Location, sidedness, and sex distribution of intracranial arachnoid cysts in a population-based sample

Clinical article

CHRISTIAN A. HELLAND, M.D., PH.D., MORTEN LUND-JOHANSEN, M.D., PH.D., AND KNU特 WESTER, M.D., PH.D.

Department of Neurosurgery, Haukeland University Hospital; and Section on Neurosurgery, Department of Surgical Sciences, University of Bergen, Norway

Object. The aim of this study was to examine the distribution of intracranial arachnoid cysts in a large and unselected patient population with special emphasis on sidedness and sex distribution.

Methods. In total, 299 patients with 305 arachnoid cysts were studied. These patients were consecutively referred to our department during a 20-year period from a well-defined geographical area with a stable population.

Results. There was a strong predilection (198 patients [66.2%]) for intracranial arachnoid cysts in the temporal fossa. Forty-two patients had cysts overlying the frontal convexity, 36 had cysts in the posterior fossa, and 23 patients had cysts in other, different locations. Of 269 cysts with clearly unilateral distribution, 163 were located on the left side and 106 on the right side. This difference resulted from the marked preponderance of temporal fossa cysts on the left side (left-to-right ratio 2.5:1; p < 0.0001 [adjusted < 0.0005]). For cysts in the cerebellopontine angle (CPA), there was preponderance on the right side (p = 0.001 [adjusted = 0.005]). Significantly more males than females had cysts in the temporal fossa (p = 0.002 [adjusted = 0.004]), whereas in the CPA a significant female preponderance was found (p = 0.016 [adjusted = 0.032]). For all other cyst locations, there was no difference between the 2 sexes.

Conclusions. Arachnoid cysts have a strong predilection for the temporal fossa. There is a sex dependency for some intracranial locations of arachnoid cysts, with temporal cysts occurring more frequently in men, and CPA cysts found more frequently in women. Furthermore, there is a strong location-related sidedness for arachnoid cysts, independent of patient sex. These findings and reports from the literature suggest a possible genetic component in the development of some arachnoid cysts. (DOI: 10.3171/2009.11.JNS081663)

Key Words • intracranial arachnoid cyst • temporal fossa • distribution • sex • cerebellopontine angle • sidedness

Abbreviations used in this paper: CPA = cerebellopontine angle; ICD = International Classification of Diseases.
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of approximately 1 million people (as of 2009). During the study period, a few patients were referred to our department from other health regions for surgical cyst decompression, but patients from our own region with cysts were not referred to other regions.

Study Design and Data Extraction

The present study is a population-based study, including all patients operated on as well as those without operations, who were referred to our department for an arachnoid cyst during the period from 1987 to 2006. Some of these patients have been presented in previous publications from our department, including 1 article addressing the same questions as this study, but using a much smaller patient sample. The patients were identified from the hospital’s computer data bank of diagnosis and treatment, with registration of every patient in the period with the diagnostic codes 348.0 (ICD 9) or G93.0 (ICD 10), the codes assigned to patients with arachnoid cysts in our hospital during the study period. These codes also include other intracranial cysts, but only patients with arachnoid cysts were included in the study. All patients were examined using either cerebral CT, MR imaging, or both. The following data were extracted from the complete medical records of the patients: demographic data, location of the cyst, Galassi type for the temporal cysts, and side.

Patient Sample

A total of 299 patients (183 males and 116 females) were included. Ten patients underwent neuroimaging without longstanding symptoms. Two patients presented with acute/subacute subdural hematomas. Six patients received an arachnoid cyst diagnosis due to a CT scan after a cerebral concussion. The arachnoid cysts in only 2 patients were truly discovered incidentally. One patient was the identical twin sister of a patient who underwent an operation for a cyst in the CPA. This patient was offered MR imaging for a scientific purpose. The last of the 10 patients was included in a functional MR imaging project as a control patient and was found to have cognitive disturbances (especially memory problems and forgetfulness). The mean age of the patients in the study was 34.4 years (median 34.6 years, range 1 month to 78 years). The mean age of the patients in the study was 34.4 years (median 34.6 years, range 1 month to 78 years). The mean age of the patients in the study was 34.4 years (median 34.6 years, range 1 month to 78 years). The mean age of the patients in the study was 34.4 years (median 34.6 years, range 1 month to 78 years). The mean age of the patients in the study was 34.4 years (median 34.6 years, range 1 month to 78 years). The mean age of the patients in the study was 34.4 years (median 34.6 years, range 1 month to 78 years). The mean age of the patients in the study was 34.4 years (median 34.6 years, range 1 month to 78 years).

