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Case Report

Third ventricular papillary meningioma: a case report and literature review

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Abstract: Papillary meningioma is a rare type of meningioma. PMs that arise in the third ventricle are exceedingly rare. This study reports a 37-year-old male with papillary meningioma presenting with generalized tonic-clonic seizures. Complete surgical resection via a left transcortical-transventricular approach, followed by gamma knife radiosurgery, was performed. Histological findings confirmed the diagnosis of papillary meningioma. Follow-up magnetic resonance imaging, 4 months after surgery, showed no signs of residual tumors or recurrence. Complete tumor resection is the recommended first-line treatment for third ventricular papillary meningioma. Close clinical and radiological follow-ups are required for such patients.

Keywords: Papillary meningioma, brain neoplasms, cerebral ventricle neoplasms, third ventricular

Introduction

Meningiomas are the most common primary central nervous system tumors, accounting for about one-third of all primary tumors of the brain and spinal cord [1]. World Health Organization (WHO) tumor classification system divides meningiomas into three groups. Most meningiomas are benign and correspond to WHO Grade I. Atypical meningiomas are classified as Grade II, while anaplastic (malignant) meningiomas are classified as Grade III meningiomas. Papillary meningioma (PM) is a rare and aggressive variant, classified as a grade III meningioma [2]. Intraventricular meningiomas are also rare tumors, accounting for only 0.5-5% of all meningiomas [3]. This present study reports an experience with an adult male patient with third ventricular PM. A brief review of relevant literature is also presented.

Case report

A 37-year-old male presented with generalized tonic-clonic seizures. He was admitted to the Department of Neurosurgery with no prior history of seizures. Neurological examinations showed no obvious neurological deficits. Brain magnetic resonance imaging (MRI) revealed an irregular intraventricular mass associated with marked peritumoral edema. Compared to the gray matter, the lesion was slightly hypointense on T1-weighted sequence and hyperintense on T2-weighted sequence. The tumor was strongly enhanced after Gd-diethylenetriamine pentaacetic acid (Gd-DPTA) administration (**Figure 1A-C**). Tumor resection was performed using the left transcortical-transventricular approach. The solid tumor mass (5.5 cm × 6.0 cm × 5.5 cm) was grayish-pink in color and had originated from the third ventricle. Gross total resection of the tumor was performed in piecemeal fashion. According to histopathological examination, tumor cells showed a perivascular pseudorosette pattern. Immunohistochemical staining demonstrated that the lesion was positive for progesterone receptor (PR), Vimentin, S-100, and Ki-67, but negative for epithelial membrane antigen (EMA) and glial fibrillary acidic protein (GFAP). Histopathological findings confirmed the diagnosis of PM (**Figure 2**). Postoperatively, the patient developed right limb asthenia, gradually resolving during the postoperative period. The patient was discharged 14 days after surgery. The patient received gamma knife radiosurgery in another hospital immediately after discharge. Follow-up MRIs, performed 4 months after surgery, confirmed

