Neurocytoma Manifesting as Intraventricular Hemorrhage
—Case Report—

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Abstract

A 43-year-old man presented with a neurocytoma manifesting as severe headache and disturbance of consciousness. Computed tomography revealed intraventricular hemorrhage, and a small mass lesion with calcification on the wall of the left lateral ventricle. The lesion appeared as mixed intensity regions on both T1- and T2-weighted magnetic resonance imaging, and heterogeneous enhancement with gadolinium-diethylenetriaminepenta-acetic acid. Angiography showed the pooling sign near the calcification in the late venous phase. Neurologically, amnestic syndrome was demonstrated in the subacute phase. Gross total removal of the lesion was performed through a transcortical approach. His transient memory disturbance resolved. The histological diagnosis was neurocytoma. Intraventricular hemorrhage is rare as the initial presentation of neurocytoma. Surgery should avoid fornix injury and the risk of permanent memory disturbance.

Key words: neurocytoma, intraventricular hemorrhage, memory disturbance

Introduction

Neurocytomas are rare, accounting for only 0.25% of all central nervous system tumors. The typical location is the anterior half of the lateral ventricle near the foramen of Monro, and occasionally in the third ventricle or intervening parenchyma. Neurocytoma often causes clinical signs associated with increased intracranial pressure, and tumor extension or obstructive hydrocephalus may cause visual disturbances, altered consciousness, and loss of memory. Neurocytomas are often only discovered incidentally. Hemorrhagic onset is quite infrequent. We describe a case of neurocytoma manifesting as intraventricular hemorrhage with transient memory disturbance.

Case Report

A 43-year-old right-handed man was admitted to our hospital with severe headache and progressive disturbance of consciousness. Neurological examination found no abnormalities other than somnolence. Computed tomography showed intraventricular hemorrhage, and a small mass lesion with calcification on the wall of the left lateral ventricle (Fig. 1A, B). Magnetic resonance (MR) imaging showed the lesion as mixed intensity with heterogeneous enhancement on T1-weighted imaging with gadolinium-diethylenetriaminepenta-acetic acid, and mixed intensity on T2-weighted imaging (Fig. 1C–F). The left body of the fornix was not visualized. Angiography showed no vascular malformations, although faint stains around the calcification were detected in the late venous phase.

His level of consciousness gradually improved, and the intraventricular hematoma diminished in size. However, the patient reported anterograde amnesia despite clear mental state, and underwent neuropsychological examinations using the Wechsler Adult Intelligence Scale-Revised (WAIS-R), Mini-Mental State Examination (MMSE), Kanahiroi Test, Word Fluency Test, Auditory Verbal Learning Test (AVLT), and Raven Colored Progressive Matrix (RCPM). Verbal intelligence quotient (IQ) was 110 and performance IQ was 102 on the WAIS-R. Scores on other tests included: 27/30 on MMSE, 35/36 on RCPM, and slightly low scores on the Kanahiroi and Word Fluency Tests. However, scores on the AVLT were significantly lower compared to other examinations, for both immediate and delayed recall, particularly for delayed recall on hospital day 9 (Table 1). Assessment of his cognitive state indicated mild impairment of attention and...
severe anterograde amnestic syndrome, especially for verbal memory.

The preoperative diagnosis was cavernous hemangioma or intraventricular tumor. Gross total removal of the tumor was performed through a transcrallosal approach on hospital day 27 with the aid of an MR imaging-guided neuronavigation system to minimize the callosal section. The tumor tissue was brown-colored, soft, and hemorrhagic with some small vessels, and contained an area of calcification. The tumor was attached to the inferolateral surface of the lateral ventricular wall, apart from the fornix.

Histological examination found compact round cells with relatively hyperchromatic nuclei and clear cytoplasm, displaying a cohesive pattern and neuropil-like pale islands (Fig. 2A). Small areas of calcification were scattered among the tumor cells with small blood vessels intervening. Tumor cells showed immunoreactivity for synaptophysin (Fig. 2B) and neurofilament protein, and a small number of cells were immunoreactive for glial fibrillary acidic protein and vimentin. MIB-1 staining index was quite low. The histological diagnosis was neurocytoma. Transudation of blood cells from the thin walls of the blood vessels was marked (Fig. 2C).

