Intraventricular meningiomas: a clinicopathological study and review of the literature

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Object. Intraventricular meningiomas are rare tumors. The origin of these tumors can be traced to embryological invagination of arachnoid cells into the choroid plexus. The authors analyzed data that they had collected to study the clinicopathological aspects and review the origin, presentation, imaging, and management of these tumors.

Methods. In this retrospective analysis, the authors describe the cases of 12 patients who had received a diagnosis of intraventricular meningioma and underwent surgery for the tumors. Nine of these patients were men and three were women. Features of neurofibromatosis Type 2 were present in two of the women. Nine of the tumors were located in the lateral ventricles, one was in the third ventricle, and two were in the fourth ventricle. Raised intracranial pressure (ICP) was the universal presentation in all the patients, and the preoperative diagnoses were confirmed on neuroimaging studies. Excision was performed using the parietooccipital (trigonal) approach for lateral ventricle tumors, the transcortical-transventricular route for the third ventricle tumor, and suboccipital craniectomy for fourth ventricle tumors. Postoperatively, one patient died and the others experienced resolution of their symptoms. Histopathological features of these tumors were similar to those seen in meningiomas in other locations.

Conclusions. Intraventricular meningiomas are slow-growing tumors that become large prior to detection. Although they are commonly seen in the lateral ventricles, they occur in the third and fourth ventricles as well. Presentation is in the form of raised ICP with no localizing features; therefore the diagnosis is based on imaging studies. Hydrocephalus occurs due to obstruction of cerebrospinal fluid pathways. Excision requires planning to avoid eloquent cortex incision. The histopathological features are varied, although most of the tumors in the study were angiomatous meningiomas. These tumors are no different histologically from tumors that are dural in origin. No recurrence has been reported.

Key Words • brain tumor • third ventricle • fourth ventricle • lateral ventricle • choroid plexus • intraventricular tumor • meningioma

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 being seen more often in the fourth ventricle and colloid cysts in the third ventricle. Tumors that are most likely to occur in the lateral ventricles are astrocytomas, subependymal giant cell astrocytomas, and ependymomas. Intraventricular meningiomas are rare tumors, accounting for 0.5 to 5% of all intracranial meningiomas. Although there are series in which lateral ventricle meningiomas have been reported, there are only case reports of third and fourth ventricle meningiomas. In this report, we present our experience with 12 cases of intraventricular meningiomas. The relevant literature is briefly reviewed.

Abbreviations used in this paper: CT = computerized tomography; ICP = intracranial pressure; MR = magnetic resonance; NF2 = neurofibromatosis Type 2; PICA = posterior inferior cerebellar artery; VS = vestibular schwannoma.

Clinical Material and Methods

We retrospectively analyzed the records of 12 patients in whom intraventricular meningioma had been diagnosed. These patients underwent treatment during a period of 12 years (1993–2005) in three different service hospitals. There were three women and nine men; all were in their third and fourth decades of life. Presentation, imaging features, surgical procedures performed, histological diagnoses, and postoperative results were analyzed. Only those patients with histologically proven meningiomas were included in the study. Whereas the symptoms in patients with lateral ventricle meningiomas were insidious and protean, the preoperative course was shorter and more acute in those who had third and fourth ventricle tumors. One patient, who had a calcified intraventricular tumor, had experienced headaches for nearly 16 years prior to detection of the tumor. Headache, intermittent or persistent, was the most common symptom and was present in all patients. One of the three women had unilateral proptosis and sensorineural deafness...
along with significant café-au-lait spots, and another woman presented only with sensorineural deafness and headache. Papilledema was observed in eight patients, and unilateral optic atrophy was seen in the woman who had proptosis in her eye. There were no other localizing signs.

Images of all patients were obtained using skull radiography and CT. Four patients also underwent MR imaging. As shown in Figs. 1 through 5, the distribution of tumors was as follows: nine in the lateral ventricle (seven in the left hemisphere and two in the right), one in the third ventricle (in the woman with NF2), and two in the fourth ventricle.