Statistical Analysis

The statistical analyses were performed using SPSS version 15.0 for Windows (SPSS Inc.). An α level of 0.05 was used for all statistical calculations. Contingency tables were analyzed using the Fisher exact test or chi-square statistics. When multiple comparisons were performed in the same group, probability values were corrected using Bonferroni correction (referred to as “adjusted” values).

Results

Intracranial Location

In the 299 patients, a total of 305 intracranial cysts were identified (6 patients had bitemporal cysts). The overall distribution of the cysts is summarized in Table 1. Of the 299 patients, 198 (66.2%) had cysts located in the temporal fossa, 42 (14.0%) had cysts overlying the frontal convexity, and 36 (12.0%) had cysts in the posterior fossa. Sixteen (5.4%) of the posterior fossa cysts were located in the CPA. Twenty-three patients (7.7%) had cysts in other locations within the neurocranium.

Sidedness

Thirty patients had cysts that were either bilateral or were located in or around the midline. These patients were therefore excluded from the sidedness analysis. Thus, a total of 269 patients each had 1 cyst that was clearly located either to the right or to the left of the anatomical midline of the brain (Fig. 1). One hundred sixty-three of these cysts were located on the left side and 106 were on the right (Table 2). This difference was statistically significant (p < 0.0001 [adjusted < 0.0005]). This left-sided preponderance is attributed solely to the distribution of cysts in the temporal fossa with a ratio of 2.5:1 (137 left, 54 right; p < 0.0001 [adjusted < 0.005]). For cysts in the frontal convexity, there was no difference in sidedness (16 left, 26 right; p = 0.124).

For posterior fossa cysts, there was a statistically significant preponderance for the right side (6 left, 19 right; p = 0.006 [adjusted = 0.03]). This difference was attributed solely to cysts located in the CPA, with 2 on the left side and 14 on the right side (p = 0.001 [adjusted = 0.005]). For cysts in other locations, there was no difference in sidedness (4 left, 7 right; p = 0.391).

Sex Distribution

There were significantly more males than females (183 vs 116, respectively; p < 0.0001) in the patient sample (Fig. 2). This difference was attributed solely to the preponderance of cysts in the temporal fossa in males (134 males/64 females; male/female ratio = 2.1:1; p = 0.002 [adjusted = 0.004]). For the other main locations (frontal convexity, posterior fossa, and other locations) there was no significant difference in sex distribution. However, for the CPA sublocation there was a significant female preponderance (5 males/11 females; p = 0.016 [adjusted = 0.032]).

Sex and Sidedness

For arachnoid cysts in the temporal fossa, both male (p < 0.0001 [adjusted < 0.0002]) and female (p = 0.001 [adjusted = 0.002]) patients demonstrated a significantly increased number of left-sided cysts (Table 2). For cysts overlying the frontal convexity, no difference in sidedness could be detected for either sex. As noted above, there was a female predominance for cysts in the CPA, with a significant tendency for right-sided localization (p = 0.026).

Discussion

Most of the previously published series on intracranial arachnoid cysts have some limitations, mainly...
because of selection bias. These reports are based on either a subgroup of patients with a distinct intracranial localization or age distribution, or a relatively small number of patients, or the inclusion of patients from before the computerized imaging era. Furthermore, in most reports there is no information on whether the study is population based. It is reasonable to assume that the intracranial distribution of arachnoid cysts previously reported does not fully reflect the true figures in the population, because cases reported before or in the early phases of the computerized imaging era were subject to considerable selection bias.

Most likely, cysts located in noneloquent areas, as well as smaller cysts, were not detected when neuroimaging was less available and the methods were less sensitive, and when patients as well as doctors paid less attention to nonprogressive symptoms such as dizziness and headache. Over the last several decades, the availability of neuroimaging technology has increased dramatically in Norway, and is still increasing. Thus, one may assume that the threshold for detecting cysts has been lowered correspondingly and will be even more so in the future.

Almost all of our patients were recruited from a well-defined population within a restricted geographical area. Thus, the study may be described as “population based.” However, whether the present results reflect the true distribution of arachnoid cysts in a population is highly uncertain, because the estimated prevalence of arachnoid cysts in a population has been estimated to be as high as 1%, 20,47,79 If this is true, our patient population represents only a small fraction of the estimated 9000 persons with arachnoid cysts in our region. Thus, our figures do not necessarily reflect the true distribution of arachnoid cysts in the overall population, but are probably a better estimate than reviews including non–population-based studies and historical reports, given the organization of health care in Norway, and the fact that all our patients were referred to our department after the introduction of CT scanning and MR imaging.

