Ganglioglioma in the Basal Ganglia Totally Resected by a Trans-Distal Sylvian Approach

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Gangliogliomas are central nervous system neoplasms composed of a mixture of neuronal and glial elements. They are rare tumors, observed in only 0.4% of all brain tumors (Zulch 1965, 1979). The most frequent location of these tumors is in the cerebral hemispheres, especially in the temporal lobe. Midline gangliogliomas, located at the basal ganglia, thalamus, hypothalamus and brain stem, are extremely rare (Henry et al. 1978; Demierre et al. 1986; Castillo et al. 1990; Silver et al. 1991; Haddad et al. 1992; Celli et al. 1993; Lang et al. 1993). Surgical resection has been advocated as the optimum treatment for patients with gangliogliomas. Extent of resection is the main prognostic factor in the treatment of gangliogliomas (Demierre et al. 1986; Russel and Rubinstein 1989), however, there have been only a few reports of patients with midline gangliogliomas resected totally.

In this report, a rare case of ganglioglioma in the basal ganglia extending into the thalamus is presented. The tumor was resected totally by a trans-distal Sylvian approach.
CASE REPORT

An 18-year-old man, a physical worker, presented with a 7-month history of progressive involuntary movement in his right hand, a 4-month history of visual field defect on the right side, and occasional headache.

Examination. Neurologic examination revealed incongruous right homonymous hemianopsia. Computed tomography (CT) showed a deep-seated cystic mass with an enhancing lesion. Magnetic resonance imaging (MRI) further defined the structure of the tumor. The solid part of this tumor was located

Fig. 1. Pre-operative magnetic resonance imaging (MRI) demonstrates a heterogeneous tumor with cystic and solid components. The solid components were localized in the basal ganglia, and the cystic components extended to the thalamus. The cystic portion was hypointense on T1-weighted images and hypointense on T2-weighted images. The solid portion was intensely enhanced with Gadolinium diethylene-triamine-pentaacetic acid (Gd-DTPA). Upper left: axial T1-weighted image; Upper right: axial T1-weighted image with Gd-DTPA enhancement; Lower left: axial T2-weighted image; Lower right: coronal T1-weighted image with Gd-DTPA enhancement.
predominantly in the basal ganglia. The tumor extended into the thalamus where it contained cystic components. The cystic portion was hypointense on T1-weighted images and hyperintense on T2-weighted images, and the solid part was enhanced intensely with Gadolinium diethylene-triamine-pentaacetic acid (Gd-DTPA) (Fig. 1). Cerebral angiography revealed an avascular area corresponding to this lesion.

Operation. Surgery was performed by a trans-distal Sylvian approach. Through a large frontotemporal craniotomy, the Sylvian fissure was dissected thoroughly, approximately 8 cm (Fig. 2). A 2 cm incision was made at the posterior part of the insular cortex near the superior insular sulcus to expose the tumor surface. The tumor consisted of both cystic and solid parts. The solid part was brown, firm, and could not be aspirated. It was resected en bloc. The
cystic parts were excised together with the cyst wall.

**Pathologic examination.** Microscopically, the tumor had a diffuse background of small to midsized astrocytic nuclei in a fibrillary mesh (Fig. 3). These cells had an eosinophilic processes and showed positive reaction to GFAP stain, confirming their identity as glial cells. There were large polymorphic cells with vesiculated nuclei, prominent nucleoli, and positive reaction in Nissl stain, which were identified as ganglion cells. Some of these were binucleate. These ganglion cells showed synaptophysin immunoreactivity surrounding the cell bodies (Fig. 4). This constellation of findings was characteristic of ganglioglioma.

**Postoperative course.** The patient fully recovered from involuntary movement and homonymous hemianopsia. Postoperative MRI revealed total resection of the tumor (Fig. 5). The patient returned to work at his previous occupation.

