

Evaluation and Management of Vermian Tumors

Ellina Hattar, MD¹, Tyler D. Alexander, MS¹, Thiago S. Montenegro, MD¹, Glenn A. Gonzalez, MD¹, Kevin Judy, MD¹, James S. Harrop, MD¹

¹Department of Neurosurgery, Thomas Jefferson University and Jefferson Hospital for Neuroscience, Philadelphia, Pennsylvania

Epidemiology From a technical standpoint, this technique allows for the selective ablation of periventricular lesions not favorable to an open approach⁽⁵⁾. Vermian tumors are rare and differ in type based on patient factors. In adults, they are most commonly due to metastasis.¹

H&P A 64-year-old male with a history of lung nodule presented with six months of progressively imbalance and dizziness and a neurologic exam significant for severe truncal ataxia.

Imaging Imaging revealed a 3.5 cm enhancing tumor of the cerebellar vermis effacing the fourth ventricle without hydrocephalus.

Procedure Two approaches can be used to access vermian lesions: the telovelar (TL) and transvermian (TR).²⁻⁴ The TL approach makes use of natural corridors through the cerebellomedullary fissure, while the TR approach is employed for tumors extending to the rostral fourth ventricle.^{5,6} In this case, the TR approach was used as the tumor extended into the rostral fourth ventricle and approached the vermian surface.

Surgical Outcomes and Follow-up Postoperative imaging demonstrated a complete resection. Pathology was consistent with metastatic carcinoma of lung origin. Postoperatively, the patient's dizziness and imbalance improved over the course of several weeks. **Keywords** cerebellar vermis, vermian tumor, telovelar, transvermian

SUBMITTED January 06, 2021. **ACCEPTED** January 21, 2021.

Transcript

SLIDE 1 [TITLE]: 0:00-0:07

Today, we will be discussing the evaluation and management of vermian tumors.

SLIDE 2: 0:07-0:25

Briefly, we will explore strategies for the evaluation and management of vermian tumors through an interesting case presentation. We will then explore relevant differential diagnoses, surgical approaches and associated anatomy, and will conclude the presentation through a brief summary of the patient's postoperative course.

SLIDE 3: 0:25-1:44

A 64-year-old man with a history of emphysema and associated malignant pleural effusions presented to our facility with worsening imbalance and dizziness that had been progressive for approximately six months. He experienced multiple recurrent pleural effusions and was found to have a lung lesion that was highly concerning for malignancy. He underwent a thoracentesis the year prior to presentation, but the fluid failed identify any evidence of malignancy. The patient was also then offered resection of the lung lesion however he elected to proceed with radiation instead. His social history was significant for tobacco use. On exam the patient had no cranial nerves deficits, was awake, able to answer questions readily. His motor and sensory functioning was intact without any evidence of limb ataxia. Upon gait examination, he had severe imbalance and truncal ataxia. Imaging and specifically MRI with and without contrast revealed a sizeable mass that was centered around the cerebellar vermis. The T1 post-gadolinium MRI axial, coronal and sagittal images shown here demonstrate that the mass was effacing but not fully occluding the fourth ventricle. Supratentorially, there was no evidence of

hydrocephalus, as shown in the last image, an axial T2 flair study.

SLIDE 4: 1:44-3:08

In this case, we must consider the most common differential diagnoses. One can group these based on their relation to the ventricular system, a strategy adapted from Anne Osborne's book titled "Diagnostic Neuroradiology". In the adult the most likely tumors surrounding the fourth ventricle are metastatic lesions. Hemangioblastoma, a highly vascular tumor often associated with a cyst, is also a possibility. In many of these cases, patients may have a positive family history for Von Hippel Lindau Syndrome. In younger adults, medulloblastoma can present in this region and is usually found to originate in the roof of the fourth ventricle. These can be either solid or cystic. An ependymoma can also occur in this location and usually originates in the floor of the fourth ventricle and subsequently spreads through the foramina of Luschka and Magendie. This tumor is often likened to toothpaste spreading through the cerebrospinal fluid apertures. Other differential diagnoses include dermoid and epidermoid tumors, choroid plexus papillomas – which interestingly in adults are most commonly found in the fourth ventricle, whereas in children and neonates are typically found in the atrium of the lateral ventricles. Subependymoma, and finally brainstem or cerebellar gliomas can also be found here, the latter being exceptionally rare in this location. In the case of children, medulloblastoma, pilocytic astrocytoma, and ependymoma are the most common tumors in this region.

SLIDE 5: 3:08-4:17

Following the performance of a suboccipital craniotomy there are two principal approaches that can be used to access vermian lesions. The first is the telovelar approach. This makes use of natural planes.

The cerebromedullar fissure and uvulotonsillar spaces are exposed. This approach is well suited for lesions of the foramen of Luschka which can be reached laterally. A C1 laminectomy can be used to enhance rostral exposure by facilitating a greater inferior to superior working angle. A second approach that can be used to access vermian lesions is the transvermian approach. This approach is typically used for large lesions that occupy the rostral 4th ventricle and it is not well suited for lesions extending laterally into the lateral recess. A portion of the vermis of the cerebellum must be divided to perform this approach and patients undergoing this surgery are at risk for vermian split symptoms, which can manifest with nystagmus, gait dysfunction, oscillation of the head and neck, and cerebellar mutism. Cerebellar mutism is more likely to be seen in children however it can rarely present in adults, as well.

SLIDE 6: 4:17-4:45

One must keep in mind the location of nuclei of the cerebellum when performing either one of these approaches. In this diagram, the dorsal surface of the cerebellum has been removed, including its central vermis. The cerebellar nuclei are a group of four paired nuclei and include the emboliform, the dentate, the globose, and the fastigial nuclei. A classic mnemonic often used to remember these is “Don't Eat Greasy Foods” which corresponds to the cerebellar nuclei in lateral to medial order.

