Multiple intracranial meningiomas

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The authors report 14 cases of multiple intracranial meningiomas representing 1.1% of all meningiomas operated on at their hospital in the past 35 years. Differentiation of multiple meningiomas, especially from meningiomatosis, must be strict. Since the introduction of computerized tomography scanning, the frequency of these cases has risen from 0.58% to 4.5% in the authors’ meningioma series. Despite the multiplicity of sites, multiple meningiomas do not differ in prognosis from benign solitary meningiomas.

KEY WORDS: multiple meningiomas • brain neoplasm • meningioma • computerized tomography

The neurosurgical frequency of multiple intracranial meningiomas is low, although it has risen since the introduction of computerized tomography (CT). In 1938, Cushing and Eisenhardt defined this entity as “something more than one meningioma and less than a diffusion of them.” Even now, however, cases reported as multiple meningiomas include a variety of conditions from which these lesions must be differentiated. These include meningioma recurrences, multiple meningiomas associated with neurofibromatosis, meningiomatosis, hereditary meningiomas, and meningiomas in patients with a history of radiotherapy. This study reports our experience with the treatment of multiple meningiomas and attempts to precisely define the disease entity.

Clinical Material and Methods

Between 1951 and 1986, 1308 histologically verified cases of intracranial meningiomas were operated on at our institution. Review of the patients' records, the operative notes, and the radiological material yielded 14 cases of multiple meningiomas (about 1% of the total). Before the advent of CT, the frequency was 0.58% and since then it is 4.5%. This study does not include meningioma cases in which more than three tumors were adjacent in one region, cases associated with neurofibromatosis, hereditary cases, cases with a history of radiotherapy to the scalp, or suspected recurrences.

The patients were followed for periods ranging from 1 to 16 years with a mean of 6.7 years. Of the 14 patients, 13 (92.8%) were women. Mean patient age on admission was 50.4 years (range 37 to 65 years).

Results

Table 1 summarizes the data on our 14 cases. In 12 patients all of the tumors were found at the first admission. In the other two the diagnosis was not established until years after removal of the first meningioma (10 years in Case 2 and 7 years in Case 4). Both of these patients were evaluated in the pre-CT era. In only one patient (Case 13) was the diagnosis incidental, due to radiological investigations for head injury. In the others the diagnosis arose from neurological signs and symptoms.

In eight cases (57%) the meningiomas affected one hemisphere. Of the total 33 tumors, 19 (57.6%) were in the right hemisphere, 10 (30.3%) were in the left, and four (12.1%) were on the midline. In 10 (71.4%) of the 14 cases only two tumors were found; one patient had four at different sites (Table 1). Table 2 gives the distribution of the tumors: 76% were in the convexity or parasagittal areas.

All 14 patients received surgical treatment. In 10 cases all of the radiologically proven meningiomas were removed in one operation, whereas in three cases more than one operation was required. In two cases one tumor was not removed: in one patient (Case 2) the
TABLE 1
Clinical findings in 14 cases of multiple meningiomas

