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Case Report

Third ventricle World Health Organization Grade II meningioma presenting with intraventricular hemorrhage and obstructive hydrocephalus: A case report and literature review

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ABSTRACT

Background: Third ventricular meningiomas are exceedingly rare intracranial tumors that may present with intraventricular hemorrhage.

Case Description: The patient is 46-year-old who initially presented with obstructive hydrocephalus from a presumed vascular lesion and who was treated with endoscopic third ventriculostomy. He presented 3 years later with acute intraventricular hemorrhage and hydrocephalus. The hemorrhage was evacuated and the third ventricular tumor was resected, and the patient made an excellent recovery. Histopathological analysis identified the tumor as the World Health Organization Grade II meningioma.

Conclusion: Third ventricular meningioma is a rare tumor that may present with hemorrhage and obstructive hydrocephalus. Surgical resection can be helpful for this rare presentation of intracranial meningioma.

Keywords: Endoscopic third ventriculostomy, intraventricular hemorrhage, intraventricular meningioma, obstructive hydrocephalus, World Health Organization grade II meningioma

INTRODUCTION

Intraventricular meningiomas are rare primary brain tumors, representing only 0.5–5% of all meningiomas.[2] Third ventricle meningiomas (TVMs) are even less common, accounting for just 0.1–0.18% of all intracranial meningiomas.[1,21] In a recent clinical series,[18] the majority of TVMs were categorized histopathologically as the World Health Organization (WHO) Grade I tumors, with only 15.6% of cases classified as the WHO Grade II. Clinically, TVM typically presents with obstructive hydrocephalus, ataxia, and hypothalamic dysfunction, along with other symptoms of elevated intracranial pressure.[14,16,18] It is exceptionally uncommon for TVMs to present with intraventricular hemorrhage (IVH), as only one case

has been reported with this presentation.[15] We present a case of a 46-year-old male with WHO Grade II TVM who presented initially with acute obstructive hydrocephalus and later represented again with acute IVH with obstructive hydrocephalus.

CASE REPORT

The patient is 46-year-old with a past medical history significant for diverticulosis who initially presented with acute obstructive hydrocephalus requiring placement of an external ventricular drain (EVD). Magnetic resonance imaging (MRI) at the time revealed a small enhancing lesion in the posterior third ventricle at the cerebral aqueduct causing obstruction [Figure 1]. An endoscopic third ventriculostomy (ETV) was performed to treat the hydrocephalus. However, on directly visualizing the lesion during endoscopy, it was decided to not perform a biopsy of the lesion since it appeared highly vascular and was considered to possibly represent a vascular malformation. No direct bleeding from the lesion was observed at the time of initial endoscopy. Cytology of the cerebrospinal fluid was obtained but assessed as negative for neoplastic cells. Diagnostic cerebral angiogram performed at a later date and did not reveal any vascular abnormalities (i.e., no aneurysm or vascular malformations were identified). Three years after the initial presentation, the patient experienced acute onset of nausea and headache, and he was found to have new obstructive hydrocephalus secondary to IVH imaged on computed tomography head and MRI [Figures 2 and 3]. An EVD was placed emergently, and the patient was taken to the operating room for an elective interhemispheric transcallosal approach to evacuate the IVH and definitively resect the posterior TVM with intraoperative image guidance [Figure 4]. Final pathology of the mass revealed it to be the WHO Grade II meningioma [Figure 5]. The patient was discharged home 9 days after surgery. At 6-month follow-up, he is doing well with no need for additional treatment of hydrocephalus.

To put this case into the appropriate context of the available literature, we performed a systematic literature search and analyzed comparable cases.

