Our evidence for doing so was based on a number of factors.

Clinical Factors

Our study did not confirm that the patient’s age at presen-

TABLE 5

<table>
<thead>
<tr>
<th>Variable</th>
<th>p Value</th>
<th>Relative Risk</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>patient age</td>
<td>NS</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>decade of surgery</td>
<td>NS</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>vermis location</td>
<td>NS</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>solid tumor</td>
<td>NS</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>transitional form of tumor</td>
<td>NS</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>pilocytic tumor</td>
<td>0.0007</td>
<td>8.9</td>
<td>2.5–31.9</td>
</tr>
</tbody>
</table>

* Based on multivariate analysis using Cox’s model.

TABLE 6

<table>
<thead>
<tr>
<th>Variable</th>
<th>p Value</th>
<th>Odds Ratio</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>preop duration of symptoms</td>
<td>0.03</td>
<td>1.0</td>
<td>0.9–1.1</td>
</tr>
<tr>
<td>decade of surgery</td>
<td>0.003</td>
<td>3.16</td>
<td>1.4–6.8</td>
</tr>
<tr>
<td>patient age</td>
<td>NS</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>ventricular dilation</td>
<td>0.008</td>
<td>3.9</td>
<td>1.4–11.1</td>
</tr>
<tr>
<td>vermis location</td>
<td>NS</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>transitional form of tumor</td>
<td>0.02</td>
<td>3.2</td>
<td>1.13–9.09</td>
</tr>
<tr>
<td>pilocytic tumor</td>
<td>NS</td>
<td>—</td>
<td>—</td>
</tr>
</tbody>
</table>

* Based on multivariate analysis using logistic regression.
Cerebellar astrocytomas

### TABLE 7

<table>
<thead>
<tr>
<th>Variable</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>patient age</td>
<td>NS</td>
</tr>
<tr>
<td>decade of surgery</td>
<td>NS</td>
</tr>
<tr>
<td>preop duration of symptoms</td>
<td>NS</td>
</tr>
<tr>
<td>tonsillar herniation</td>
<td>NS</td>
</tr>
<tr>
<td>solid astrocytoma</td>
<td>NS</td>
</tr>
<tr>
<td>vermis location</td>
<td>NS</td>
</tr>
<tr>
<td>transependymal resorption</td>
<td>NS</td>
</tr>
<tr>
<td>degree of ventricular dilation</td>
<td>NS</td>
</tr>
<tr>
<td>transitional form of tumor</td>
<td>0.006</td>
</tr>
<tr>
<td>complete tumor excision</td>
<td>0.0003</td>
</tr>
<tr>
<td>postop pseudomeningocoele</td>
<td>NS</td>
</tr>
<tr>
<td>postop CSF leak</td>
<td>NS</td>
</tr>
<tr>
<td>postop meningitis</td>
<td>0.002</td>
</tr>
<tr>
<td>tumor recurrence</td>
<td>0.001</td>
</tr>
</tbody>
</table>

* Based on chi-square test.

Entation was related to that individual's prognosis, as has been previously reported. Rather, we showed that it was a short preoperative duration of symptoms, regardless of patient age, that significantly improved the length of survival and long-term outcome. The basis of this is unclear but may relate to a reduction in neurological sequelae due to a decreased duration of intracranial hypertension and to a diagnosis being made prior to significant brainstem extension.

### Radiological Characteristics

Solid astrocytomas have been associated with a poorer prognosis and shorter survival time. Our analysis did not support this contention. We showed that prognosis was significantly related to the presence of the transitional form of tumor. In our series these lesions were more often solid, and it is probably this fact that accounted for the negative prognosis attributed to the solid form by other authors.

### Histological Characteristics

Cerebellar astrocytomas are subdivided by the World Health Organization's classification into pilocytic and nonpilocytic tumors with the nonpilocytic subtype tending to be more diffuse and more infiltrative. Hayostek and colleagues and others found that pilocytic astrocytomas are associated with improved survival and recurrence rates. Austin and Alvord and others, however, showed no correlation between histological variables and tumor recurrence or cure.

We similarly found no correlation between histological subtype and prognosis. We did note, however, a higher incidence of the nonpilocytic subtype in the transitional form of the tumor (14.5% compared with 4.9%), and it is probably this fact that accounts for the poorer prognosis attributed to this subtype by some authors.

