Mixed Cavernous Angioma and Glioma (Angioglioma) in the Hypothalamus
—Case Report—

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Abstract

A 54-year-old female presented with a huge mixed cavernous angioma and astrocytoma in the hypothalamus manifesting as headache, visual field defect, gait disturbance, and convulsion. Radiological studies revealed a huge suprasellar tumor encasing all the major cerebral vessels. Craniotomy disclosed a hemorrhagic tumor poorly demarcated from the surrounding brain which was partially removed. Histological examination of the operative specimen revealed cavernous angioma with low grade glioma in the periphery. The residual tumor responded to radiation therapy remarkably well. An autopsy conducted 3 years later revealed a small hypothalamic astrocytoma with abundant vasculature.

Key words: mixed tumor, cavernous angioma, hypothalamic astrocytoma, angioglioma, vascular encasement

Introduction

Cavernous angioma can arise anywhere in the central nervous system but is very rare in the hypothalamus. Tumors with the characteristics of both cavernous angioma and glioma are also rare and are occasionally called angioglioma, a term introduced to describe a highly vascular cerebellar neoplasm and subsequently used for glial neoplasm possessing angioma-like vasculature.

We describe a patient with cavernous angioma in the hypothalamus with features of glioma.

Case Report

A 54-year-old female developed headache, visual disturbance, and unsteady gait a few months prior to a generalized seizure. She was admitted to our hospital on December 18, 1987. Past medical history included diphtheria at the age of 10 years. Physical examination revealed an emaciated anemic woman with a hard pelvic mass which was removed subsequently and proved to be a leiomyoma of the uterus. She was disoriented about time and lethargic with right homonymous hemianopsia. Routine laboratory examinations were within normal limits except for mild anemia. Endocrinological studies disclosed an elevated serum prolactin level (64 ng/ml) with anterior hypophyseal hypofunction. Serum alpha-fetoprotein, carcinoembryonic antigen, and human chorionic gonadotropin levels were all normal.

A skull roentgenograph showed erosion of the dorsum sellae. Computed tomography (CT) showed dilated lateral ventricles and a huge low density suprasellar mass (maximum coronal diameter 6.6 x 7.0 cm) with postcontrast heterogeneous enhancement (Fig. 1). Magnetic resonance (MR) imaging revealed a relatively well-demarcated lesion appearing as a heterogeneously low intensity area on T1-weighted images and a diffusely high intensity area on T2-weighted images (Fig. 2). The tumor encased all the major cerebral vessels, including the bilateral internal carotid, anterior cerebral, middle cerebral, posterior cerebral, and basilar arteries. Angiography showed superior displacement of the bilateral inter-
nal carotid bifurcations and posterior displacement of the distal portion of the basilar artery. The lateral and medial lenticulostrate arteries were irregular but there was no identifiable tumor stain, arteriovenous shunt, or abnormal vein (Fig. 3). A left persistent primitive trigeminal artery was incidentally found.

After admission, she became progressively obtunded and semicomatous. A ventriculoperitoneal shunt was installed resulting in improved conscious level postoperatively. On January 14, 1988, the tumor was partially removed (approximately 20%) via the orbitozygomatic approach. The tumor was soft and hemorrhagic. Despite the well-demarcated appearance on MR images, the tumor was ill-defined and infiltrated the surrounding brain. The tumor enclosed the optic nerves, chiasm, bilateral internal carotid arteries, left anterior cerebral artery, and left oculomotor nerve. The pituitary stalk and basilar artery were included in the tumor. Hemostasis was difficult because of the numerous fine vessels in the tumor.

Histological examination of the operative specimen showed most of the tumor consisted of ectatic vascular channels of various sizes without lamina elastica or muscularis. There was no intervening glial tissue in the main portion of the tumor (Fig. 4 upper). There were many areas of hemorrhage with fibrin deposit with prominent papillary endothelial proliferations. The periphery of the tumor included areas of microcysts and atypical cells mixed with abnormal vessels (Fig. 4 lower). Such atypical cells had irregular nuclear size and shape, and included occasional bizarre large cells. These cells were scattered and irregularly aggregated in some areas. Many cells in these areas were positively stained for glial fibrillary acidic protein (GFAP). These findings are compatible with glioma rather than extensive gliosis.