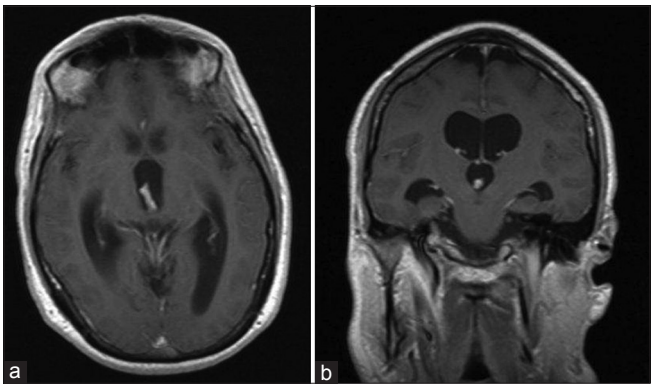

Figure 1: Magnetic resonance imaging from initial presentation with hydrocephalus (HCP). (a) T1-weighted image (T1WI) with contrast axial view. Demonstrates uniformly enhancing lesion within the posterior aspect of the third ventricle obstructing aqueduct. This results in triventricular HCP. (b) T1WI with contrast coronal views. Demonstrates uniformly enhancing mass within the cortical aspect of the third ventricle blocking the aqueduct. This results in triventricular HCP. No transependymal flow was noted on FLAIR sequence, suggesting HCP to be longstanding.

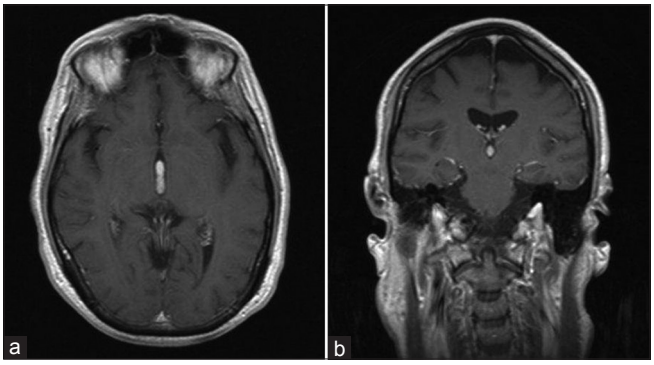

Figure 2: Last magnetic resonance imaging (MRI) before admission. (a) T1WI with contrast axial view (similar cut as initial MRI). Significant improvement in obstructive HCP following successful ETV. The uniformly enhancing mass within the third ventricle is stable in size compared to prior MRI. (b) T1WI with contrast coronal view (similar cut as initial MRI). The uniformly enhancing mass within the third ventricle appears to be stable in size, and spans almost the entire length of the third ventricle.

Methods

A PubMed search up to December 2018 using the key phrase "TVMs" returned 137 papers. From this initial search, we identified individual papers that included new cases of TVM and case series of intraventricular meningiomas that had TVM reports within them. This allowed us to identify 109 cases who had been reported in the literature. However, after carefully reviewing all of the identified records, we noticed that some reviews and case series had inadvertently duplicated prior reports when determining the total amount of cases of TVM in the literature. As a result, we cross-referenced all of the identified records and did not include reports based on the following exclusion criteria: if the case had been previously reported literature or if the TVM was not primary and had originated from a lateral ventricle or elsewhere. Doing this, we identified 60 unique cases of primary TVM in the literature (Figure 6 for the PRISMA-guided study flow diagram).

We also wished to investigate and quantify the surgical approach across these studies and correlate them with outcomes. We found that the outcomes across the records were highly variable in how they were reported. As a result, we pooled together outcomes that were described as "good" - describing a range from simple survival to independent functioning - into an

"optimal" category. Similarly, we pooled together outcomes described as "bad" - describing a range from death to dependent functioning - into a "suboptimal" category. We then constructed comparative tables detailing the outcomes. We additionally collected data across all 60 cases describing the patient characteristics and symptomatology of TVM.

