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http://www.ajnr.org/content/12/4/749
Intracerebral Gangliogliomas in Patients with Partial Complex Seizures: CT and MR Imaging Findings

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The clinical and radiologic findings in 19 patients with partial complex seizures and surgically proved intracerebral gangliogliomas were reviewed to characterize the radiologic features of these lesions. The CT and MR findings were not specific. On CT the gangliogliomas can be hypodense with no enhancement and they often have calcifications. On MR these tumors have a wide variety of signals. In five of our cases the tumor had a high-intensity signal with a cystlike component on proton density- and T2-weighted images. In five cases the lesion had an inhomogeneously intense signal on proton density-weighted images and high signal intensity on T2-weighted images. The tumor had high-intensity signal on both proton density- and T2-weighted images in four patients. Finally, in two cases the MR findings were normal.

We recommend MR as the examination of choice for patients with partial complex seizures because it allows an artifact-free evaluation of the temporal region. However, CT should also be performed in order to recognize calcifications that may be missed on the MR examination.


Gangliogliomas are rare, slow-growing, relatively benign neoplasms consisting of a mixture of adult ganglion cells and glial tumor cells. They are often responsible for a prolonged clinical course, characterized primarily by seizures. To better define the imaging characteristics of these tumors, we reviewed the clinical and radiologic findings in 19 patients presenting with partial complex seizures who were subsequently found to have surgically proved gangliogliomas.

Materials and Methods

From January 1987 to August 1990, 19 patients, 11 men and eight women, 7 to 44 years old (mean age, 21 years), were referred to our institute for intractable partial complex seizures (PCoS). PCoS are one of the features found in temporal lobe epilepsy. They often, but not always, start with motor arrest typically followed by orocutaneous automatism lasting less than 1 min, which in turn is followed by post-ictal confusion and amnesia [1]. These patients received a complete clinical and radiologic work-up and underwent surgery for removal of intracerebral space-occupying lesions histologically proved to be gangliogliomas. EEG findings, skull radiographs, and CT scans were available for all patients. CT examinations were performed with an EMI scanner 1010 and a GE 9800 Quick scanner. In eight cases the CT scans were obtained without contrast enhancement; in six cases the examinations were performed after injection of contrast medium. Five patients had both plain and enhanced CT examinations. Eighteen patients were studied by MR imaging; in one patient (case 4) the examination was stopped because the boy had a seizure. In five cases, the MR examinations were performed with a Philips Gyroscan 0.5-T unit, and T1-weighted, 500/30/2 (TR/TE/excitations), proton density-weighted, and T2-weighted (1500/30,60/2) images were obtained. In 13 cases, a 1.5-T unit was used, and T1-weighted (500/30/2), proton density-weighted, and T2-weighted (2100/30,60/2) images were obtained. Proton density- and T2-weighted images were obtained in axial and coronal views in all cases. T1-weighted images.
were available only in seven cases. None of the MR studies were contrast enhanced, since this study was carried out before the clinical use of gadopentetate dimeglumine in Canada.

Results

Clinical and EEG Findings (Table 1)

The familial and medical histories were unremarkable in 16 patients in our series. One patient (case 5) was born 3 weeks post-term without any evidence of perinatal sequela. Another patient (case 18) was suffering from Rosenthal syndrome caused by deficiency of factor XI. A 33-year-old woman (case 17) developed PCoS after an episode reported as encephalitis at age 13.

All the patients were referred for intractable PCoS. In seven cases, these episodes were associated at an older age with tonic-clonic seizures and secondary generalization. Case 1 presented with PCoS and left focal motor seizures. In one patient (case 5) the PCoS were associated with drop attacks, and in another (case 15) with absences, characterized by short episodes of loss of contact without automatism.

The age of onset of seizure disorders ranged from 3 months to 31 years. The frequency of the attacks, when specified, ranged from four attacks per day to two per year at the time of admission.

All the patients had a prolonged clinical course with an important free interval between the onset of the seizures and the radiologic diagnosis. This time interval ranged from 3 to 26 years (mean, 11½ years).