Whereas one lateral ventricle tumor was solid and densely calcified, another was partially cystic. Others were solid and were intensely enhanced after addition of intravenous contrast agents. The intraventricular tumors in the two women who had NF2 were caused by that condition, as were their multiple intracranial tumors. One of these women had a third ventricle meningioma along with an optic nerve sheath meningioma, VS, and two small cerebral convexity meningiomas; the other woman had lateral ventricle meningioma and bilateral VSs.

All patients underwent excision of the intraventricular tumors. Total excision was achieved in all.

**Surgical Procedures**

**Lateral Ventricle Tumors.** All of the tumors were in the atrium (trigone) of the lateral ventricle. They were approached via a transcortical route, through an incision over the parietooccipital cortex to open the ventricle. The capsule was extensively coagulated using bipolar current, and the tumor capsule was emptied in the area of coagulation so that it could be turned to expose its feeding vessels originating from the posterior choroidal artery. These vessels were then coagulated and divided, and the tumor was excised in one piece.

**Third and Fourth Ventricle Tumors.** The only third ventricle tumor was excised in one piece using a transcortical–transventricular route. One of the two fourth ventricle tumors had the PICA draped over it, and this was causing severe brainstem distortion in the patient. The artery was carefully mobilized and protected. The tumor was coagulated and hollowed out so that it could be dissected in the arachnoid plane and separated from the floor of the fourth ventricle. The tumor in the other patient received its vascular supply from the choroid plexus on the left side of fourth ventricle. The tumor was coagulated and divided, then excised piecemeal.

In all patients, the dura mater was closed primarily or by duraplasty in which a pericranium or temporalis fascia graft was used.

**Results**

Results were analyzed for immediate postoperative outcome and long-term follow up as well as to determine the histological variants of the tumor.

**Lateral Ventricle Tumors**

Of nine patients with this type of tumor, one patient died in the postoperative period due to intraventricular hemorrhage, one patient had persistent postoperative homonymous hemianopia, and recovery was uneventful in seven. Two patients had transient speech disturbances following surgery, which resolved within 4 weeks. In one of the two women who had NF2, a retromastoid craniectomy and excision of VS was done prior to excision of the lateral ventricle meningioma.
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**Third Ventricle Tumor**

In the one patient with this type of tumor (one of the two women who had NF2) recovery was difficult due to a postoperative cerebrospinal fluid leak and a case of meningitis that required a prolonged antibiotic regimen. After recovery, the patient underwent a retromastoid craniectomy for excision of VS, and the optic nerve meningioma was removed in a separate surgical procedure.

**Fourth Ventricle Tumor**

One patient had postoperative respiratory dysfunction and oropharyngeal paralysis, probably due to injury to the

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**Fig. 3.** Three Gd-enhanced MR images revealing meningioma in the trigone of the lateral ventricle.

**Fig. 4.** Preoperative CT scans demonstrating third ventricle meningioma as part of NF2 complex, with accompanying optic nerve sheath meningioma and VS.
floor of the fourth ventricle. The patient underwent ventilation therapy for 8 days, and was weaned thereafter. He required a nasogastric feeding tube, and remained dependent on it after discharge. Recovery was uneventful in the other patient.

Pathological Findings

Eleven of the tumors were sent to a pathology laboratory as fragmented tissues, and the other, a calcified intraventricular tumor that had been removed from a lateral ventricle, was sent as a single, rock-hard mass. The fragmented tumors were fixed in formalin and subjected to routine processing. In all cases, H & E–stained sections were used for final diagnosis. Reticulin stains were done in cases of angiomatic meningioma to differentiate them from hemangiopericytoma. The rock-hard completely calcified tumor was cut into small pieces with an oscillating saw and fixed in formalin.

Histopathological findings in the tumors were varied, as seen in Figs. 6 and 7. Of the lateral ventricle tumors, four were angiomatic, three were meningothelial, one was psammomatous/osteoblastic, and one was macrocystic angiomatic. The sole third ventricle tumor was angiomatic. Of the two fourth ventricle tumors, one was fibroblastic and one was meningothelial.

In nine of the patients, tests for epithelial membrane antigen and vimentin were positive. The psammomatous meningioma showed densely packed psammoma bodies in an ossified matrix.