Postoperatively, her neurological status was essentially unchanged. She received local radiation therapy (35 Gy) postoperatively. She received replacement therapy of corticosteroid and thyroid hormone and regained weight. Serial follow-up CT scans (Fig. 5) revealed a residual large suprasellar tumor essentially unchanged in size (maximum coronal diameter 5.1 × 3.1 cm) until April, 1990. She subsequently did not seek medical attention and was found dead at her home on October 4, 1991.

The autopsy was performed by coroners from Osaka University Medical School, Department of Legal Medicine. She was extremely cachexic and dehydrated. The brain was carefully removed but there was no extracerebral skull base tumor or tumor bulging from the hypothalamus. Coronal section revealed ventricles of normal size and a small soft xanthochromic tumor in the left hypothalamic area.
Histological examination of the autopsy specimen showed the tumor was well-demarcated from the surrounding brain and consisted of irregularly distributed GFAP-positive astrocytic cells with pleomorphic nuclei (Fig. 7). There were areas of numerous thick-walled hyalinized vessels, some of which were thrombosed, but there was no area which could be defined as cavernous angioma although multiple sections were examined.

Discussion

Only three cases of mixed cavernous angioma and glioma have been reported, a frontal lobe oligodendroglioma associated with a cavernous angioma, and two cavernous angiomas, one with oligoden-
droglioma and one with astrocytoma referred to as "angioglioma." However, angioglioma is a controversial concept and is not a generally accepted histological entity. The term includes hypervascular low grade glioma, and glioma with hemangioblastoma, cavernous angioma, or arteriovenous malformation components. Bonnin et al. proposed that the term should be limited to only true mixed tumors of glial and vascular tissue origin. Lombardi et al. recently reviewed their experience of angioglioma (highly vascular low grade glioma) and suggested the term be abandoned because there was no difference in clinical or prognostic features between gliomas with and without angiomatous components. In contrast, Mathern et al. distinguished this entity from glioma with cavernous angioma or hemangioblastoma and proposed that angioglioma is a subgroup of cerebral glial neoplasm. The definition of angioglioma is thus confusing. However, astrocytoma, particularly arising in the wall of the third ventricle, is intensely vascular and the vasculature may be the predominant component, so we believe that there is a group of highly vascular low grade gliomas which may require a specific name such as angioglioma. In our patient, the extent of the vascular component and absence of intervening glial tissue in the main portion of the tumor suggested cavernous angioma rather than secondary vascular proliferation. Therefore, our final diagnosis was mixed cavernous angioma and glioma, but the term angioglioma is applicable because there was no clear distinction between the mixed cavernous angioma and glioma, and highly vascular low grade glioma. Reactive gliosis, particularly oligodendroglial proliferation associated with vascular malformation, is relatively common and occasionally atypical glial cells are seen. These cases must be excluded from angioglioma or mixed angioma and glioma. In our case, the microcystic change and atypicality of the astrocytic cells suggested tumor rather than gliosis. The postmortem examination revealed predominantly astrocytic tumor with hyalinized angiomatous component.

Cavernous angioma located in the hypothalamus is rare, with only nine cases reported. Cavernous angioma usually appears as a high density mass on precontrast CT due to calcification or hemorrhage, but in our patient the tumor appeared as a low density mass with heterogeneous enhancement and ill-defined borders. These findings may be due to mixed angioma and glioma. The efficacy of radiation therapy on cavernous angioma has not been established, but Shibata and Mori reported a series of radiosensitive cases. In one of these, the tumor disappeared 19 months after irradiation, suggesting a delayed effect of radiation on this tumor. Our case was another example of extremely radiosensitive cavernous angioma. Considering the risk of radical surgery, biopsy or optic nerve decompression followed by radiation therapy is the first choice for treatment of this type of tumor.

Extensive vascular encasement was another unusual aspect of this case. This feature is occasionally seen in meningioma and pituitary adenoma in the suprasellar region. Mixed cavernous angioma and glioma should be included in the differential diagnosis of the tumor encasing cerebral vessels.

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