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Third Ventricular Meningioma

—Case Report—

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Abstract

A 63-year-old man presented with headache. Magnetic resonance imaging showed a mass lesion homogeneously enhanced with gadolinium, which occluded the route from the third ventricle to the aqueduct. The patient underwent surgery for removal of the tumor via the right frontal transcortical-transventricular approach to the third ventricle via the transchoroidal route. Intraoperative diagnosis was meningioma. Total removal of the tumor was achieved in piecemeal fashion (Simpson grade 1). The histological diagnosis was meningothelial meningioma. The patient was discharged without neurological deficits. Third ventricle is a rare and difficult site to remove tumor totally. However, total removal was needed in this case of benign meningioma, so the operative strategy and the differential diagnosis before operation is considered to be very important.

Key words: meningioma, third ventricle, intraventricular meningioma, arachnoid cell, choroid plexus

Introduction

Cerebral ventricles are unusual sites for the occurrence of tumors in the central nervous system. The incidence of tumors follows a regional preference, with ependymomas and primitive neuroectodermal tumors occurring more often in the fourth ventricle and colloid cysts in the third ventricle. Tumors that are most likely to occur in the lateral ventricles include astrocytomas, subependymal giant cell astrocytomas, and ependymomas. Intraventricular

meningiomas are rare tumors, accounting for 0.5% to 5% of all intracranial meningiomas.²⁾ Although some series have reported lateral ventricle meningiomas, there are few case reports of third and fourth ventricle meningiomas. We present our experience with a case of third ventricle meningioma.

Case Report

A 63-year-old man presented with headache. Neurological examination showed no apparent abnormality other than headache, and no apparent findings of neurofibromatosis

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Fig. 1 Coronal (A), axial (B), and sagittal (C) T₁-weighted magnetic resonance images with gadolinium revealing a homogeneously enhanced mass lesion arising in the region of the third ventricle and extending into the aqueduct.

Fig. 2 Right vertebral angiograms, lateral view, showing the tumor fed by the right medial posterior choroidal artery (arrows) in the arterial phase (A) and tumor staining in the late phase (B).

type 2 (NF 2), such as café-au-lait spots or auditory disturbance. The patient had a history of prostatic cancer, but positron emission tomography detected no recurrence or metastatic lesions. Head computed tomography (CT) showed a homogeneously isodense mass lesion in the third ventricle, which caused slightly obstructive hydrocephalus. Magnetic resonance (MR) imaging showed the mass lesion was homogeneously enhanced with gadolinium, which occluded the route from third ventricle to aqueduct (Fig. 1). Angiography showed the tumor was fed by the right medial posterior choroidal artery (Fig. 2).

The patient underwent surgery for removal of the tumor via the right frontal transcortical-transventricular approach to the third ventricle via the transchoroidal route. Intraoperatively, the tumor appeared grossly to be a white, non-vascular rich and relative soft mass (Fig. 3A). The tumor, fed by the right medial posterior choroidal artery, was attached to the third ventricular posterior wall. In addition, no tumor invasion to the right thalamus, or floor and lateral wall of third ventricle was observed. Intraoperative diagnosis indicated meningioma. Total removal of the tumor was achieved in piecemeal fashion (Simpson grade 1) (Figs. 3B and 4).

Histological examination found that no malignant features such as atypical nuclei or mitosis. The tumor consisted of meningotheial cells with whorl formation, which is a typical finding of meningioma. Immunohistochemical

Fig. 3 Operative photographs showing the white soft tumor (T) within the third ventricle (A). The tumor was removed in piecemeal fashion (B). CP: choroid plexus, FOM: foramen of Monro, TSV: thalamostriate vein.

Fig. 4 Coronal (A), axial (B), and sagittal (C) T₁-weighted magnetic resonance images with gadolinium revealed disappearance of the mass lesion.

Fig. 5 Photomicrograph showing the tumor consisted of meningotheial cells with whorl formation, and absence of malignant features such as atypical nuclei and mitosis. Hematoxylin-eosin stain, $\times 20$.

staining was positive for epithelial membrane antigen (EMA), vimentin, and S-100 protein, but negative for carcinoembryonic antigen, glial fibrillary acidic protein, cytokeratin (CK) 7, CK 20, and synaptophysin. The histological diagnosis was meningotheial meningioma (Fig. 5).

No adjuvant therapy was performed, because intraoperative observation confirmed the tumor was grossly removed and the histological finding was benign. The patient was discharged without neurological deficits.

