Cystic pilocytic astrocytomas of the cerebral hemispheres

Surgical experience with 51 cases and long-term results

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This study concerns 51 cases of cystic pilocytic astrocytoma of the cerebral hemispheres. At operation these tumors closely resembled cerebellar astrocytomas, presenting as a big cyst contiguous with the lateral ventricle and containing a small mural nodule. The nodule was in a deep location in 26 cases, and always showed marked contrast enhancement on computerized tomography scans. The most frequent presenting symptom was epilepsy (68%), and the most common sign was papilledema (85%). The major goal of surgery was extirpation of the nodule. The follow-up period of the 34 cases available for long-term review ranged from 6 to 31 years (mean 17 years). Of these, 28 patients (82%) had a good outcome; four (12%) had a fair result, and two (6%) had a poor response. An analysis of these results shows that total extirpation of the mural tumor was associated with the best outcome, whether or not the cyst wall was completely removed. Radiation therapy was irrelevant to the prognosis for these patients. On the other hand, partial excision of the nodule, correlated with the deep location of the tumor, was the cause of the two poor results in this series and resulted in multiple operations for recurrences in two other patients. Nevertheless, two of these patients are still alive and well many years after incomplete surgical treatment. On the basis of this study, the importance of recognizing the occurrence of this "benign" tumor of the cerebral hemispheres is stressed, and the hypothesis of a common origin from subependymal glia of all pilocytic astrocytomas is supported.

KEY WORDS • brain tumor • pilocytic astrocytoma • astrocytoma • cerebral hemisphere • cerebellum

Pilocytic astrocytoma, the most benign variant of astrocytic tumors in the new World Health Organization classification, corresponds to the term “spongioblastoma” of Zülch and Wechsler and “juvenile pilocytic astrocytoma” of Russell and Rubinstein. This tumor is usually found in the cerebellum and optic-hypothalamic region, but it occasionally can be found in the cerebral hemispheres of young patients where it presents as a big cyst surrounding a small mural tumor.

There have been very few reports of surgical experience with this peculiar cerebral astrocytoma since Cushing's original observations. We are reporting a series of 51 cases of cystic pilocytic astrocytoma of the cerebral hemispheres that were operated on at the Institute of Neurosurgery of Rome University over a 32-year period.

Summary of Cases

Clinical Material

These 51 cases of cystic pilocytic astrocytoma represented 3% of 1678 gliomas of the cerebral hemispheres, and slightly less than 1% of 5441 brain tumors operated on at the Institute between 1951 and 1983. The mean age of patients at presentation was 18 years; two-thirds of the cases occurred in the first two decades of life, which was very similar to the age of onset in cerebellar astrocytoma cases at this Institute (Fig. 2). The average clinical history was 14 months. The most frequent presenting symptoms were epilepsy (68%), headache (63%), and vomiting (51%); the most common neurological sign was papilledema (85%), followed by hemianopsia (30%), and pyramidal tract signs (29%).

The temporal lobe was involved in 49% of the 51
cases and the parieto-occipital lobe in 35% (Table 1). The mural nodule was in a superficial location in 26 cases; most of these were subcortical but a few were visible on the surface of the cortex. In 43% of cases the mural nodule was located on the medial aspect of the cyst, in two cases the position of the nodule was not recorded, and in one case no mural tumor was found inside the cyst (Table 1).

The mural tumor always showed a marked area of contrast enhancement on computerized tomography (CT) scanning (Fig. 3), but carotid angiography seldom disclosed a tumor blush. It should be pointed out that the mural tumor was not always a prominent round nodule. In some cases it presented as a plaque on the cyst wall (Fig. 4 left), and in others as multiple masses, irregularly packed together (Fig. 4 right). In all cases the medial aspect of the cyst was contiguous with the lateral ventricle, which was separated from the cyst by the ependymal lining. The cysts contained between 90 and 320 ml of fluid, usually xanthochromic and coagulating on aspiration, but sometimes brown, as if secondary to previous hemorrhage. In some cases traces of past bleeding were noted on inspection of the cyst wall. The mural tumor itself was often described as reddish-brown and richly vascularized, thus explaining the enhanced CT image.

Treatment

At surgery, the cyst contents were aspirated, and the mural nodule was excised. The contiguous ventricular wall was then opened and removal of the cyst wall itself was attempted where possible, especially in the area of the nodule. The cyst wall was incompletely removed in half of the cases without adverse consequences. In six cases (12%) extirpation of the nodule was only partial because of the apparent involvement of critical areas. With increasing experience it was learned that separation from the brain tissue can also be achieved in deep areas almost with impunity. Partial extirpation, however, did not exclude a benign course with long survival. Radiation therapy was prescribed in 12 of the 51 patients, most of whom were over 15 years of age and had undergone partial resection.

Histological Study

Microscopic examination of all the surgical specimens showed a pattern that is usually seen in cerebellar astrocytoma, including loose and compact areas, microcysts, elongated or round cells, oligodendrogial cells, Rosenthal fibers, and cytoid or granular bodies (Fig. 5). This tumor entity was previously called “spongionblastosoma” in the German literature or “juvenile pilocytic astrocytoma” by Russell and Rubinstein, and is presently included under the name of “pilocytic astrocytoma” by the World Health Organization classification.

