Multiple Brain Tumors of Different Cell Types with an Unruptured Cerebral Aneurysm

—Case Report—

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Abstract

A rare case of coexistent Burkitt-type lymphoma and meningioma associated with an unruptured cerebral aneurysm is presented. A 49-year-old male complaining of headache and right hemiparesis was admitted to our hospital. Neuroradiological examination revealed a multinodular mass in the left frontal convexity and an unruptured cerebral aneurysm at the M₁ portion of the left middle cerebral artery. He underwent an operation for tumor removal and aneurysm clipping. Histological examination revealed the tumor to be a typical meningotheliomatous meningioma without malignancy. However, a second operation was necessary for another tumor invading into the left frontal lobe, which proved to be a Burkitt-type lymphoma. The second tumor may have been resulted from irritative effect of the first tumor, but the aneurysm was considered purely coincidental.

Key words: multiple brain tumors, lymphoma, meningioma, cerebral aneurysm

Introduction

Multiple tumors of the central nervous system (CNS) with different histologies are uncommon in patients without phacomatoses or a history of radiation therapy. The Burkitt-type CNS lymphoma is extremely rare, and few cases have been reported.⁸⁾ A rare case of coexistent Burkitt-type lymphoma and meningioma occurring in the left frontal lobe and frontal convexity, without phacomatoses or previous radiation therapy, is described. An unruptured aneurysm at the junction of the left middle cerebral and lateral lenticulostriate arteries was also present.

Case Report

A 49-year-old male was admitted to our hospital on May 9, 1989, complaining of headache, memory disturbance, and weakness in his right lower extremity. His physical status was normal. Neurological examination found motor weakness in the right leg,

motor aphasia, and slight papilledema in the left optic fundus. Blood tests were normal, including T- and B-cell percentages in the peripheral lymphocytes and a T-helper to T-suppressor cell ratio. Computed tomographic (CT) scans showed an irregular, highly enhanced nodular mass in the left frontal lobe. An axial T₁-weighted magnetic resonance (MR) image demonstrated a round low-intensity mass in the left frontal lobe (Fig. 1 *left*). A T₂-weighted MR image demonstrated the mass as a mixed intensity area surrounded by high intensity (Fig. 1 *right*). A left carotid angiogram disclosed an aneurysm at the junction of the middle cerebral and lateral lenticulostriate arteries (Fig. 2). A malignant glioma or meningioma associated with an aneurysm at the origin of the left lenticulostriate artery was diagnosed.

A left frontotemporal craniotomy was performed on May 16, 1989. Incision of the dura mater exposed a whitish-gray, well-defined tumor attached to the frontal dura. The tumor was clearly demarcated from the frontal cortex and easily dissected. Following total removal of the tumor, the tumor bed appeared anemic, but showed no definite evidence for

Fig. 1 *left*: T₁-weighted (inversion recovery; TR 2000 msec, TI 600 msec, TE 35 msec) axial MR image, demonstrating a round low-intensity mass in the left frontal lobe. *right*: T₂-weighted (spin echo; TR 2000 msec, TE 92 msec) axial MR image, showing mixed intensity mass surrounded by high intensity.

Fig. 2 Left carotid angiogram, disclosing an aneurysm (arrow) at the junction of the middle cerebral and lenticulostriate arteries associated with marked shift of the anterior cerebral artery to right.

another mass lesion. A small saccular aneurysm at the junction of the middle cerebral and the lateral lenticulostriate arteries was successfully clipped.

He made an uneventful recovery but complained of headache postoperatively. A CT scan taken 2 weeks later showed a high-density mass in the left frontal lobe under the craniotomy site, and his neurological status became aggravated again.

On June 6, 1989, a left frontotemporal craniotomy was performed as before. A grayish, nodular tumor

Fig. 3 Photomicrograph of a tumor specimen from the first operation, showing a typical meningotheliomatous meningioma, with lobulated arrangement of tumor cells and conspicuous whorl formations. HE stain, $\times 200$.

Fig. 4 Photomicrograph of a tumor specimen from the second operation, showing diffuse proliferation of undifferentiated lymphocytes with little variation in size and shape. Macrophages containing cell debris are scattered throughout the tumor cells. HE stain, $\times 200$.

was found at the previous site. The main mass was well demarcated, but diffuse invasion into the left frontal lobe had occurred. The tumor was less vascular and was grossly totally removed.