Discussion

The presence of arachnoid cell nests in the normal

choroid plexus stroma has been reported, and thorough examination of the choroid plexus usually reveals small or sometimes larger collections of these cells.^{6,14} The presence of these cells in the choroid plexus has been explained variously.^{6,14} The choroid plexus develops initially from an invagination of the mesenchyme in the thin, roof area of the myelencephalon during the 6th week of gestation.¹⁹ In the 7th to 9th weeks, the telencephalic choroid plexus has started to develop a loose mesenchymal stroma, which is covered by a layer of cells derived from the ependyma.⁴ Arachnoid tissue is transported together with the choroid plexus as the ventricular system invaginates, and by 20 to 40 weeks, the central stroma of the choroid plexus contains meningocytes, connective tissues, and blood vessels.^{2,14} Meningiomas arise from arachnoid cap cells, which are specialized cells in arachnoid granulations. Similarly, intraventricular meningiomas arise from arachnoid cells contained within the choroid plexus. Meningothelial inclusion bodies are normally found in the arachnoid and choroidal tela, and meningiomas arise from this mesenchymal stroma of the choroid plexus.⁸ Third ventricle tumors may arise from the tela of the velum interpositum, which is the space between the two layers of tela in the roof of the third ventricle that contains the posterior medial choroidal arteries and internal cerebral veins. According to the Brain Tumor Registry of Japan, 12th edition (1984–2000), the ventricles are rare sites for the occurrence of meningiomas (1.6% of all meningioma cases), the third (0.1%) and fourth ventricles (0.1%) are less common than the lateral ventricles (1.4%), because the choroid plexus is more bulky in the lateral ventricles.

Intraventricular meningiomas can be any of the various histopathological tumor types (predominantly fibrous, fibroblastic, meningothelial, or psammomatous) defined by the World Health Organization classification of meningiomas. The structure of psammoma bodies in the choroid plexus is very similar to that in meningiomas. In general, meningiomas are diagnosed based only on the morphological features. In this case, immunohistochemistry showed EMA, vimentin, and S-100 protein, consistent with the typical pathological characteristics of meningioma. EMA, derived from human milk fat globule protein, is present in secretory cells and is a marker of meningioma.¹⁰ Vimentin is positive in cells of mesenchymal origin, S-100 protein is a calcium-binding protein and positive in glial cells,¹⁶ and CK is the marker of epithelial cells. The various immunohistochemical responses against these various antigens show the variety of the origin of meningioma, although arachnoid cells of the choroid plexus are assumed to be the origin of intraventricular meningiomas.

Most clinical symptoms of ventricular meningiomas are due to increased intracranial pressure. Lesions grow into the ventricular cavity until obstructive hydrocephalus results. Visual field deficits may be seen in 25% of patients and seizures are rare.² Hypothalamic features with or without endocrinopathy may be seen in third ventricle tumors. Manifestations of NF 2 may be evident on clinical and neurological evaluation. Meningiomas are more common in women, and intraventricular meningiomas are

common in childhood⁷ and rare in adulthood.

The rarity of intraventricular meningiomas in contrast to those with attachment may be due to as yet unknown factors. Multiple meningiomas arising from the dura mater are probably caused by the inherent multicentricity of the dural foci, possibly influenced by hormonal factors.⁵ Although multiple intraventricular meningiomas are unreported, such tumors have been known to coexist with spinal meningiomas,¹² vestibular schwannomas, and optic nerve sheath meningiomas that accompany NF 2. NF 2 was not present in our case.

According to the Brain Tumor Registry of Japan, 12th edition (1984–2000), tumors types homogeneously enhanced with gadolinium on MR imaging in the third ventricle and in middle age include astrocytoma (13.8%), anaplastic astrocytoma (4.4%), malignant lymphoma (3.6%), ependymoma (2.0%), oligodendroglioma (1.9%), and others (18.3%). These tumors and meningioma should be considered in the differential diagnosis of this case. Tumors in the atrium are supplied by both anterior and posterior choroidal arteries. Third ventricle tumors are supplied by medial posterior choroidal vessels, whereas fourth ventricle tumors are supplied by choroidal branches of the posterior inferior cerebellar artery.^{2,9} Tumor staining was faint in this case. Soluble interleukin-2 receptor level in the cerebrospinal fluid was within normal limits in this case.

Surgical management of intraventricular meningiomas requires careful planning. The strategy should be to reach the blood supply with minimal brain resection, coagulation of the tumor prior to incision, internal decompression, and occlusion of the feeding vessels. Magnification greatly aids in tumor excision. The optic radiation runs lateral and inferior to the atrium. Visual symptoms can be caused by larger tumors in this tract or occur following resection of these tumors through the temporal lobe. Total removal of third ventricle meningiomas can be achieved through the interhemispheric transcallosal, interhemispheric trans-lamina terminalis, or transcortical-transventricular approaches.^{1–3,11,13,15,17,18} The transcortical-transventricular approach requires cortical incision to reach the lateral ventricle. This approach is best used for lesions with a significant intraventricular component within the third ventricle and associated enlarged lateral ventricles. The most frequent complication of this approach is the occurrence of postoperative seizures with a frequency as high as 27%,¹⁷ presumably as a result of the cortical incision which creates an epileptogenic focus. Neurological deficits and memory loss have also been reported following this approach. The interhemispheric-transcallosal approach permits entry to the third ventricle without a cortical incision. Ventriculomegaly is not a prerequisite for this approach. Hemiparesis, mental status changes, memory disturbances, confusion, and mutism have been reported following this approach.

We chose the transcortical-transventricular approach because mild hydrocephalus was found on CT, and the frontal bridging veins were rich and difficult to preserve in the interhemispheric approach. To avoid hypothalamic injury, there was no traction on the tumor. Large tumors

are poor candidates for excision due to the morbidity associated with neuroendocrine and hypothalamic dysfunction, and the possible deep venous system injury associated with surgery of large third ventricle tumors. The tumor was internally decompressed to allow separation from the brainstem and total excision. Hemostasis has to be meticulous to prevent postoperative intraventricular hematoma.

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