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>No. &amp; Size of Tumors</th>
<th>Location of Tumors</th>
<th>Histological Findings</th>
<th>Year of Surgery</th>
<th>Diagnostic Radiological Studies†</th>
<th>Follow-Up Period (yrs)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>55, F</td>
<td>1 large</td>
<td>rt parasagittal</td>
<td>endotheliomatous</td>
<td>1960</td>
<td>angiography</td>
<td>13</td>
</tr>
<tr>
<td>2</td>
<td>44, F</td>
<td>1 small</td>
<td>rt parietal</td>
<td>endotheliomatous</td>
<td>1960</td>
<td>angiography</td>
<td>16</td>
</tr>
<tr>
<td>3</td>
<td>49, F</td>
<td>1 large</td>
<td>lt parietal</td>
<td>fibromatous-endotheliomatous</td>
<td>1971</td>
<td>angiography, CT</td>
<td>14</td>
</tr>
<tr>
<td>4</td>
<td>45, F</td>
<td>1 small</td>
<td>rt occipital</td>
<td>fibromatous-endotheliomatous</td>
<td>1971</td>
<td>angiography, CT</td>
<td>14</td>
</tr>
<tr>
<td>5</td>
<td>29, F</td>
<td>1 large</td>
<td>rt parasagittal</td>
<td>fibromatous</td>
<td>1979</td>
<td>angiography, CT</td>
<td>7</td>
</tr>
<tr>
<td>6</td>
<td>59, M</td>
<td>1 large</td>
<td>rt frontal</td>
<td>endotheliomatous</td>
<td>1980</td>
<td>angiography, CT</td>
<td>6</td>
</tr>
<tr>
<td>7</td>
<td>65, F</td>
<td>1 small</td>
<td>medial falx</td>
<td>endotheliomatous</td>
<td>1980</td>
<td>angiography, CT</td>
<td>4</td>
</tr>
<tr>
<td>8</td>
<td>53, F</td>
<td>1 large</td>
<td>rt frontal</td>
<td>fibromatous</td>
<td>1982</td>
<td>angiography, CT</td>
<td>4</td>
</tr>
<tr>
<td>9</td>
<td>57, F</td>
<td>3 small</td>
<td>rt frontal</td>
<td>fibromatous</td>
<td>1982</td>
<td>angiography, CT</td>
<td>3</td>
</tr>
<tr>
<td>10</td>
<td>54, F</td>
<td>1 small</td>
<td>rt parietal</td>
<td>endotheliomatous</td>
<td>1983</td>
<td>angiography, CT</td>
<td>3</td>
</tr>
<tr>
<td>11</td>
<td>37, F</td>
<td>1 large</td>
<td>rt occipital</td>
<td>fibromatous</td>
<td>1983</td>
<td>angiography, CT</td>
<td>3</td>
</tr>
<tr>
<td>12</td>
<td>57, F</td>
<td>1 small</td>
<td>medial tuberculum sellae</td>
<td>endotheliomatous</td>
<td>1985</td>
<td>angiography, CT</td>
<td>1</td>
</tr>
<tr>
<td>13</td>
<td>57, F</td>
<td>1 large</td>
<td>rt parietal</td>
<td>endotheliomatous-psammomatous</td>
<td>1985</td>
<td>angiography, CT</td>
<td>1</td>
</tr>
<tr>
<td>14</td>
<td>44, F</td>
<td>1 small</td>
<td>rt parietal</td>
<td>endotheliomatous-psammomatous</td>
<td>1986</td>
<td>angiography, CT, MRI</td>
<td>1</td>
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<tr>
<td>15</td>
<td>57, F</td>
<td>1 small</td>
<td>rt parietal</td>
<td>endotheliomatous</td>
<td>1986</td>
<td>angiography, CT</td>
<td>1</td>
</tr>
<tr>
<td>16</td>
<td>44, F</td>
<td>2 large</td>
<td>rt parietal</td>
<td>fibromatous</td>
<td>1986</td>
<td>angiography, CT</td>
<td>1</td>
</tr>
</tbody>
</table>

* Large tumors were ≥ 2 cm; small tumors were < 2 cm.
† CT = computerized tomography; MRI = magnetic resonance imaging.

Histologically, there were no differences between the multiple and benign solitary meningiomas. Of the 31 tumors removed, 12 (38.7%) were endotheliomatous, 12 were fibroblastic (see Table 1), and the other seven were mixed. There have been no recurrences. At long-term follow-up evaluation four patients (28.6%) had residual neurological deficits; however, this was not severe enough to affect the quality of life, since all the patients returned to their jobs.

Discussion

In 1889, Anfimow and Blumenau⁵ reported the first case of multiple meningiomas, and other single case reports followed. However, it was Cushing and Eisenhardt⁶ who first defined them as a nosological entity, clearly distinguishing the condition from diffuse meningiomatosis, neurofibromatosis, and recurrences.