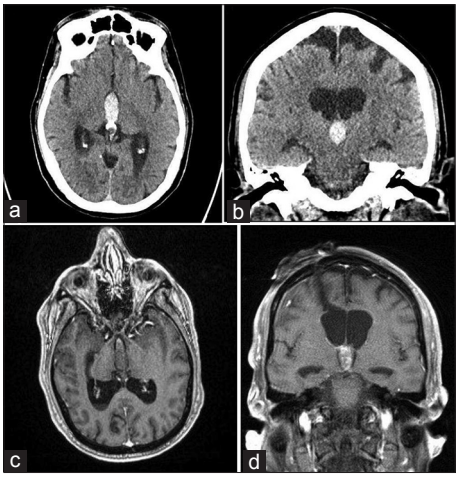

Figure 3: (a) CTH and magnetic resonance imaging during this admission. (a) Axial noncontrast head computed tomography (CT). IVH is noted within the third ventricle and aqueduct. Triventricular obstructive HCP is also noted. (b) Coronal non-contrast CTH. IVH is noted within the third ventricle which results in obstructive triventricular HCP. (c) T1WI with contrast axial view. IVH is noted within the third ventricle and occipital horns with resultant triventricular obstructive HCP. Contrast-enhancing lesion is noted within the anterior superior aspect. (d) T1WI with contrast coronal view. IVH is noted within the third ventricle with resultant triventricular obstructive HCP. The contrast-enhancing lesion is noted within the anterior superior aspect of the third ventricle.

RESULTS

The patient characteristics and symptomatology of TVMs were evaluated across the 60 reviewed cases. 37 patients were male and 21 were female (1.76 male/1.0 female ratio), and in two cases, the sex was not reported. The average presenting age was 30.1 years old (Standard deviation=±18.4). The most common presenting symptoms were headache in 29 cases (51.8%), followed by gait ataxia in 15 cases (26.8%) and visual changes or disturbance in 10 cases (26.8%). Hypothalamic changes, including the development of diabetes insipidus, were a symptom in only 4 cases (7.2%). Various other neurologically related symptoms were also found and are detailed in Table 1.

Surgical approaches for the treatment of TVMs were also investigated, along with the associated resection extent and outcomes for each technique. The frequency of use, resection extent (gross total vs. subtotal), and associated outcomes for each surgical approach of TVM are described and reported in Table 2. Overall, a transcallosal approach was the most commonly utilized, general approach (39.3%). Among the specific approaches, the transcallosal interforniceal approach was the most often used (14.8%). This approach was also the most consistently successful, with a 100% (9/9) positive outcome success rate and a gross total resection extent rate of 67% (6/9 cases). Other specific transcallosal approaches were used with various frequencies and success rates, including transcallosal transventricular transchoroidal (8.2%, 4/5 success rate), transcallosal unspecified (8.2%, 2/3 success rate), posterior transcallosal (3.2%, 1/2 success rate), bifrontal transcallosal (3.2%, 1/2 success rate), and interhemispheric transcallosal transchoroidal (current case, 1.6%, 1/1 success rate). The resection extents for each of these approaches are also detailed in Table 2.

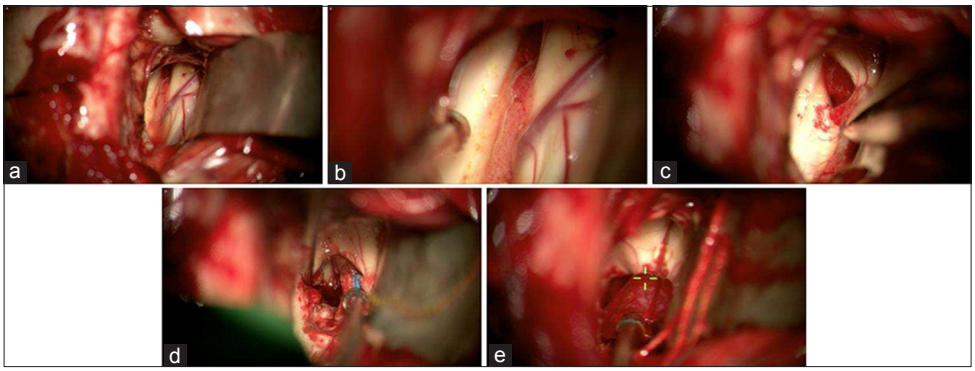

Figure 4: Intraoperative pictures of right sided interhemispheric transcallosal approach to the third ventricular mass. (a) Zoomed out view of the left lateral ventricle with foramen of Monro at 12 o' clock, anterior septal vein at 3 o' clock, and choroid plexus at the center of the picture. (b) zoomed in view of the left lateral ventricle with better visualization of the foramen of Monro, anterior septal vein, and choroid plexus. The red/purplish tumor is starting to come into view within the foramen of Monro. (c) The third ventricular tumor is better seen within the foramen of Monro. It appeared to be more red and vascular, as supposed to the IVH clot which looked dark purple. (d) The tumor is better visualized at the 1 o'clock position. (e) The tumor being removed. After the tumor was removed, the third ventricle was explored, and the IVH clot was noted to be more posterior. It was then suctioned out.