The patient was discharged on hospital day 55, without adjuvant therapy. Postoperative neuropsychological examinations were performed on days 35, 101, and 269 after symptom onset. General higher cortical function and attention were not disturbed postoperatively and the gradual improvements were noted in the AVLT for both immediate and delayed recall (Table 1).

**Discussion**

Spontaneous hemorrhage was detected in 94 of 1841 primary brain tumors (5.1%), most commonly in pituitary adenoma followed by glioblastoma. Of the 45 cases excluding pituitary adenomas, only five tumors were located in the ventricular system, in the lateral (n = 1), third (n = 2), and fourth ventricles (n = 2). However, histological findings were not described. Intraventricular hemorrhage often occurs with neurocytomas, as the tumor affects the central structures of the ventricular system such as the foramen of Monro or third ventricle. Only seven cases of neurocytomas presenting with intraventricular hemorrhage have been reported (Table 2). Sudden death in one patient occurred due to intraventricular hemorrhage of neurocytoma.

The cause of bleeding in neurocytomas is unclear. Numerous thin-walled tumor vessels may be responsible for the bleeding. High vascularity, hemodynamic stress caused by arteriovenous shunting, fragile tumor vessels, and venous occlusion by tumor encasement are all factors leading to hemorrhage in gliomas. However, microvessels in neurocytoma consist of endothelial cells without fenestrations but with tight junctions, similar to the vessels in low-grade gliomas. Vessel structure is thus variable in neurocytomas. The present case did
### Table 1 Results of neuropsychological examinations

<table>
<thead>
<tr>
<th></th>
<th>WAIS-R</th>
<th>MMSE</th>
<th>Kanahiroi test</th>
<th>Word fluency</th>
<th>AVLT</th>
<th>RCPM</th>
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<tbody>
<tr>
<td></td>
<td>VIQ</td>
<td>PIQ</td>
<td></td>
<td>Category</td>
<td>Letter</td>
<td>Immediate</td>
</tr>
<tr>
<td>Day 9</td>
<td>110</td>
<td>102</td>
<td>27</td>
<td>16-11-10</td>
<td>10-11-8</td>
<td>4-5-4-4-3</td>
</tr>
<tr>
<td>Day 35</td>
<td>—</td>
<td>—</td>
<td>25</td>
<td>18-13-7</td>
<td>10-6-8</td>
<td>3-5-5-5-5</td>
</tr>
<tr>
<td>Day 101</td>
<td>—</td>
<td>—</td>
<td>29</td>
<td>15-11-8</td>
<td>3-8-8</td>
<td>5-7-8-7-10</td>
</tr>
<tr>
<td>Day 269</td>
<td>—</td>
<td>—</td>
<td>27</td>
<td>18-12-11</td>
<td>11-13-8</td>
<td>4-8-7-7-11</td>
</tr>
</tbody>
</table>


Fig. 2 Photomicrographs of the surgical specimen showing compact round cells with relatively hyperchromatic nuclei and clear cytoplasm with cohesive pattern and neuropil-like pale islands (A: HE stain, ×200), immunoreactivity for synaptophysin (B: ×200), and a thin-walled capillary with transudation of red blood cells (C: HE stain, ×200).

not appear highly vascular on angiography, but had mild vascularity at surgery, and numerous thin-walled tumor vessels were found by histological examination. These fragile blood vessels were considered responsible for the hemorrhage in the present case.