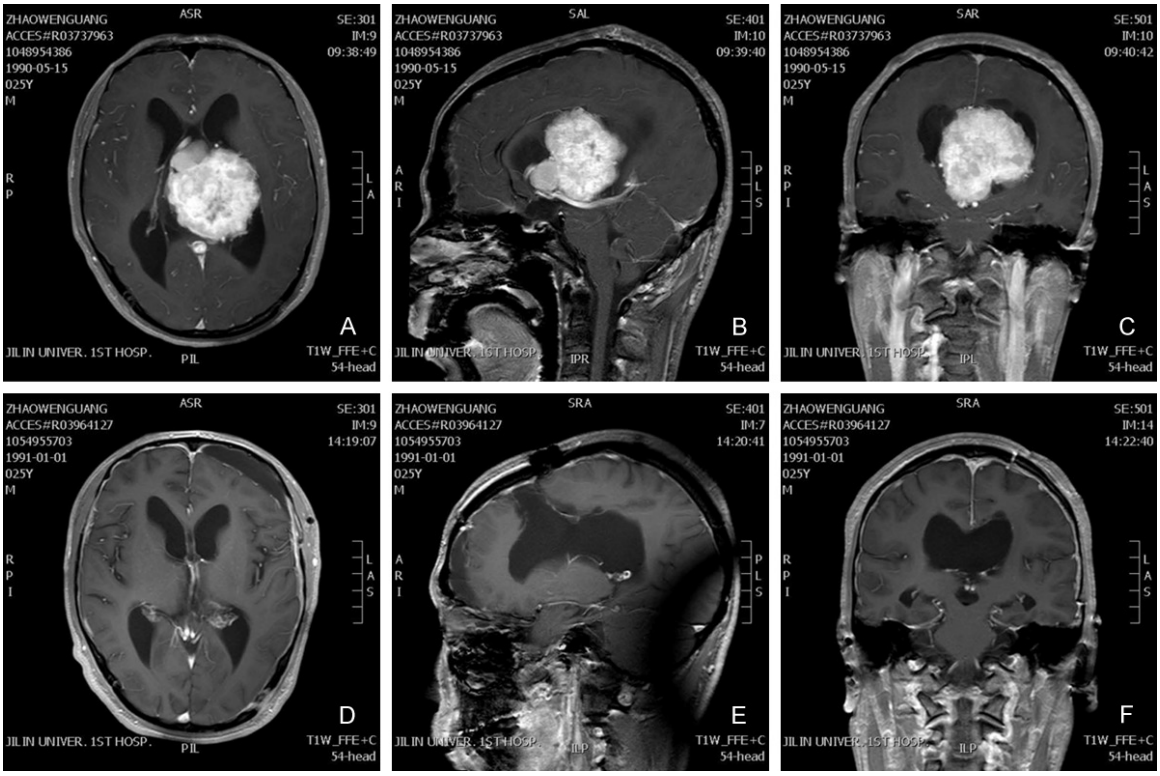

Figure 1. Preoperative and postoperative MRI findings. Preoperative contrast-enhanced brain MRI (Gd-diethylenetriamine pentaacetic acid) showing an enhanced irregular intraventricular lesion (A. Axial view; B. Sagittal view; C. Coronal view). Postoperative MRI at 4 months showing no signs of recurrence (D. Axial view; E. Sagittal view; F. Coronal view).

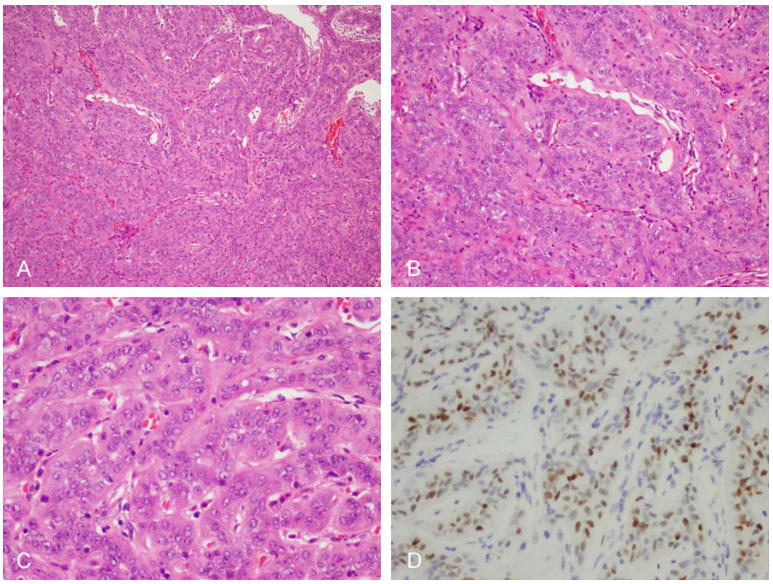

Figure 2. Histopathological examination of surgical specimen. Hematoxylin and eosin stained sections of the resected tumor tissue showing a perivascular pseudorosette arrangement of tumor cells (A. × 10; B. × 20; C. × 40). Positive immunohistochemical staining for progesterone receptor (D. × 40).

complete resection of the tumor with no signs of recurrence (**Figure 1D-F**). Six months of follow-up after surgery was uneventful.