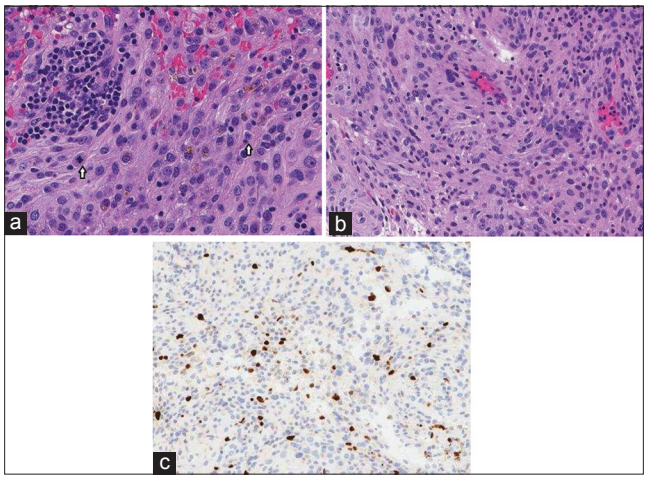

Figure 5: Histopathology of resected tumor specimen concluded to be a WHO Grade II meningioma. (a) The neoplasm is composed of pleomorphic cells with irregular nuclei and prominent nucleoli. Hemosiderin pigment is present in the center of the field, indicating prior hemorrhage. Two mitoses (white arrows) are present in this high magnification photomicrograph. (H and E, ×300). (b) The tumor does not have the morphology of a low-grade meningioma, but there is an attempt at whorl formation in the bottom left in this field. (H and E, ×250). (c) The Ki67 (Mib-1) nuclear labeling index is significantly higher that would be expected in low grade meningiomas. (Ki67 immunohistochemistry, ×125).

Table 1: Pooled patient characteristics and symptomatology from all documented studies of third ventricle meningioma.

Patient characteristic	*n* (%)
Male:female ratio	37:21
Average age (SD)	30.1 (±18.4)
Symptom:	
Headache	29 (51.8)
Gait ataxia	15 (26.8)
Visual changes/disturbance	10 (17.9)
Mental changes - unspecified	8 (14.3)
Somnolence	7 (12.5)
Raised ICP - not specified	7 (12.5)
Hemiparesis	6 (10.7)
Nausea/vomiting	6 (10.7)
Vertical gaze palsy	5 (8.9)
Hypothalamic disturbance	4 (7.2)
memory deficits	3 (5.4)
Urinary incontinence	3 (5.4)
Hearing loss/changes	3 (5.4)
Endocrine dysfunction	2 (3.6)
Mild dementia	2 (3.6)
Personality change	2 (3.6)
Meningismus	1 (1.8)
Dysgraphia	1 (1.8)
Drop attacks	1 (1.8)
Developmental delay	1 (1.8)
Vertigo	1 (1.8)

