Central Neurocytoma with Craniospinal Dissemination

Kumiko Ando1*, Reiichi Ishikura1, Tsutomu Morikawa1, Norio Nakao1, Jota Ikeda2, Tsuyoshi Matsumoto3, and Norio Arita4
1Department of Radiology, Hyogo College of Medicine
Mukogawa-cho, Nishinomiya, Hyogo 663-8501, Japan
2Department of Radiology, Ashiya Municipal Hospital
3Department of Neurosurgery Goushi Hospital
4Department of Neurosurgery, Hyogo College of Medicine
(Received September 26, 2002; Accepted October 30, 2002)

We report a case of central neurocytoma that manifested tumor recurrence with craniospinal dissemination. Imaging studies before surgery showed a markedly enhanced tumor with small cysts and calcification, which invaded the adjacent brain parenchyma, located in the posterior horn of the right lateral ventricle. Proton MR spectroscopy showed markedly elevated choline and lactate peaks with a strongly diminished N-acetylaspartate peak. Two years after neurosurgical intervention, the tumor showed multiple craniospinal dissemination in the middle cranial fossa and the intradural extramedullary space of the spine.

Keywords: central neurocytoma, cranio-spinal dissemination, MRI, MR spectroscopy

Introduction

Central neurocytoma is a neoplasm that usually arises from the anterior part of the lateral ventricle near the foramen of Monro in young adults. Central neurocytoma corresponds histologically to WHO grade I–II and usually has a favorable clinical course. Its malignant clinical course is rare.1,2

We report a case of central neurocytoma with cellular pleomorphism that manifested tumor recurrence with craniospinal dissemination two years after surgical intervention.

Case Report

A 31-year-old female suffered from progressive headaches and nausea of one month’s duration. On admission, she was in the 8th month of pregnancy and exhibited a decreased level of consciousness. She had left hemiparesis and bilateral abducens nerve palsy. Laboratory data were normal except for an elevated AFP level (197.8 ng/ml) due to pregnancy.

Her own history and familial history were otherwise unremarkable. CT images showed a large heterogenous low-density mass in the right cerebrum with spotty calcification (Fig. 1). MRI revealed the tumor was mainly located in the posterior horn of the right lateral ventricle and extended into the adjacent ipsilateral thalamus (Fig. 2a–c). The tumor

Fig. 1. A non-contrast CT image shows a large heterogenous hypodense mass in the right cerebrum with spotty calcification.
showed areas of low to high signal intensity in T₁-weighted images (Fig. 2a), and showed heterogeneous high signal intensity on T₂-weighted MRI images (Fig. 2c). The tumor showed intense and heterogenous enhancement with Gd-DTPA (Fig. 2b). These findings suggested that the mass had a solid portion with small cysts and hemorrhaging.

MR spectroscopy was performed with a 1.5T unit and the CSI method (TR/TE = 2000/270 ms, excitation = 1, FOV = 22 cm, matrix = 16 × 16, thickness = 2 cm). MR spectroscopy within the tumor revealed markedly elevated choline (Cho) peaks at 3.2 ppm and very low N-acetylaspartate (NAA) peaks at 2.0 ppm. Elevated lactate (Lac) and lipid peaks at 1.3 ppm were also observed (Fig. 3).

Following a successful delivery by Caesarian section in the 36th week of pregnancy, a suboccipital craniotomy was performed and the tumor was subtotally removed with a supracerebellar approach. The tumor was located in the right posterior horn and extended into the right thalamus.

On histological examination, the tumor cells, which appeared round, displayed a clear cytoplasm and large nuclei. Cellular pleomorphism was observed.

GFAP immunostain was negative, but synaptophysin immunostain was positive (not shown). These findings are consistent with central neurocytoma. The MIB-1 labeling index (MIB-1 LI) was 15%.
Imaging studies after the surgical procedure showed a residual mass in the right lateral ventricle and right thalamus (not shown). The patient received trans-arterial chemotherapy (Mesna 7,500 mg, Ifosfamido 16,750 mg, Cisplatin 300 mg, Etoposido 950 mg) and 50 Gy of radiation therapy.

Two years after the surgical intervention, the patient showed motor weakness in both lower extremities. Cranial and spinal MR images showed a local recurrent mass with multiple craniospinal disseminated tumors in the middle cranial fossa and in the intradural extramedullary space of the spine (Fig. 4a, b).

The patient received additional chemotherapy but died three years after the initial surgical operation.

Discussion

Central neurocytoma accounts for 0.25% to 0.5% of central nervous system tumors. It arises from the septum pellucidum or the wall of the ventricle. The patient’s prognosis is usually favorable and a malignant clinical course is rare.

The typical central neurocytoma is a sharply demarcated, lobulated intraventricular tumor with cysts and calcifications. On MR images, central neurocytoma shows heterogenous intensity both on T1- and T2-weighted images and is variably enhanced with contrast media.

Our case showed cranio-spinal dissemination two years after the surgical intervention. Cellular pleomorphism was revealed histologically. The tumor location was atypical, as it was located in the posterior horn of the right lateral ventricle. Imaging findings were typical, except for the extra-ventricular extension of the tumor into the adjacent
The literature reports five cases of central neurocytomas with craniospinal dissemination. All five reported cases were histologically benign neurocytomas. Thus, histological malignancy may not be related to the dissemination.

All cases manifested craniospinal dissemination after surgical intervention. In five of six cases, including our case, the tumor removal was incomplete. Surgical intervention with residual tumor is suggested as one risk factor for craniospinal dissemination of the central neurocytoma. Time to recurrence from first surgery varies from two months to three years. Central neurocytoma requires long-term follow-up after surgical intervention.

Tumor location was typically in the lateral ventricle near the foramen of Monro in four cases. In the other two cases, including our case, the tumor was located in the trigone, which is an unusual location for neurocytoma.

Extraventricular extension of the tumor was observed in four of six cases with craniospinal dissemination. Two of the cases were histologically benign typical neurocytoma. Wichmann et al. suggested that extraventricular extension of a central neurocytoma indicates malignant transformation of the tumor. Extraventricular extension of the tumor may be observed in a benign central neurocytoma, and it might be a useful predictor of poor clinical outcome.

The metabolic features of central neurocytomas are reported in three cases. All three reported cases are typical benign neurocytomas without craniospinal dissemination. The spectrum of reported cases showed an elevated Cho peak and a severely decreased NAA peak with or without lactate peak (Lac). These spectral features are the same as those of our case and were not deemed useful for predicting the poor prognosis of the neurocytoma.

The elevated Cho peak reflects high proliferation potential in gliomas. In our case, the MIB-1 LI was 15%, very high for a central neurocytoma. However, in Hartmann’s case, the Cho peak was also elevated, but the MIB-1 rate was less than 1%. As Warmuth-Metz commented, the elevated Cho peak may reflect high cellularity in neurocytoma.

It is interesting to note that the NAA peak of the central neurocytoma is decreased, even though the central neurocytoma has a neuronal nature. NAA is assumed to be produced from mature neurons. Low NAA levels in the central neurocytoma may be due to its cellular immaturity.

In conclusion, we reported a case of central neurocytoma that manifested tumor recurrence with craniospinal dissemination two years after the surgical intervention. Imaging studies before surgery showed a tumor in the posterior horn of the right lateral ventricle that invaded the adjacent brain parenchyma. Proton MR spectroscopy showed markedly elevated choline and lactate peaks with a strongly diminished N-acetylaspartate peak, but the MR spectroscopic findings were the same as those of neurocytomas with a favorable clinical course.

References