Ependymomas: Clinical and Pathological Aspects

ERNEST C. FOKES, JR., M.D., AND KENNETH M. EARLE, M.D.
Division of Neurological Surgery, Medical College of Georgia, Augusta, Georgia, and Branch of Neuropathology, Armed Forces Institute of Pathology, Washington, D.C.

The propriety of grading ependymomas by a system based upon histological anaplasia, and the effectiveness of irradiation as an adjunctive measure in treatment, are two of the more controversial topics related to this tumor.

Utilizing material from the Armed Forces Institute of Pathology (AFIP) supplemented by cases from the Medical College of Georgia (MCG), we will discuss certain aspects of the classification, pathology, diagnosis, and therapy of ependymomas.

Source of Material

One hundred and eighty cases were available for study: 167 from the AFIP; 13 from the MCG. The AFIP material consisted of histological slides and in many cases gross material from which additional slides could be made along with a clinical summary. The patient population from which this material was taken consisted primarily of military personnel and their dependents, veterans, and occasional civilians where slides had been sent to AFIP for consultation. Talmadge Memorial Hospital at the MCG, on the other hand, is a general referral hospital for the State of Georgia.

Cases Excluded. Of the 180 cases, 47 were excluded for one or more of the following reasons:
1. Inadequate pathological material to permit accurate diagnosis.
2. Disagreement with the original diagnosis. Most of these cases were seen before 1940 and originally classified as "spongioblastoma ependymal," they are now considered "glioblastoma multiforme" and so have been omitted. All cases in which the degree of anaplasia was such that the basic ependymal origins were not recognizable were also excluded.
3. A few cases in the series had been classified as "subependymal glioma,"

Received for publication April 22, 1968.

"subependymal spongioblastoma," or "subependymal spongioneuroblastoma," after Kuhlenbeck, 6, 7, 11 He, of course, described this neoplasm as consisting primarily of spongioblastic elements, with only occasional tubule or rosette formations as evidence of any ependymal component. We have not included these tumors in the present series for we feel that one of the problems in the past has been the tendency of some to call a tumor an ependymoma in a supratentorial location with relatively little support, such as the presence of a single perivascular pseudorosette. Such tumors are probably glioblastomas from the outset and simply happen to imitate the ependymoma pattern in a few areas. The converse is probably also true, that in some cases where an ependymoma pattern is present it has gone unrecognized. This is well illustrated in the proceedings of the Cancer Seminar on Intracranial Tumors; 5 histopathologic diagnoses submitted by mail on two cases of ependymoma included malignant choroid plexus tumor, chromophobe adenoma, angiosarcoma, medulloblastoma, and 20 "others." Again, tumors that did not primarily show the patterns described below were excluded.

4. Scanty or no clinical data available.

Cases Included. One hundred and thirty-three cases remained for this study. Because clinical information was not available for all of these 133 cases, however, small differences in totals will be noted in the accompanying statistics.

Tumor Location

The most frequent site of the tumors in this series as in others was the fourth ventricle. 3, 6 The location of the tumor was known in 124 cases. Of this number, 86 (69%) were in the posterior fossa; 32 cases (26%)
TABLE 1

Tumors filling the ventricular system

<table>
<thead>
<tr>
<th>Patient's Age (yrs)</th>
<th>Tumor Type</th>
<th>Findings</th>
<th>Duration of Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>20</td>
<td>mixed</td>
<td>filled 3rd ventricle; extended into rt. lat. vent.; filled if. lat. vent. to temporal horn</td>
<td>Broad-based gait for 6 mos, with tendency to fall to rt., headaches</td>
</tr>
<tr>
<td>41</td>
<td>mixed</td>
<td>filling to tips of both temporal horns and all of 3rd ventricle</td>
<td>Dizziness, seizures, incontinence, 1f. side weakness; 6 mos duration</td>
</tr>
<tr>
<td>44</td>
<td>mixed</td>
<td>partially filled 3rd and lat. vent. extending into aqueduct; cerebellar implant (5 cm)</td>
<td>Weakness in legs, diplopia, dizziness dysphasia, ataxia, but not laterализing; 10 yrs duration</td>
</tr>
<tr>
<td>28</td>
<td>subependymoma</td>
<td>filled lateral and 3rd ventricle</td>
<td>Frontal headaches, episodic loss of coordination; 5 yrs duration. Nausea and vomiting; 5 months duration. Severe rt. side headache before admission</td>
</tr>
</tbody>
</table>

were supratentorial; and 6 cases (5%) arose from the spinal cord or cauda equina.

In most cases of ependymoma of the fourth ventricle, the point of origin was probably the floor, occasionally the cerebellopontine angle. At surgery, however, the exact point from which the tumor arose could not always be determined due to the size of the lesion, its impingement upon the cerebellum, its spread out from the lateral recesses, and extension down over the cervical cord. The surgeon may have the impression that the tumor readily shells away from all adjacent structures.

In 19 of 32 supratentorial tumors, proximity to the ependymal lining of the ventricle and filling or partial filling of the ventricular system were noted either at operation or postmortem examination. Four cases (Table 1) are particularly noteworthy examples. In these, the tumor arose from the septum pellucidum or the ventricular system; one case also had a "tongue" of tumor protruding into the aqueduct with a 5 cm tumor implant in the cerebellum. Three of these tumors were mixed ependymomas and the fourth a subependymoma. Ringertz and Reymond have previously pointed out that relation to the ventricles is infrequent in supratentorial tumors. In the present series, however, this relationship was observed in slightly more than one-half of the cases.

Five tumors were found in the cauda equina and one in the dorsal spinal cord.

Age of Patients

The ages of 113 patients varied from 4 weeks to 64 years (Table 2). The mean age of patients with supratentorial lesions was 27.8 years (30 patients), and that of patients with infratentorial lesions was 18.7 years (83 patients). This is in contrast to Ringertz and Reymond's data which showed no age difference in patients with the two tumors, although there was a predominance of infratentorial tumors in the first decade, as in our series. As these authors pointed out,