SD: Standard deviation

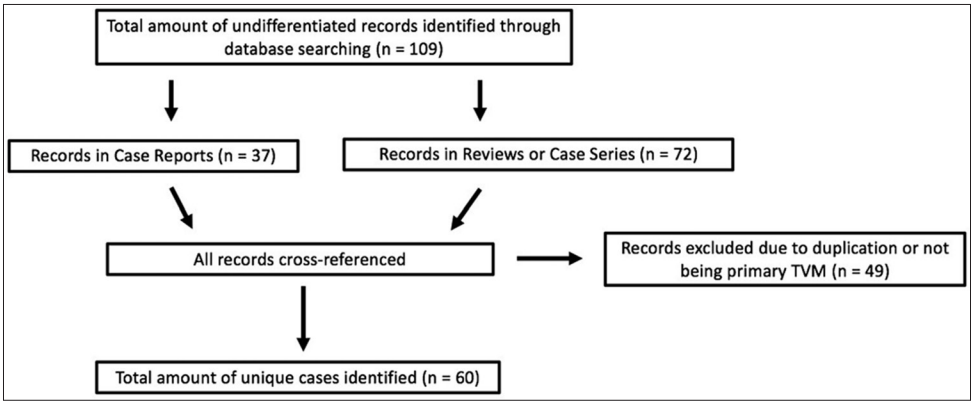

Figure 6: Preferred reporting items for systemic reviews and meta-analysis-guided study diagram.

Transventricular approaches were the second most common surgical approach employed (24.6% of cases). The transventricular transforaminal approach was the most common specific transventricular technique used (9.8%, 4/6 success rate). A transventricular, unspecified approach (9.8%, 4/6 success rate) was also reported in 6 cases. This was followed by the transcortical transventricular approach (4.9%, 3/3 success rate).

Several other approaches were also utilized and described within the literature with varying frequency and success rates, including the supracerebellar infratentorial approach (8.2%, 5/5 success rate) and occipital transtentorial approach (6.6%, 2/4 success rate), among other approaches that are described in Table 2. In 14.8% of cases (9/61), the surgical approach was not described. Total pooled analysis also showed that most patients had a good outcome with surgery (65.6%, 40/61 cases).

DISCUSSION

TVMs are rare tumors. To date, 60 unique cases have been reported in the literature. Most cases are reported in isolated

Table 2: Comparative table of resection extent and outcomes for different surgical approaches of third ventricle meningioma. These data were pooled and collected from all previously published cases of TVM.[1,4-10,14,16-18,23]

Surgical approach		Resection extent (*n*)			Outcome (*n*)		
	Frequency (*n*)	Subtotal	Gross total	Unspecified	Suboptimal	Optimal	Unspecified
Transcallosal interforniceal	9	3	6	0	0	9	0
Transcallosal transventricular, transchoroidal	5	2	3	0	1	4	0
Transcallosal, unspecified	5	1	2	2	1	2	2
Posterior transcallosal	2	0	1	1	1	1	0
Bifrontal transcallosal	2	0	1	1	0	1	1
Interhemispheric transcallosal transchoroidal (present case)	1	0	1	0	0	1	0
Transcallosal total	24	6	14	4	3	18	3
Transventricular, unspecified	6	0	0	6	2	4	0
Transventricular transforaminal	6	1	5	0	2	4	0
Transcortical transventricular	3	0	3	0	0	3	0
Transventricular total	15	1	8	6	4	11	0
Supracerebellar infratentorial	5	1	4	0	0	5	0
Occipital transtentorial	4	1	3	0	2	2	0
Right parieto-occipital	2	0	0	2	0	0	0
Occipital approach, unspecified	1	0	1	0	0	1	0
Posterior parietal	1	0	0	1	0	0	1
Unspecified	9	0	0	9	4	4	1
Pooled total	61	9	29	22	13	40	4

case reports or case series describing all types of intraventricular meningiomas.[1,4,5,7,9,14,16-18,23] TVM most commonly not only presents with symptoms of HCP but can also present with ataxia, hemiparesis, and hypothalamic dysfunction. Our patient presented with acute IVH and obstructive HCP. There is only one other reported patient with TVM presenting in this way,[15] and there are no documented cases of Grade II TVM with this presentation. In one pathological series, only five total cases of Grade II TVMs were found on a literature review, none of which presented with IVH.[14]