Meningiomatosis is a diffusion of small tumor nodules, is often associated with von Recklinghausen’s disease, and as a rule has a highly proliferative potential resulting in a poorer prognosis.¹²,¹³,¹⁷,²¹,²² Although there are transitional forms between multiple meningiomas and meningiomatosis,⁶,¹⁶,²⁴ every effort must be made

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to differentiate the two conditions. This is why we “arbitrarily” excluded cases with more than three confluent tumors in a single region of the brain. Some authors suggest that multiple meningiomas are a forme fruste of von Recklinghausen’s disease.11,23 We are convinced that this theory is untenable in the absence of cutaneous stigmata, other associated tumors (especially eighth cranial nerve neuroma),10 and objective genetic or biochemical evidence.15 We also believe that the growth of a new tumor, not just at the site of a previous operation but also in its immediate neighborhood, should be considered a recurrence and therefore be excluded from the category.

Most of the published series put the frequency of multiple meningiomas at between 1% and 3% of all meningiomas.1,8,12,23 Nahser, et al.,29 reviewed the world literature and found a mean frequency of 2.4%. This frequency decreases if we consider the cases in which several meningiomas in separate compartments of the neuraxis were associated. Indeed, these are mentioned in the literature only as case reports.2,5,9,14,27

Since the introduction of CT scanning, frequencies of multiple meningiomas ranging between 5.9% and 10.5% have been reported.11,18,20 The highest incidence (16%) is that reported by Wood, et al.,26 before CT became available; theirs was a postmortem series comprising an elderly population with no neurological symptoms, so it is unlikely to be paralleled in ordinary clinical practice. It is doubtful that objective conclusions on the clinical and biological features of multiple meningiomas will emerge from a comparison of these series because the material often includes cases of probable meningiomatosis,1,10,18,23,24 recurrences,17 neurofibromatosis,12,23 hereditary cases,20 or cases without histological verification.11,15 Such cases were excluded from our series. Our overall frequency rate is 1.1%, but this is the mean of 0.58% before CT and 4.5% since it became available, a difference in incidence that tallies with the data reported by Sheehy and Crockard.23 Routine CT scanning undoubtedly enabled us to detect tumors that would previously have been missed by conventional radiology, especially small and/or asymptomatic tumors. We agree with Lusins and Nakagawa18 that CT findings confront us with a new dilemma: that is, whether or not to treat asymptomatic lesions.

Women accounted for 92.8% of our patients, a percentage similar to that found by others.12,18,23,24 Hormonal sensitivity might explain this particular proliferative condition, for it is known that meningiomatous tissue may contain hormonal receptors; however, to date it has not been possible to reach definite conclusions and hence to offer suggestions for treatment.22

There were no histological differences between multiple meningiomas and benign solitary meningiomas in our series, in accordance with the findings of most other workers.3,10–12,20,23 Our clinical follow-up data revealed a benign course, as was true in the series of Sheehy and Crockard.23 This contrasts to the findings of Andrioli, et al.,3 who reported a poor prognosis in four of their five patients, but these were probably cases of meningiomatosis.

Three theories have been advanced to explain the etiology and pathogenesis of multiple intracranial meningiomas: spontaneous or surgical blood-borne spread, spontaneous or surgical spread via the cerebrospinal fluid, and multicentricity of dural foci.3,11,17,18 Spontaneous dissemination via blood or cerebrospinal fluid is unlikely because the tumor is histologically benign. Seeding after surgery has been suggested in some cases of multiple meningiomas,17,23 but in our series 12 patients (85.7%) had multiple meningiomas on first presentation; in the two cases of “successive” multiple meningiomas the first diagnosis and operation antedated CT and quite possibly there were already multiple tumors. Most authors find the multicentricity theory acceptable, and this theory has gained credibility since the demonstration by Borovich and Doron17 of multiple minute dural foci around meningiomas operated on as “solitary.” The ratio of multiple meningiomas is low, probably because only a small minority of these foci develop into tumors and these often remain asymptomatic and undiagnosed.

References


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