Discussion

Meningiomas are the most common benign intracranial neoplasms, accounting for 13-26% of all primary intracranial tumors [4]. Meningiomas are classified based on dural site of origin, involvement of adjacent tissues, or histological findings. Ventricles are unusual sites for occurrence of meningiomas [3]. Intraventricular meningiomas (IMs) tend to develop in the lateral ventricles, as the choroid plexus is bulkier in the

Table 1. Published case reports of intraventricular papillary meningioma

Authors	Age/Sex	Location	Symptoms	Resection	Follow-up	Status
Regel et al. [9]	17/M	Lateral ventricle	Limb asthenia, headache and nausea	Complete	25 months	Recurrence
Zhi et al. [10]	23/F	Lateral ventricle	Headache and dizziness	N/A	N/A	N/A
Karki et al. [11]	50/M	Third ventricle	Limb asthenia and headache	N/A	23 months	Recurrence

lateral ventricles. However, these can develop anywhere in the ventricular system [3, 5]. Liu et al. reported a series of 25 patients with IMs. Of these, only one patient had IIM located in the fourth ventricle, while the others were in the latter ventricle [6]. In a study by Li et al. [7], ventricular meningiomas and third ventricular meningiomas accounted for 2.1% and 0.11% of all meningiomas, respectively. Ventricles of the brain provide space for tumor expansion, hence manifestations are typically mild and nonspecific. Most IMs are slow-growing tumors, although most are large at the time of detection due to benign symptomatology. MRI is the investigation of choice. Imaging characteristics are typically non-specific. IMs are always hypo- or isointense on T1-weighted sequences and hyper- or isointense on T2-weighted sequences. These typically show strong homogeneous enhancement. Surgical excision of tumors is the preferred approach to treatment. Bhatoe et al. suggested that the surgical strategy should be to reach the blood supply with minimal brain resection, coagulation of the tumor prior to incision, internal decompression, and occlusion of feeding vessels [3].

Papillary meningioma is a rare but well-recognized histological variant of meningioma. It is characterized by the presence of a perivascular pseudopapillary pattern [2]. PMs tend to occur in young patients. Brain invasion is always noteworthy and the recurrence rate is relatively high. To the best of our knowledge, this case was the fourth report of intraventricular PM and the second report of a third ventricular PM. **Table 1** lists previously published reports. All previously reported patients with intraventricular PM have developed recurrence at least once. It is believed that the clinical aggressiveness of PM is the first reason for the high rate of recurrence. The second reason is that it is hard to achieve Simpson grade I or II resection of intraventricular PMs. Some residual tumor cells in the ventricles are liable to disseminate via the cerebrospinal fluid. Eom et al. reported a patient with intraventricular rhabdoid papillary meningioma, in whom diffuse craniospinal leptomeningeal metastatic lesions were detected five years after surgery [8].

In the present case, the tumor was very large and irregular. Based on MRIs, meningioma was the first preoperative diagnosis, but glioma could not be excluded. The surgical approach to resection of a third ventricular meningioma is guided by the site of tumor, its relation to anatomically contiguous structures, patient conditions, and individual surgeon preferences [7]. This patient was given the left transcortical-transventricular approach for complete resection. Achievement of total resection is typically challenging with a right-sided approach but is relatively safe. Surgery and the follow-up period were uneventful. Follow-up of 6 months is not enough, however. Based on the pathological diagnosis of PM, further follow-ups are needed.

In conclusion, intraventricular PM is rare type of meningioma. This study reports a third ventricular PM in a male patient, presenting with generalized tonic-clonic seizures. Diagnosis of PM was confirmed through histopathological examination. Complete tumor resection was performed, as it is the recommended first-line treatment. Close clinical and radiological follow-ups are required for these patients.

Disclosure of conflict of interest

None.

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