The third ventricle is an abnormal location for meningiomas to be found because the third ventricle lacks a dural attachment. TVMs are postulated to arise from the tela choroidea of the velum interpositum.[2] This forms a component of the roof of the third ventricle, which also houses the internal cerebral veins and the medial posterior choroidal arteries.[1,17] These meningiomas are thought to develop from arachnoid cells within the choroid plexus, specifically meningothelial inclusion bodies of the tela choroidea.[2,22] The vascular supply of a TVM is the most often derived from the medial posterior choroidal artery. However, the involvement of the lateral posterior and anterior choroidal arteries has also been observed.[2,8,12] Hemorrhage of TVMs is thought to be infrequent since the blood supply is derived from these small, distensible choroidal branches,[17] which might explain why these tumors rarely present this way. It is also very uncommon for lateral intraventricular meningiomas to present with hemorrhage, with only 11 cases reported in other over 600 cases in the literature of lateral ventricle meningiomas.[19,20] While the exact mechanism of hemorrhage in these lateral ventricle meningiomas is unknown, several theories have been proposed, which may help to provide an explanation of our third ventricle case. These mechanisms include abnormal development of the vascular network within the meningioma,[11] internal necrosis due to rapid growth resulting in venous thrombosis and hemorrhage,[10] and disruption of the vascular blood supply due to tortuous and over-dilated arteries that lose their ability to regulate blood pressure.[3]

On histopathological analysis, the resected tumor consisted of sheets of pleomorphic cells with prominent nucleoli and frequent mitoses. Structures typically associated with meningiomas (e.g., cellular whorls, psammoma bodies, and nuclear pseudoinclusions) were absent. The mitotic rate exceeded 4 per 10 high power fields, thus designating it as the WHO Grade II meningioma [Figure 6]. To our knowledge, the patient is the second published TVM with IVH and the first reported WHO Grade II TVM presenting in this way. Overall, studies have demonstrated that the WHO Grade I is the most common histopathological diagnosis of TVM with an incidence ranging from 50 to 84.6%.[14,16] The WHO Grades II and III are more uncommon, with incidences between 15.6–25% and 0–25%, respectively.[14,16] In these studies, the Grade II histopathological subtypes were either choroidal, fibrous, or atypical. Our patient harbored an atypical TVM.

On literature review, the surgical approaches and management of TVM were investigated. We found that the transcallosal approach was the most commonly utilized approach across all 61 identified cases, including the present case. Interestingly, the transcallosal interforniceal approach was the most consistently successful and also the most commonly used overall [Table 2]. Several anatomical studies have detailed this technique and found it the optimal surgical approach for mass-occupying lesions of

the third ventricle since it facilitates avoiding damage to fibers of the hippocampal and anterior commissure.[24] It has also been shown that this approach can help reduce surgical complications when treating masses in the third ventricle.[13] Overall, surgery is successful with the majority of patients making a good recovery; we found that 65.5% of cases resulted in a good outcome [Table 2], suggesting that surgical intervention is helpful for treatment of this rare tumor.

Our patient presented with an initial episode of symptomatic HCP 3 years before the second hemorrhagic event. He was initially treated with ETV, but we refrained from a biopsy because the tumor appeared to be highly vascular and the patient was clinically nonfocal (other than the hydrocephalic presentation). There are also no previously documented instances of TVM presenting with hemorrhage after being monitored for years with serial imaging, which lead us to follow the lesion expectantly instead of resecting it. Ultimately, our patient underwent interhemispheric transcallosal transchoroidal approach with resultant gross total resection of the tumor after presenting with IVH. We chose this approach because the fornices were adherent to each other in our patient, making the interforniceal approach challenging. Our patient presented with IVH after several years of stable imaging. Surgery was not pursued earlier because of the risks associated with surgical tumor resection in a mostly neurologically intact patient. We would recommend serial imaging follow-up at 3 months, 6 months, and annually. Patients should be informed that hemorrhage is possible even if tumor remains stable in size. Without tissue diagnosis, we do not recommend stereotactic radiosurgery as the primary treatment modality.

CONCLUSION

TVM is an uncommon neurologic tumor. It most commonly presents with symptoms of increased intracranial pressure, ataxia, hemiparesis, or visual disturbance. Infrequently, it may present with IVH. Surgical management can be successfully achieved with a transcallosal approach.

Disclosures

None.

IRB: Per Dartmouth-Hitchcock Medical Center, IRB was not required for single patient case report.

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Section of Neurological Surgery, Department of Surgery, Dartmouth-Hitchcock Medical Center, Lebanon, New Hampshire.

Conflicts of interest

There are no conflicts of interest.

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