The EEGs showed epileptiform abnormality in all the cases in our series. In seven patients the EEG reports described an abnormal discharge with a right temporal focus, and in seven cases the focus was found in the left temporal region. In case 7 the abnormal discharge was from both temporal lobes, and in case 2 the abnormality was from the right posterior temporo-occipital area. In only three patients (cases 15, 16, and 17) did the EEG demonstrate an abnormal discharge lateralized to one cerebral hemisphere with no evidence of focal abnormalities.

Radiologic Findings (Table 2)

Skull Radiographs. In no case did the plain films reveal any evidence of raised intracranial pressure or mass effect with displacement of the calcified pineal gland. In four patients florid calcifications were visualized in the middle cranial fossa on the same side of the lesion. In cases 8 and 11, the plain films documented the previous craniotomy.

CT Scans. The CT findings were available in all cases. In 15 patients it was positive: four lesions (cases 1, 4, 15, 16) were hypodense (Figs. 1 and 2); two lesions (cases 7 and 8) were ring-enhancing (Fig. 3), and in six patients (cases 2, 3, 6, 9, 10, 13) the examination demonstrated only calcifications (Fig. 4). Two CT studies (cases 14 and 18) showed asymmetry of the temporal horn (Fig. 5), the larger being on the tumoral side, and in case 12 the CT study demonstrated only a prominent left hippocampus. In three cases (5, 17, 19) the examination was negative, and, finally, in case 11 there was only evidence of the previous surgery.

MR Images. Seventeen MR examinations were completed and available and the tumor was identified in 15 patients.
TABLE 2: Radiologic Findings

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Skull</th>
<th>CT Plain</th>
<th>Enhanced</th>
<th>MR T1 Proton Density</th>
<th>T2 Proton Density</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>7</td>
<td>M</td>
<td>Neg</td>
<td>Hypodense</td>
<td>–</td>
<td>–</td>
<td>[Ts + cystlike]</td>
<td>R temp</td>
</tr>
<tr>
<td>2</td>
<td>11</td>
<td>F</td>
<td>Neg</td>
<td>Calcifications</td>
<td>No enh</td>
<td>–</td>
<td>Inh is + cystlike</td>
<td>–</td>
</tr>
<tr>
<td>3</td>
<td>12</td>
<td>M</td>
<td>Calcifications</td>
<td>No enh</td>
<td>–</td>
<td>–</td>
<td>Inh is + cystlike</td>
<td>L temp</td>
</tr>
<tr>
<td>4</td>
<td>12</td>
<td>M</td>
<td>Neg</td>
<td>Hypodense</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>L temp</td>
</tr>
<tr>
<td>5</td>
<td>14</td>
<td>M</td>
<td>Neg</td>
<td>Calcifications</td>
<td>No enh</td>
<td>–</td>
<td>–</td>
<td>L temp</td>
</tr>
<tr>
<td>6</td>
<td>15</td>
<td>F</td>
<td>Neg</td>
<td>Calcifications</td>
<td>No enh</td>
<td>–</td>
<td>–</td>
<td>L temp</td>
</tr>
<tr>
<td>7</td>
<td>16</td>
<td>F</td>
<td>Neg</td>
<td>Calcifications</td>
<td>Ring enh</td>
<td>Iso is</td>
<td>[Ts + cystlike]</td>
<td>L temp</td>
</tr>
<tr>
<td>8*</td>
<td>17</td>
<td>M</td>
<td>Calcifications</td>
<td>Ring enh + calcification</td>
<td>–</td>
<td>–</td>
<td>[Ts + cystlike]</td>
<td>L temp</td>
</tr>
<tr>
<td>9</td>
<td>17</td>
<td>F</td>
<td>Calcification</td>
<td>Calcification</td>
<td>No enh</td>
<td>Iso is</td>
<td>[Ts + small void sig]</td>
<td>R temp</td>
</tr>
<tr>
<td>10</td>
<td>18</td>
<td>M</td>
<td>Calcifications</td>
<td>Calcifications</td>
<td>–</td>
<td>–</td>
<td>Inh is + void sig</td>
<td>R temp</td>
</tr>
<tr>
<td>11*</td>
<td>21</td>
<td>M</td>
<td>Calcifications</td>
<td>Calcifications</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>L temp</td>
</tr>
<tr>
<td>12</td>
<td>25</td>
<td>M</td>
<td>Neg</td>
<td>–</td>
<td>Prominent hippocampus</td>
<td>Iso is</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>13</td>
<td>25</td>
<td>F</td>
<td>Calcifications</td>
<td>Calcifications</td>
<td>–</td>
<td>–</td>
<td>Inh is + void sig</td>
<td>R temp</td>
</tr>
<tr>
<td>14</td>
<td>27</td>
<td>M</td>
<td>Neg</td>
<td>Asymmetric temporal horn R &gt; L</td>
<td>–</td>
<td>–</td>
<td>Neg</td>
<td>R temp</td>
</tr>
<tr>
<td>15</td>
<td>27</td>
<td>F</td>
<td>Neg</td>
<td>–</td>
<td>Hypodense</td>
<td>Hypo is + cystlike</td>
<td>[Ts + cystlike]</td>
<td>R temp</td>
</tr>
<tr>
<td>16</td>
<td>32</td>
<td>M</td>
<td>–</td>
<td>Hypodense</td>
<td>No enh</td>
<td>–</td>
<td>Inh is + cystlike</td>
<td>–</td>
</tr>
<tr>
<td>17</td>
<td>33</td>
<td>F</td>
<td>Neg</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Neg</td>
<td>L temp</td>
</tr>
<tr>
<td>18</td>
<td>34</td>
<td>M</td>
<td>Neg</td>
<td>Asymmetric temporal horn R &gt; L</td>
<td>–</td>
<td>–</td>
<td>Inh is + cystlike</td>
<td>R temp</td>
</tr>
<tr>
<td>19</td>
<td>44</td>
<td>M</td>
<td>Neg</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Is</td>
<td>R temp</td>
</tr>
</tbody>
</table>