Postoperative recovery was uneventful. Radiation therapy and adjuvant chemotherapy with cyclophosphamide, doxorubicin, vincristine, and prednisolone, with intrathecal methotrexate, were started. No other tumor was found clinically. A CT scan taken 6 months after the second operation found no evidence of recurrence.

Histological examination of the first tumor found a typical meningotheliomatous meningioma, with

lobulated arrangement of tumor cells and conspicuous whorl formations (Fig. 3). The second tumor, however, had a diffuse proliferation of undifferentiated lymphocytes with little variation in size and shape. Macrophages containing cell debris were scattered among the tumor cells (Fig. 4). The tumor cells demonstrated marked cytoplasmic pyroninophilia with methyl green pyronic stain but no reaction to the periodic acid-Schiff or naphthol-AS-D-chloroacetate reagents. Immunoperoxidase staining with peroxidase-antiperoxidase technique was positive for immunoglobulin M and kappa chain only. The histological diagnosis was diffuse Burkitt-type lymphoma.

Discussion

Many multiple brain tumors with different cell types have been reported, but most were coexistent meningioma and glioma.^{1,6,7,18,24} Possible etiologies of multiple brain tumors with different histologies include: 1) carcinogenesis causing neoplastic transformation in different cells¹⁷; 2) embryonal rests causing different brain tumors at different times²; and 3) meningeal tumor inducing a malignant transformation in the underlying brain parenchyma.²¹ Only 10 cases of preoperatively confirmed multiple brain tumors with different cell types have been reported.^{1,4-6,10,14,15,18,19,23,24} Close locations, frontal lobe astrocytoma and sphenoid ridge meningioma,¹⁸ intrasellar meningioma and craniopharyngioma,⁶ and frontal lobe astrocytoma and sphenoid ridge meningioma,²⁴ occurred in three of the 10 cases. However, definite preoperative diagnosis of two adjacent tumors was made in only one.²⁴

Malignant lymphomas are very often accompanied by immunosuppressive therapy or autoimmune disorders. Burkitt-type lymphoma is an undifferentiated, B-cell type malignant lymphoma predominantly occurring in childhood and usually involving multifocal lesions, such as abdominal viscera, retroperitoneal soft tissue, bone, thyroid glands, salivary glands, or the CNS.³ Increased Epstein-Barr virus reaction is frequently demonstrated in Africa, but elsewhere is infrequently associated with Burkitt-type lymphoma.²⁸ In general, the prognosis for Burkitt-type lymphoma is poor, but it responds to radiation therapy and chemotherapy. Few cases of primary intracranial Burkitt-type lymphoma have been reported.^{8,12,16,26} Giromini *et al.*⁸ reported an interesting case of Burkitt-type lymphoma and astrocytoma in an 11-year-old boy. A left temporo-occipital lymphoma was found 6 months after resection of a left temporoparietal astrocytoma, with

no direct communication between the tumors.

Brain tumors associated with cerebral aneurysms are not uncommon, with a reported incidence of 0.3–0.7% of all brain tumors.^{9,25} Pituitary adenoma is the most frequent brain tumor associated with cerebral aneurysms.²⁷ The etiology of such association remains unknown. Mechanisms such as increased blood supply to the tumor or the direct mass effect on the vascular wall have been postulated.^{11,20} High serum levels of human growth hormone in acromegaly or an “intrinsic dysgenetic metabolic” defect inducing atherosclerotic changes in the arterial wall may also be factors in aneurysm formation.^{13,22}

The development of two brain tumors with different cell types in the same region is unique to this case. There was no evidence of phacomatoses, immunodeficiency, or previous radiation therapy. In addition, the case was complicated by an unruptured aneurysm, and one brain tumor was a quite rare, diffuse Burkitt-type lymphoma. A definite diagnosis of two different tumors by preoperative CT and MR imaging was difficult, because the tumors developed in almost the same place. The tumor removed in the first operation was a typical meningotheliomatous meningioma without malignancy, but an irritative effect may have resulted in the malignant lymphoma. The association of the cerebral aneurysm was, however, considered purely coincidental. There was no direct contact between the tumors and the aneurysm, and no increased blood supply from the middle cerebral artery to the tumors was found by preoperative angiography or surgery. Aneurysms associated with brain tumors are known to have a significantly high risk of hemorrhage after tumor removal.²⁵ Therefore, we recommend a single radical operation to treat both the aneurysm and the brain tumor. The tumor bed was not thoroughly explored at the first operation, and the lymphoma may have existed unrecognized. Adjuvant chemoradiotherapy immediately after the second operation effectively diminished the Burkitt-type lymphoma.

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