Long-Term Results

Two of the 51 patients died after the operation, five others were lost to follow-up review, and nine patients were operated on less than 5 years before this study and were therefore excluded. One other patient died sud-

TABLE 1
Location of the cyst and position of the mural nodule in 51 cases

<table>
<thead>
<tr>
<th>Location of Cyst</th>
<th>Position of Mural Nodule</th>
<th>Total Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Superficial</td>
<td>Deep</td>
</tr>
<tr>
<td>temporal lobe</td>
<td>12</td>
<td>13</td>
</tr>
<tr>
<td>parieto-occipital lobe</td>
<td>10</td>
<td>5</td>
</tr>
<tr>
<td>frontal lobe</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>total cases</td>
<td>26</td>
<td>22</td>
</tr>
</tbody>
</table>
Cerebral hemispheric pilocytic astrocytomas

FIG. 3. Computerized tomography scans before (left) and after (right) infusion of contrast medium, showing marked contrast enhancement of the mural tumor.

FIG. 4. Computerized tomography scans of cerebral pilocytic astrocytomas after infusion of contrast medium. Left: The mural tumor, appearing as a plaque lying on the inferomedial aspect of the cyst, is seen beyond the midline. Right: Example of a tumor presenting as multiple masses, irregularly packed together.

FIG. 5. Photomicrograph of a cerebral pilocytic astrocytoma showing the typical histological features, including pilocytic, stellate, and oligodendroglial cells, microcysts, cytoid or granular bodies, and Rosenthal fibers. H & E, x 400.

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Discussion

This study and an analysis of data in the literature raises two interesting points in our view. The first concerns the definite confirmation that cystic gliomas above the tentorium may be similar in all aspects to cerebellar astrocytomas. Such a similarity includes both clinical and pathological features. These are tumors of children and juveniles; 75% of our cases occurred in the first two decades of life, and only two of our patients were more than 40 years old. The increased incidence of cerebellar astrocytomas during the first decade (Fig. 2) can reasonably be explained by the earlier clinical onset of the symptoms generally seen in posterior fossa tumors. Other salient clinical features of these cystic cerebral astrocytomas that are similar to their cerebellar counterpart are their mainly benign biological behavior which results in the best long-term outcome for tumors in the astrocytoma group after surgical treatment, the possibility of long survival with good function even after partial excision, and the fact that the prognosis is unchanged whether or not the cyst wall has been completely removed or postoperative radiation is given.

Schisano, et al.,26 published the only other previous study of a similar size, and their conclusions are identical to ours. As they have already noted, pilocytic astrocytomas of the cerebral hemispheres can be solid tumors, as in the cerebellum. This was probably less well recognized in the past, so that only a total review of the old series would give a true idea of the incidence of this tumor variant. In this study, we considered only cystic pilocytic astrocytomas, and excluded seven cases of solid pilocytic tumors that we recently treated. The true incidence of cystic pilocytic astrocytoma among astrocytic tumors of the cerebral hemispheres, although admittedly low, remains difficult to assess because the specific reported surgical experience is scanty. The figure of 3% of cerebral gliomas in our series is similar to that reported in two other studies,6,10 but remains considerably lower than the 5% to 8% described by Olivecrona.14 It should further be pointed out that this figure is considerably higher in the pediatric age group, where we have found that this kind of tumor represented 22% of hemispheric gliomas and 11% of all supratentorial tumors.16

The second comment we want to make concerns the nature of pilocytic astrocytomas, cerebral as well as cerebellar. Without presuming to solve an old neuropathological controversy,16-19,23 in our view, the localization in the cerebral hemispheres of a tumor type more commonly observed in the cerebellum clearly speaks in favor of a common origin of both from the same cell line. We are inclined to agree with others23 that the subependymal glia is the common cell line that gives origin to the pilocytic astrocytoma both below and above the tentorium, whether occurring on one side or in the midline. This view is supported by considering the close anatomical relationship of these tumors with the ventricular wall that we and others20 have observed in every case. This proposition has recently received further support from the findings of Kuhlen- dahl, et al.,10 who stressed this peculiarity of subependymal glia, constituting the so-called “circumventricular organ” and normally presenting a fusiform shape (tanycytes).7 They also found that normal tanycytes share some microscopic features with cells of both third ventricle and cerebellar tumors.10 In appreciation of the World Health Organization efforts aimed at promoting an international classification of tumors of the nervous system, we have adopted the term “pilocytic astrocytoma” for the tumors that form the subject of this study.

<table>
<thead>
<tr>
<th>Factors</th>
<th>Outcome</th>
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<tr>
<td></td>
<td>Good</td>
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<tr>
<td>total excision</td>
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<tr>
<td>with cyst wall</td>
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<td>1</td>
</tr>
<tr>
<td>without cyst wall</td>
<td>14</td>
<td>1</td>
</tr>
<tr>
<td>partial excision</td>
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<td>2</td>
</tr>
<tr>
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<tr>
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<tr>
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<tr>
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<tr>
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<tr>
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<tr>
<td>11–20 yrs</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>&gt; 20 yrs</td>
<td>10</td>
<td>1</td>
</tr>
<tr>
<td>total cases</td>
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References

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Manuscript received October 8, 1984.

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