Two-thirds of the cysts were located in the temporal fossa. This observation is in accordance with previous publications and may be caused, at least in part, by the pronounced curving of the fetal brain in the formation of the sylvian fissure. There are also some interesting findings in our data that have, to our knowledge, not been previously addressed. The skewed sidedness of cysts in the temporal fossa is well known, as is the sex distribution with more temporal cysts occurring in male patients. We have addressed the issue of cyst distribution in 2 previous reports. The first report is a review based on the literature published after the introduction of the CT scanner. The second publication is a report of a population of 126 patients, in which we found that the left sidedness of intracranial arachnoid cysts was present only in males, and that females had an even distribution of temporal cysts between the 2 sides. However, in the larger population studied in the present article, we cannot confirm this, as we found the same left-sidedness distribution for temporal cysts in both sexes. This finding of a sex-independent, location-dependent sidedness for cysts in the temporal fossa indicates that if a genetic mechanism is involved in the formation of temporal cysts, it is probably not genes located on the sex chromosomes, as the previous results otherwise might imply.

Cysts in the CPA appear to constitute a separate entity, with the reverse distribution pattern from that of temporal cysts (female overrepresentation and preponderance for the right side). From our findings, it appears that temporal and CPA arachnoid cysts have a sex-independent but location-dependent sidedness, and that they therefore possibly constitute separate entities. Mirror-image cysts in the CPA in monozygotic twins have been reported.

### TABLE 1: Intracranial distribution of 305 arachnoid cysts in 299 patients

<table>
<thead>
<tr>
<th>Location</th>
<th>No. of Cysts (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>temporal fossa*</td>
<td>204 (66.9)</td>
</tr>
<tr>
<td>frontal convexity</td>
<td>42 (13.8)</td>
</tr>
<tr>
<td>posterior fossa</td>
<td>36 (11.8)</td>
</tr>
<tr>
<td>CPA</td>
<td>16 (5.2)</td>
</tr>
<tr>
<td>other intracranial locations</td>
<td>23 (7.5)</td>
</tr>
</tbody>
</table>

* Six patients had bitemporal cysts.

### TABLE 2: Sex and sidedness in different locations

<table>
<thead>
<tr>
<th>Sex</th>
<th>Temporal Fossa</th>
<th>Frontal Convexity</th>
<th>Posterior Fossa</th>
<th>CPA</th>
<th>Other</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>male</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>lt side</td>
<td>94</td>
<td>8</td>
<td>3</td>
<td>0</td>
<td>1</td>
<td>106</td>
</tr>
<tr>
<td>rt side</td>
<td>36</td>
<td>14</td>
<td>4</td>
<td>5</td>
<td>2</td>
<td>61</td>
</tr>
<tr>
<td>total</td>
<td>130</td>
<td>22</td>
<td>7</td>
<td>5</td>
<td>3</td>
<td>167</td>
</tr>
<tr>
<td>female</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>lt side</td>
<td>43</td>
<td>8</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>57</td>
</tr>
<tr>
<td>rt side</td>
<td>18</td>
<td>12</td>
<td>1</td>
<td>9</td>
<td>5</td>
<td>45</td>
</tr>
<tr>
<td>total</td>
<td>61</td>
<td>20</td>
<td>2</td>
<td>11</td>
<td>8</td>
<td>102</td>
</tr>
</tbody>
</table>
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Furthermore, this location has been described to co-occur in a variant form of hereditary spastic paraplegia associated with a mutation in the spastic paraplegia Type 4 (SPG4) gene.38

Cysts in the temporal fossa have been reported to have a familial occurrence, in some cases co-occurring with other genetically determined diseases.30,42,44,61 Arachnoid cysts have also been reported to co-occur with other diseases linked to genetic aberrations, such as autosomal dominant polycystic kidney disease.2,5,7,88

The sex-independent but location-dependent sidedness, the familial occurrence, and the linkage with other genetically determined diseases support the hypothesis of an underlying genetic mechanism in the development of at least some of these cysts. This is an observational finding that should be investigated further with modern gene expression techniques to bring to light the mechanisms of cystogenesis in intracranial arachnoid cysts.

Conclusions

Intracranial arachnoid cysts in the temporal fossa and in the CPA demonstrate a sex-independent sidedness. These findings, in addition to previous data, might indicate a genetic mechanism behind the formation of arachnoid cysts.

Disclosure

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