**DISCUSSION**

The most common symptom of supratentorial ganglioglioma is seizure (Henry et al. 1978; Johannsson et al. 1981; Castillo et al. 1990; Silver et al. 1991; Haddad et al. 1992; Celli et al. 1993; Lang et al. 1993). In contrast, deep-seated gangliog-
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Ganglioglioma is characterized by headache (Lang et al. 1993) or focal neurologic deficit (Johannsson et al. 1981; Haddad et al. 1992; Lu et al. 1993). In our case, involuntary movement was considered to be a focal sign of the basal ganglia. Incongruous homonymous hemianopsia was due to the compression of a part of the optic radiation.

Ganglioglioma appears as a low- or isodensity mass with contrast enhancement on CT. Calcification is seen in 25 to 56% of cases (Demierre et al. 1986; Castillo et al. 1990; Silver et al. 1991; Haddad et al. 1992; Celli et al. 1993). Approximately 44 to 66% of cases are cystic lesions (Demierre et al. 1986; Castillo et al. 1990; Haddad et al. 1992; Celli et al. 1993). MRI has been demonstrated to be more precise in identifying tumor presence compared to CT. Ganglioglioma appears as a hypointense mass on T1-weighted images, hyperintense on T2-weighted images, and is significantly enhanced with Gd-DTPA administration (Demierre et al. 1986; Silver et al. 1991; Haddad et al. 1992). However, a preoperative diagnosis of ganglioglioma is rarely made radiologically, since imaging findings are nonspecific.

The diagnosis of ganglioglioma is usually made by histologic proof of neoplastic ganglion and glial cells. To be proven a ganglioglioma, a tumor must contain both glial and ganglion cell elements (Russel and Rubinstein 1989). Astrocytic components are usually verified by GFAP immunoreactivity demonstrating im-

Fig. 4. Anti-synaptophysin immunoreactivity was demonstrated in the ganglion cell component (original magnification ×400).
munopositive cell bodies and processes. The identification of neoplastic ganglion cells using light microscopy is also difficult. Recently, monoclonal antibodies for neuronal cells have been reported as useful markers for neoplastic ganglion cells, anti-synaptophysin in particular (Diepholder et al. 1991; Miller et al. 1993).

Most studies have suggested that patients with gangliogliomas have long survival times (Johannsson et al. 1981; Haddad et al. 1992). Surgical resection has been advocated as the optimum treatment for patients with ganglioglioma (Demierre et al. 1986; Haddad et al. 1992; Celli et al. 1993; Lang et al. 1993). Most of the patients in these reports have tumors in the temporal lobe, frontal lobe and parietal lobe, not in deep gray matter. Less is known about the optimum treatment of patients with midline gangliogliomas (Lang et al. 1993). Surgical treatments for midline gangliogliomas remain difficult. However, the recent

Fig. 5. Post-operative MRI confirms total resection of the tumor. Normal post-operative changes are observed, but there is no evidence of residual tumor. Upper left: axial T1-weighted image; Upper right: axial T1-weighted image with Gd-DTPA enhancement; Lower left: axial T2-weighted image; Lower right: coronal T1-weighted image with Gd-DTPA enhancement. Arrows indicate the route of a trans-distal distal trans-Sylvial approach.
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development of microsurgical techniques and increased knowledge of microsurgical anatomy have led to more frequent success in the resection of these lesions.

Some literature has recommended a transcallosal approach for thalamic tumors (Apuzzo et al. 1982; Ehni 1984). In our case, a well-circumscribed lesion extended from the basal ganglia to the outer portion of the thalamus. The solid portion of the tumor existed predominantly in the basal ganglia. The tumor in the thalamus was made up of cystic components. We thought that the cystic portion in thalamus could be resected in the same approach to the basal ganglia. Therefore, we employed a trans-Sylvian approach. Through the distal portion of the Sylvian fissure, we resected the thalamic lesion without damaging the pyramidal tract. The tumor was totally resected by this method, and the patient showed considerable improvement of his symptoms. Even with midline gangliogliomas, maximum resection should be attempted by an approach with minimum damage to normal brain tissue.

References