Transient verbal memory disturbance in this case may have been related to dysfunction of the fornix as MR imaging indicated that the left corpus callosum and fornix appeared to have been damaged by hemorrhage. Unilateral fornix injury can cause transient or permanent memory disturbances, as dysfunction of the contralateral fornix may occur in addition to disconnection of the ipsilateral thalamo-limbic circuit if the fornix is injured. Characteristics of memory disturbance associated with fornix injuries include predominantly anterograde episodic memory, in contrast to hippocampal memory disturbance, which shows both antero- and retrograde amnesia. Dominance of visual memory by the right fornix and verbal memory by the left fornix may also be apparent, as with the hippocampus. Damage to the bilateral fornices by trauma or infarction results in severe amnesia. Partial preservation of the memory function in the present case may be because the left fornix was injured but not completely destroyed, since the site of tumor attachment was inferolateral to the fornix rather than inferomedial.

Surgery should be planned to avoid injury to the fornix, as recent memory disturbances can improve gradually over the long term. Two approaches can be used for lateral ventricular tumors, the interhemispheric transcallosal approach and the transcortical approach. The transcallosal approach is suitable for small lesions or lesions attached to the surface of the thalamus, due to the low risk of fornical injury during tumor removal and the small callosal section, which may not cause any new damage to the higher cortical functions such as disconnection syndrome. The risk of memory disturbance is similar for the two approaches, and permanent memory disturbance is considered to be attributable to preoperative cognitive dysfunction. The transcortical approach carries the risk of postoperative seizure and subcortical fiber injury in the dominant frontal lobe. Neuronavigation systems may be useful to minimize dissection of the corpus callosum and enter the lateral ventricle without injuring the fornix. Detecting the location of the tumor attachment using preoperative imaging is crucial for removal of the neurocytoma without memory loss.

Neurocytoma should be considered in the
Table 2  Clinical summary for seven cases of neurocytoma manifesting as intraventricular hemorrhage

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>Age (yrs)/Sex</th>
<th>Symptom</th>
<th>Angiography</th>
<th>Macroscopic findings</th>
<th>Histological findings</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Smoker et al. (1991)</td>
<td>26/M</td>
<td>memory disturbance</td>
<td>early vein</td>
<td>ND</td>
<td>hemosiderin deposits</td>
<td>total resection resection</td>
</tr>
<tr>
<td>Goergen et al. (1992)</td>
<td>18/F</td>
<td>sudden headache</td>
<td>not remarkable</td>
<td>ND</td>
<td>ND</td>
<td>total resection resection</td>
</tr>
<tr>
<td>Okamura et al. (1995)</td>
<td>23/M</td>
<td>none</td>
<td>not remarkable</td>
<td>soft hemorrhagic tumor</td>
<td>numerous thin-walled capillary</td>
<td>total resection resection (twice)</td>
</tr>
<tr>
<td>Balko and Schultz (1999)</td>
<td>38/M</td>
<td>sudden death</td>
<td>not performed</td>
<td>large hemorrhagic tumor</td>
<td>hemosiderin deposits, arteriosclerosis</td>
<td>no surgery</td>
</tr>
<tr>
<td>Jamshidi et al. (2001)</td>
<td>22/F</td>
<td>sudden headache</td>
<td>vascular blush</td>
<td>intratumoral blood clot</td>
<td>ND</td>
<td>subtotal resection resection</td>
</tr>
<tr>
<td>Metellus et al. (2001)</td>
<td>38/M</td>
<td>consciousness disturbance</td>
<td>not remarkable</td>
<td>ND</td>
<td>ND</td>
<td>total resection resection</td>
</tr>
<tr>
<td>Hanel et al. (2001)</td>
<td>35/F</td>
<td>consciousness disturbance</td>
<td>ND</td>
<td>soft hemorrhagic tumor</td>
<td>numerous thin-walled capillaries</td>
<td>total resection resection</td>
</tr>
<tr>
<td>Present case</td>
<td>43/M</td>
<td>transient memory disturbance</td>
<td>delayed stain</td>
<td></td>
<td></td>
<td>total resection resection</td>
</tr>
</tbody>
</table>

ND: not described.

differential diagnosis of tumor manifesting as intraventricular hemorrhage. The fragility of the tumor vessels may be the origin of hemorrhage in neurocytoma. Surgical treatment for neurocytomas should be planned to minimize fornix injury and avoid the risk of permanent memory disturbance.

References


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