SLIDE 7: 4:45-7:14

We have created this helpful diagram to highlight the rich anatomy of the fourth ventricular floor. The fourth ventricular floor is rhomboid in shape. Its rostral two thirds are located just posterior to the pons while its caudal third is posterior to the medulla. The floor can also be thought of in relation to its cerebrospinal fluid inflow and outflow

connections which can be inferred in this diagram. At the rostral apex of the floor is the cerebral aqueduct. At its caudal tip is the obex, just anterior to the foramen of Magendie. The lateral angles of the floor lead to the lateral recesses and ultimately the foramina of Luschka in the cerebellopontine angles. The lateral recesses are horizontally connected by the striae medullaris. The floor of the fourth ventricle is vertically divided at its midline by the median sulcus with each resultant half further divided by the sulcus limitans. The sulcus limitans separates the median eminence medially from the vestibular area laterally. The sulcus limitans deepens just lateral to the facial colliculus and the hypoglossal triangles to form dimples called the superior and inferior fovea. In the location of the superior fovea the median eminence forms an elongated protrusion, the facial colliculus which overlies the nucleus of the abducens nerve and an ascending portion of the facial nerve. The locus ceruleus is located at the rostral end of the sulcus limitans. This structure has a bluish hue due to the presence of pigmented neurons that contain neuromelanin. These are also responsible for a majority of norepinephrine production in the brain. More inferiorly, the hypoglossal triangle, vagal triangle, and area postrema can be found. Interestingly this group of structures forms a shape reminiscent of the nib on an ink pen and are also referred to as the calamus scriptorius. The hypoglossal and vagal triangles overly nuclei of the same name. The area postrema, when stimulated, can lead to nausea and vomiting. Patients undergoing surgery in this area may therefore experience a self-limited course of postoperative nausea and emesis for which the use of anti-emetic medications is especially efficacious. Not shown here is the auditory tubercle which is a protrusion of the vestibular area and overlies the dorsal cochlear nucleus and the cochlear part of the vestibulocochlear nerve. Also not shown here

is the posterior inferior cerebellar artery which emerges under the cerebellar tonsils. Given the rich functional anatomy of the floor of the fourth ventricle care must be taken to avoid damage to these important structures as excessive pressure or bipolar cautery, for example, may lead to postoperative deficits.

SLIDE 8: 7:14-8:35

In this case, we elected to use the transvermian approach as the tumor was found in the more rostral aspects of the fourth ventricle. It appeared to be extending to the vermian surface and did not extend to the lateral recesses. On postoperative imaging a gross total resection was achieved as demonstrated in this post-gadolinium MRI. In these first 3 images, corresponding to T1 post-gadolinium coronal, sagittal and axial sections, there was no evidence of nodular enhancement. There was some hyperintense signal within the resection cavity due to a small amount of residual blood products – as the cavity was lined with surgicel to enhance hemostasis following surgery. The limited amount of postoperative blood products is confirmed when we compare the T1 post-gadolinium axial image to a T1 pre-gadolinium image showing a hyperintense signal in the operative bed in both studies. The patient's ultimate pathology revealed the

diagnosis of metastatic carcinoma with lung origin. Postoperatively, the patient was found to be at his baseline exam without any evidence of cranial nerve deficits, nystagmus or any changes with his ataxia. His dizziness and imbalance gradually improved following his participation in a short rehabilitation program at an inpatient facility after which the patient was able to be discharged home. He is currently doing well and is awaiting postoperative stereotactic radiation.

SLIDE 9: 8:35-8:56

In conclusion, the differential diagnosis for a vermian mass in the adult patient includes metastasis, hemangioblastoma and medulloblastoma. Both the telovelar and the transvermian approaches may be employed to access tumors of the fourth ventricle. A strong understanding of the structures in the floor of the fourth ventricle is necessary for the safe resection of these tumors.

SLIDE 10: 8:56-9:05

These are helpful references used in the making of this presentation.

SLIDE 11: 9:05-9:07

Thank you for your time.

References

1. Osborn A. Diagnostic Neuroradiology. Mosby; 1994.
2. Ebrahim KS, Toubar AF. Telovelar approach versus transvermian approach in management of fourth ventricular tumors. Egypt J Neurosurg. 2019;34(1):10. doi:10.1186/s41984-019-0036-9
3. Maloumeh EN, Khoshnoud RJ, Ebrahimzadeh K, et al. Surgical management of the fourth ventricular tumors using telovelar approach and the role of neuroendoscopy: Post-operative outcome and long-term results in a series of 52 cases. Clin Neurol Neurosurg. 2021;201:106419. doi:https://doi.org/10.1016/j.clineuro.2020.106419
4. Tomasello F, Conti A, Cardali S, La Torre D, Angileri FF. Telovelar Approach to Fourth Ventricle Tumors: Highlights and Limitations. World Neurosurg. 2015;83(6):1141-1147. doi:https://doi.org/10.1016/j.wneu.2015.01.039
5. Ghali MGZ. Telovelar surgical approach. Neurosurg Rev. Published online 2019. doi:10.1007/s10143-019-01190-5
6. Ferguson SD, Levine NB, Suki D, et al. The surgical treatment of tumors of the fourth ventricle: a single-institution experience. J Neurosurg JNS. 2017;128(2):339-351. doi:10.3171/2016.11.JNS161167

Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: all authors. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Senior author.

Correspondence

Ellina Hattar: Thomas Jefferson University Hospital, Philadelphia, PA. Ellina.Hattar@jefferson.edu.