All 11 surviving patients were followed for 3 to 10 years, with no evidence of recurrence. The two patients with NF2 had surgery for associated tumors.

Discussion

The presence of arachnoid cell nests in the normal choroid plexus stroma has been illustrated in the literature, and a thorough examination of the choroid plexus usually reveals small or sometimes larger collections of these cells. The presence of these cells in the choroid plexus has been explained by various investigators. The choroid plexus develops initially from an invagination of a mesenchyme in the thin, roof area of the myelencephalon during the 6th week of gestation. 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. 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 tissue, and blood vessels. Meningiomas arise from arachnoid cap cells, which are specialized cells in arachnoid granulations. In a similar fashion, 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. 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. Fourth ventricle meningiomas characteristically arise from choroids or interior tela choroidea.

Ventricles are rare sites for the occurrence of meningiomas. The incidence of meningiomas of lateral ventricles in adults is variously reported as between 0.5 and 5% of all intracranial meningiomas. For unknown reasons, meningiomas of the lateral ventricles occur more frequently on the left (up to 60% of all lateral ventricle meningiomas) than the right. Because the choroid plexus is more bulky in the lateral ventricles, incidence of lateral ventricle meningiomas is higher compared with those in the third or fourth ventricles. Meningiomas are rare in the third and fourth ventricles; in
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our extensive review of the literature, we found reports of only 83 meningiomas in the third ventricle and 35 meningiomas in the fourth.\textsuperscript{20}

Ventricles of the brain provide space for tumor expansion, and until the cerebrospinal fluid pathways are mechanically occluded, manifestations are mild and nonspecific. Cushing and Eisenhardt\textsuperscript{2} described five clinical features of lateral ventricle tumors: 1) pressure symptoms with ipsilateral headache; 2) contralateral macula splitting homonymous hemianopia; 3) contralateral sensorimotor paresis, more marked sensory involvement, and numbness over trigeminal distribution; 4) cerebellar involvement in more than half of patients; and 5) paralexia, worsened by surgical intervention in left-sided tumors.

Regardless of their locations, most of the clinical symptoms are due to increased IP. Lesions grow into the ventricular cavity until they produce obstructive hydrocephalus. Visual field deficits may be seen in 25\% of patients,\textsuperscript{8} and seizures are rare.\textsuperscript{31} Cerebellar signs may be seen in fourth ventricle tumors, and hypothalamic features with or without endocrinopathy may be seen in third ventricle tumors. Manifestations of NF2 may be evident on clinical and neurological evaluation. Although meningiomas are more common in women, in our 12-patient group only three were women who had intraventricular meningiomas; two of these women had features of NF2. Although intraventricular meningiomas are reported to be common in childhood,\textsuperscript{3,13} there were no children with these tumors referred to our center.

The rarity of intraventricular meningiomas in contrast to those with dural attachment may also be due to a yet unknown factor. Multiple meningiomas arising from the dura mater are regarded as being caused by the inherent multicentricity of the dural foci, possibly influenced by hormonal factors.\textsuperscript{11} Although there are no reports of multiple intraventricular meningiomas, such tumors have been known to coexist with spinal meningiomas,\textsuperscript{22} VSs, and optic nerve sheath meningiomas that may accompany NF2.

Neuroimaging is usually necessary for diagnosis of these tumors. Intraventricular meningiomas may occasionally be densely calcified so as to be visible on plain skull radiographs.\textsuperscript{2} Although CT scans may show calcification, obstructive hydrocephalus, and uniform contrast enhancement, MR imaging is needed for diagnosis of the tumor. The MR images reveal the tumor to be hypo- or isointense on T1-weighted images, and the tumor enhances densely and uniformly after the addition of gadolinium. The MR spectroscopy and blood volume–time intensity maps alone or together may increase the certainty of preoperative diagnosis. A high alanine-to-creatinine ratio has been reported as a specific MR spectroscopic finding for meningiomas.\textsuperscript{17} Fibroblastic meningiomas, the most common type in the lateral ventricles, are hypointense on T2-weighted images.\textsuperscript{19} Macrocystic tumors are rare, and have been reported only twice; 4 both of those tumors and the macrocystic tumor seen in our series were aggressive and malignant. Additional tumors such as those that occur in cases of NF2 are well delineated on MR images. Digital subtraction angiography can be used for assessment of vascularity and to determine the location of the principal feeder vessel.