### Extent of Surgical Resection

Although the transitional form was not shown to be a significant independent risk factor in relation to tumor recurrence, it was associated with a higher incidence of incomplete resection (37.5% compared with 3.1%) for the purely cerebellar form and, consequently, a higher incidence of recurrence (20% compared with 6.3%).

Complete surgical resection is an important prognostic factor in relation to tumor recurrence and patient survival. We similarly showed that it is the sole independent variable that significantly determines the risk of tumor recurrence. Despite this, some authors still contend that a complete resection is not essential, citing several cases in which there was no tumor recurrence several years after partial removal.

Related to this issue is the management of the cyst wall. Contrast enhancement or its absence in this series did not accurately reflect the presence or absence of tumor in the cyst wall. In light of this and the fact that complete surgical resection significantly alters the risk of recurrence, we currently remove any macroscopically apparent abnormal cyst wall regardless of the tumor's radiological appearance. However, this issue is contentious and is currently the basis of a prospective study.

In our series, patients with transitional tumors had a higher proportion of incomplete resection with a higher proportion of tumor recurrence and, subsequently, a significantly poorer length of survival and poorer long-term neurological outcome. Thus via a combination of factors this group of patients behaved in a different fashion than those patients with a tumor located purely within the cerebellum, regardless of other tumor features. Having identified these children as belonging to a unique category of posterior fossa astrocytomas, our attention needs to be turned to improving their length of survival by reducing the risk of recurrence and to improving their quality of life.

### Radiation Therapy

The role of radiotherapy in preventing recurrence remains controversial because it is often administered in an inconsistent and uncontrolled manner, which occurred in our five patients (who represented only 26% of patients with an incomplete tumor resection). This is not surprising given the inhomogeneity of opinion in the literature and the lack of appropriate statistical analysis.

Some authors maintain that postoperative radiotherapy is beneficial after incomplete excision in obtaining local tumor control, delaying the time to recurrence, and improving the length of survival. The series conducted by Marsa and associates, Garcia and Fulling, and Wallner, et al., however, show a recurrence rate on the order of 34% in spite of a radiation dose of at least 40 Gy. In addition, Undjian and colleagues and Sgouros, et al., did not find any improvement in prognosis after postoperative radiotherapy even when it was administered to patients with incomplete excision and brainstem involvement. Unfortunately, our numbers were too small to be of statistical use.

At this stage, because of side effects that are not insignificant and the lack of adequate data to support its use, we do not propose the use of radiotherapy after surgery even if the excision is incomplete. Whether radiotherapy has a role in cases of tumor recurrence remains debatable; we would prefer repeated surgery in these cases.
Long-Term Outcome

A poorer long-term outcome was found by us to be associated with a long preoperative duration of symptoms, severe hydrocephalus, and the presence of brainstem involvement. Together these probably represent cumulative and somewhat irreversible insults on the nervous system, resulting in a reduced IQ and neurological deficit.

As surgeons we are able to have a direct influence on only one of these three parameters—the management of hydrocephalus. There are some authors who have proposed the systematic preoperative placement of shunts in patients.2,9,13 During the evolution of surgical therapy in our institution lasting more than 40 years we have found that ventricular shunts can be advantageously replaced by endoscopic third ventriculostomy. This has now become the procedure of choice at our institution. The need for preoperative treatment of hydrocephalus remains frequent (14 of 168 patients), despite an incidence of hydrocephalus of 93.4% in our series. At present we advocate performing a third ventriculostomy before definitive tumor excision only in those patients with hydrocephalus in whom it is probable that the cerebrospinal fluid (CSF) pathways will be breached during tumor removal.

Postoperative shunt placement, similarly, remains a rare event in our series and in those of others.15,26,57 We found incomplete excision, recurrence with a second operative procedure, and postoperative meningitis to be significant factors, suggesting a secondary disturbance of CSF resorption rather than a primary obstruction to CSF flow. These factors are largely within surgical control.

Conclusions

This study was undertaken to determine the factors influencing the outcome of children surgically treated for benign cerebellar astrocytomas. We discovered that the most important factor was the presence of brainstem involvement because it had a direct bearing on the length of survival and on neurological outcome as well as an indirect bearing on tumor recurrence because these patients were more likely to have had an incomplete surgical excision. We thus believe that these tumors should be regarded as a separate entity, as part of a spectrum of posterior fossa astrocytomas with a prognosis that is intermediate between that of the cerebellar astrocytoma and the benign brainstem glioma.

References

Cerebellar astrocytomas

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