Note.—neg = negative examination, – = examination not done, no enh = no enhancement after injection, inh is = inhomogeneously intense signal, iso is = isointense signal, hypo is = hypointense signal, [Is] = high-intensity signal, [Is] = low-intensity signal, void sig = void signal, R = right, L = left, temp = temporal lobe.

* Previous partial resection in another center.

The gangliogliomas demonstrated a wide variety of signals. In five patients (cases 1, 3, 7, 8, 15) the lesion had a signal similar to CSF on proton density-weighted images and higher than CSF on T2-weighted images (Figs. 1–3). In five cases the tumor had an inhomogeneously intense signal on proton density-weighted images that became hyperintense on the T2-weighted images (cases 5, 10, 13, 16, 18). Finally, the lesion had a high-intensity signal on both proton density- and T2-weighted images in four patients (cases 6, 9, 12, 19).

MR detected calcifications in only two patients (cases 9 and 13) (Fig. 4). T1-weighted images were available in only seven patients: they showed the lesions to be isointense in three cases, hypointense in two cases, and isointense with CSF in one case (Fig. 2).

Discussion

The first description of ganglioglioma was given by Courville in 1930 [2]. The term ganglioglioma clearly defines the structure of these uncommon tumors as a mixture of nerve cells and glial elements. They constitute just 0.4% of all CNS tumors [3, 4]. This percentage has been reported to be as high as 4.3% [5] and 7.6% [6] in studies that factor out the growing fraction. One study [7] reported a prevalence of 4.1% in a consecutive series of brain tumors. Other studies have reported a prevalence of 1.3% [8] and 0.3% [9] in a consecutive series of brain tumors. The prevalence of ganglioglioma in a series of intracranial tumors was 0.9% [10]. The prevalence of ganglioglioma in a series of intracranial tumors was 0.9% [10]. The prevalence of ganglioglioma in a series of intracranial tumors was 0.9% [10]. The prevalence of ganglioglioma in a series of intracranial tumors was 0.9% [10].