Lateral ventricle tumors are fed by lateral–posterior choroidal vessels, whereas those in the temporal horn receive their blood supply from the anterior choroidal artery. 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 derive their blood supply from choroidal branches of the PICA.\textsuperscript{14}

Surgical management of intraventricular meningiomas requires careful planning. In cases of tumors involving the dominant lobe, preoperative neuropsychological testing should be performed. Meningiomas are solid, discrete lesions that can be totally excised. 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. A plethora of surgical approaches for meningiomas of the lateral ventricle have been developed to address the risk to visual fields, the left-sided preponderance of the tumors with their potential for speech and cognitive deficits, and the need for early control of the vascular pedicle. These approaches may be made through the cerebral convexity (temporal, parietotemporal, and parietooccipital lobes), which would involve occipital lobectomy, or through the corpus callosum.\textsuperscript{8} Most of these approaches lead to iatrogenic morbidity in the form of visual field deficit, disconnection syndrome, or speech and cognitive deficits. The parietooccipital approach follows a cranio-caudal orientation parallel to the optic radiation over the cerebral convexity and is least likely to damage the optic radiation. Moreover, this is often the thinnest region overlying the trigone and the tumor. Debulking allows the tumor to be turned so that the feeding choroidal vessel can be controlled effectively; early attempts at control may result in undesirable brain retraction and its sequelae. Although early control of the feeding vessel is desirable, that may not be possible until the tumor is debulked and turned to expose the vessel. With this approach, a tumor can be completely excised, and speech and cognitive deficits, if any, are mild and transient. A high cortical incision can be made in the dominant hemisphere to avoid speech disturbances, and frameless stereotaxy can be used to secure a safe trajectory.\textsuperscript{23} Small third ventricle tu-
In general, meningiomas are diagnosed based on morphological features alone. The expression of estrogen receptors is low in these tumors, and two thirds of patients who have such tumors are positive for progesterone receptors. It remains to be seen whether these findings, reported after complete excision, are no unique clinical features on presentation, and raised intracranial pressure. To avoid hypothalamic injury, there should be no traction on the tumor. Large tumors are poor candidates for excision due to the morbidity associated with neuroendocrine, hypothalamic dysfunction and the possible deep venous system injury associated with surgery of large third ventricle tumors. Fourth ventricle tumors are best approached by suboccipital craniectomy, splitting of the vermis, and coagulation of surface vessels, while preserving important vessels like the PICA. The tumor is internally decompressed so that it can be separated from the brainstem and excised totally. Hemostasis has to be meticulous to prevent postoperative intraventricular hematoma.

Intraventricular meningiomas can be any of the histopathological tumor types (predominantly fibrous, fibroblastic, meningothelial, or psammomatous) defined by the World Health Organization classification of meningiomas. Angiomatous tumors were the most common type seen in our series, followed by meningothelial, fibroblastic, and psammomatous/osteoblastic types. The structure of psammoma bodies in the choroid plexus is very similar to that in meningiomas. In general, meningiomas are diagnosed based on morphological features alone. The expression of estrogen receptors is low in these tumors, and two thirds of patients who have such tumors are positive for progesterone receptors. It remains to be seen whether these findings, and cytogenetic features like deletion of chromosome 1 and 14 that occur in meningiomas at other sites, can be applicable to intraventricular meningiomas.

Conclusions

Intraventricular meningiomas are rare tumors, occasionally seen as part of the NF2 complex. Most authors have reported a preponderance of meningiomas in women. There are no unique clinical features on presentation, and raised IP is usually evident at that time. A diagnosis must be established using MR imaging, and surgery requires planning to avoid eloquent area damage. Early control of the vascular supply to the tumor is critically important, and the tumor can usually be removed intact without damage to vital areas around the ventricles. No recurrences have been reported after complete excision. The histology of these tumors is in no way different from that of meningiomas with dural attachment.

References


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