Intracranial Ependymomas: Factors Influencing Prognosis*

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Our purpose in this paper is to analyze the case histories of a large number of patients with intracranial ependymomas in order to determine which clinical, pathological and therapeutic factors influence prognosis. Some of the general data on this group of patients have been reported elsewhere.10

Review of the Literature

Among large series of intracranial gliomas, the incidence of ependymomas has varied from approximately 2 per cent2 to 8 per cent.21 The average age of 126 patients with intracranial ependymomas reported by Mabon et al.13 was 23.4 years; thus emphasizing their predominance in early life. Indeed, the supratentorial ependymoma is considered by some to be the most common cerebral hemispheric glioma of children.17,24

Ependymomas usually grow slowly and, as with other relatively benign neoplasms, the prognosis is in a large part dependent upon the location of the tumor. In various series approximately 60 per cent of intracranial ependymomas have been infratentorial in location.5,8,17 Most of the tumors arise in the midline in the 4th ventricle, usually from the floor, but sometimes from the roof. They often fill the 4th ventricle and frequently project extraventricularly into the cisterna magna. Extension about the upper segments of the cervical cord is relatively common, while spread into the cerebellopontine angle, into the central canal of the spinal cord and into and through the aqueduct of Sylvius occurs less frequently. Ringertz and Reymond17 stated that only rarely do these tumors extend predominantly into the cerebellar vermis or hemispheres. Grossly these tumors usually are well defined, soft and somewhat lobulated.

Supratentorial ependymomas were found most commonly in the parietal lobe in one investigation2 but other reports have failed to disclose any characteristic location.19 The growth of these tumors often is by expansion toward the centrum semiovale rather than into the ventricular cavity.2 Ringertz and Reymond17 found that more than half of the supratentorial tumors in their series were unrelated visibly to the ventricles. Unlike infratentorial ependymomas, those in the cerebral hemispheres often are partly or extensively cystic.

Three histological forms of ependymoma—cellular, epithelial and papillary—have been defined by some authors.7 Although one architectural type may predominate, most frequently the cellular variety, many of these tumors contain representations of more than one type. In view of this, and because of the failure of other investigators18,17 and ourselves to find any correlation between the histological varieties of ependymomas and their biological behavior, we have not classified our cases in this fashion.

There is controversy as to whether papilloma of the choroid plexus is a fourth type of ependymoma or a separate tumor. We recognize papillomas of the choroid plexus as neoplasms distinct from ependymomas; hence they are not included in this report.
TABLE 1
Intracranial ependymomas (70 patients)

<table>
<thead>
<tr>
<th>Age (Years)</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Under 15</td>
<td>38</td>
</tr>
<tr>
<td>15-25</td>
<td>12</td>
</tr>
<tr>
<td>25-35</td>
<td>11</td>
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<tr>
<td>35-45</td>
<td>6</td>
</tr>
<tr>
<td>45-55</td>
<td>1</td>
</tr>
<tr>
<td>Over 55</td>
<td>2</td>
</tr>
</tbody>
</table>

**Material**

A review of the records of surgical neuropathology of the Neurological Institute from 1943 to 1960 yielded 65 cases of intracranial ependymomas. Five additional patients seen between 1930 and 1943 were brought to our attention by virtue of autopsy records or recurrence of tumor in recent years. Pathological material was available for review in 63 cases; in the 7 remaining the original pathological report was used. Eighteen patients had supratentorial, and 52 infratentorial tumors. One was lost to follow-up immediately upon discharge from the hospital. The remaining 69 were followed either to the present time or to the patient's death. All were followed for more than 3 years. Fifty-nine patients were available for a follow-up of longer than 5 years. Of these, 45 had infratentorial tumors while 14 had supratentorial tumors. Forty-six were followed for 10 years, and 12 for more than 20 years.

**Age**

The average age of all patients when first seen was 16.4 years, with a range of 4 months to 66 years. Patients with supratentorial tumors had an average age of 18.8 years, while the average age of those with infratentorial tumors was 15.4 years. Table 1 shows the distribution of ages of these patients. Customarily, 15 years of age is taken as the dividing line between children and adults; thus 54 per cent of the ependymomas occurred in the pediatric age group.

**Sex**

While the total group of tumors was almost equally divided between males and females, 36 and 34 respectively, 12 of the 18 supratentorial tumors were present in females; 30 of the 52 infratentorial tumors occurred in males.

**Symptoms**

The average duration of symptoms associated with supratentorial ependymomas prior to diagnosis was 7 months with a range of 2 weeks to 8 years. Symptoms were dependent primarily upon the location of the lesion and usually consisted of headache, seizures and visual disturbance. Patients with infratentorial tumors presented most frequently symptoms of increased intracranial pressure such as vomiting, headache or decreased visual acuity. However, nearly one-fifth had complaints of either vertigo or loss of balance. The average duration of symptoms in the infratentorial group was 9 months with a range of 2 weeks to 2 years. Of interest is the fact that patients with infratentorial tumors that were found to be grossly invasive at operation had symptoms for an average of 5.4 months, whereas patients with tumors that were not grossly invasive had an average duration of symptoms of 11.0 months.

**Locus and Growth**

Twelve supratentorial tumors were solid, while 6 were cystic. The neoplastic masses varied in size from 3 cm. to 10 cm. There did not seem to be any favored ventricular site of origin, except for a slight predilection for the 3rd ventricle (Fig. 1). Ependymomas of