In childhood, gangliogliomas can occur in any number of locations in the CNS, and can involve both brain and spinal cord; however, the most common locations in the adult population seem to be the temporal lobe and the floor of the third ventricle [3, 8]. In the present series we included only lesions causing PCoS, and for this reason a preference for the temporal lobe, particularly the hippocampal gyrus, was found.

In childhood, gangliogliomas seem to occur more frequently in the medulla and spinal cord [9] and the differential diagnosis with the more benign gangliocytoma has to be considered. This latter tumor, rarely supratentorial [10, 11] and purely neuronal without a glial component, is an extremely rare entity, accounting for 0.1% of intracranial tumors [12] with a slight male predilection [13], and it cannot be radiologically differentiated from ganglioglioma.

The clinical course suggests that gangliogliomas are slow-growing neoplasms, and a malignant transformation is very uncommon. When it happens it is usually the glial element that is responsible for the change [14]. Metastasis in the CNS...
appears to be exceptional [14]. Some authors have also raised the possibility that gangliogliomas are hamartomatous lesions since they have been found in temporal lobes of patients with a long history of seizures [15] and because collections of heterotopic gray matter similar to ganglioglioma can cause seizures [16]. Nowadays, the literature seems to agree that gangliogliomas are true neoplasms and their removal has to be attempted [5, 17].

The EEG findings usually suggest the focal abnormality that must be confirmed by the imaging findings. Radiologically, gangliogliomas do not have a specific appearance. Calcifications can be visualized on a skull radiograph in 10% of patients [18]. Although the skull radiograph is a superfluous examination, it was performed in all our patients and we observed four cases (21%) with flocular calcifications in the middle cranial fossa.

CT findings also are nonspecific. Gangliogliomas are usually hypodense [19] with calcifications [20]. In our series CT scans were positive in 12 patients (63%); six of them (32%) demonstrated only abnormal calcifications, and the other six showed a wide variety of findings. In no case in our series did CT demonstrate edema or mass effect.

In a small series [21] of four patients studied with MR imaging, two lesions had a cystic appearance while the other two displayed an increased signal on T1-weighted images and a decreased signal on T2-weighted images. The authors did not try to explain these findings. In a larger series of 14 patients reported by Castillo et al. [22] there were four cystic lesions (low signal on T1-weighted images and high signal on T2-weighted images) and 10 solid masses that demonstrated high-intensity signal on T2-weighted images. The authors did not mention whether the cystic nature of the lesions observed on MR was confirmed later at surgery.

In our series, gangliogliomas presented different signals because of their heterogeneous components of glial and neuronal cells associated with calcifications. In five (29%) of the 17 patients who were studied with MR imaging, the gangliogliomas had a cystlike component (Figs. 1 and 2) and...
in five patients (29%) the lesions displayed inhomogeneous signal on proton density-weighted images and high-intensity signal on T2-weighted images. Finally, in four cases (24%) the tumors had high signals on both proton density- and T2-weighted images. In only two patients (12%) was MR able to detect calcifications (Fig. 4). We prefer to use the term cystlike appearance to describe the MR characteristic of a well-defined area with signal intensity similar to CSF on proton density-weighted images and higher than CSF on T2-weighted images. This is because the content of the lesions is not fluid but solid; therefore, it cannot be drained but has to be removed, as was the case in the patients in our series.

MR failed to show the tumor in two (12%) of the 17 cases. Owing to their frequent attacks of seizures and their EEG results, these patients eventually underwent temporal lobectomy; the histologic reports revealed the presence of the
lesion. The CT and MR findings in these patients demonstrated an asymmetry of the temporal horns in one patient (case 14) and were completely normal in the other (case 17).

In conclusion, gangliogliomas are uncommon lesions, but their presence has to be considered in patients with a long history of uncontrollable seizures. Since most of these tumors are located in the middle cranial fossa, MR is the examination of choice because it allows images free from bone artifact. Nevertheless, false-negative diagnoses are possible and we think that CT should also be performed in order to detect calcification, which can be missed on MR.
REFERENCES