

An early diagnosed medulloblastoma of the vermis

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Abstract

Introduction: Medulloblastoma is categorized into different histological and molecular subtypes with unique characteristics that make diagnosis challenging. We present a case of a 30-year-old male with continuous nausea and vomiting for 3 months ago. Imaging revealed a brief signal change in the vermis. He had normal results of neurologic examinations. The lesion was completely removed surgically. The histopathology exam revealed classic medulloblastoma, non-SHH, and non-WNT, and the patient was referred for adjuvant therapy. In this case, the low intensity of changes confined to the vermis, without edema and gadolinium enhancement ruled out medulloblastoma. The WNT was a more probable subtype, but histopathology ruled that out. It seems that the decision to operate on this patient was the best approach because this surgery sped up the diagnosis and his chemoradiotherapy started sooner. In conclusion, in the case of lesions in the posterior fossa, medulloblastoma should always be kept in mind because an early diagnosis can improve the prognosis.

Keywords: medulloblastoma, imaging, vermis, non-WNT, non-SHH

Introduction

Medulloblastoma is the most common malignant pediatric brain tumor [1] that arises from progenitor cells [2, 3] and mostly occurs in children but also can be seen in adults [4]. Medulloblastoma has five histologic subtypes [5] and four molecular subclasses [6] with different genetic and demographic properties, clinical symptoms, and prognosis. There are major differences between medulloblastoma in adults and children. In adults, the average age of diagnosis is 29 years [7], and the most common location of tumor is in the cerebellar hemispheres because of the predominance of Sonic hedgehog (SHH) subtype in this population [8]. The Wingless-related (WNT) subtype accounts for 15% of adult cases; patients with tumors situated mostly in the midline have the best prognosis [9].

The gadolinium enhancement with different patterns and degrees is seen in 85 to 100% of patients,

and peri-tumoral edema is present in 50 to 90% of medulloblastoma cases [10]. A common presenting symptom in medulloblastoma is headache and nausea/vomiting because of tumor expansion to the 4th ventricle and obstructive hydrocephalus; truncal ataxia is also prominent in these patients [11]. We report a case with unusual imaging characteristics and subtle manifestation. Also, its genetic subtype was incongruent with common patterns.

Case presentation

The patient is a 30-year-old man who had continuous nausea and vomiting for 3 months before admission, with no cause confirmed in gastrointestinal investigations. Due to lack of response to treatment, brain imaging was performed. Signal changing in the vermis with no enhancement or peri-tumoral edema was the sole positive finding on imaging (Fig. 1A–C). On the neurological examination, the patient had no neurological deficit and the results of cerebellar-related examination were normal. The patient did not have truncal ataxia, and his gait was normal.

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Received: 27 January 2024; Accepted: 4 April 2024; Early publication: 24 April 2024

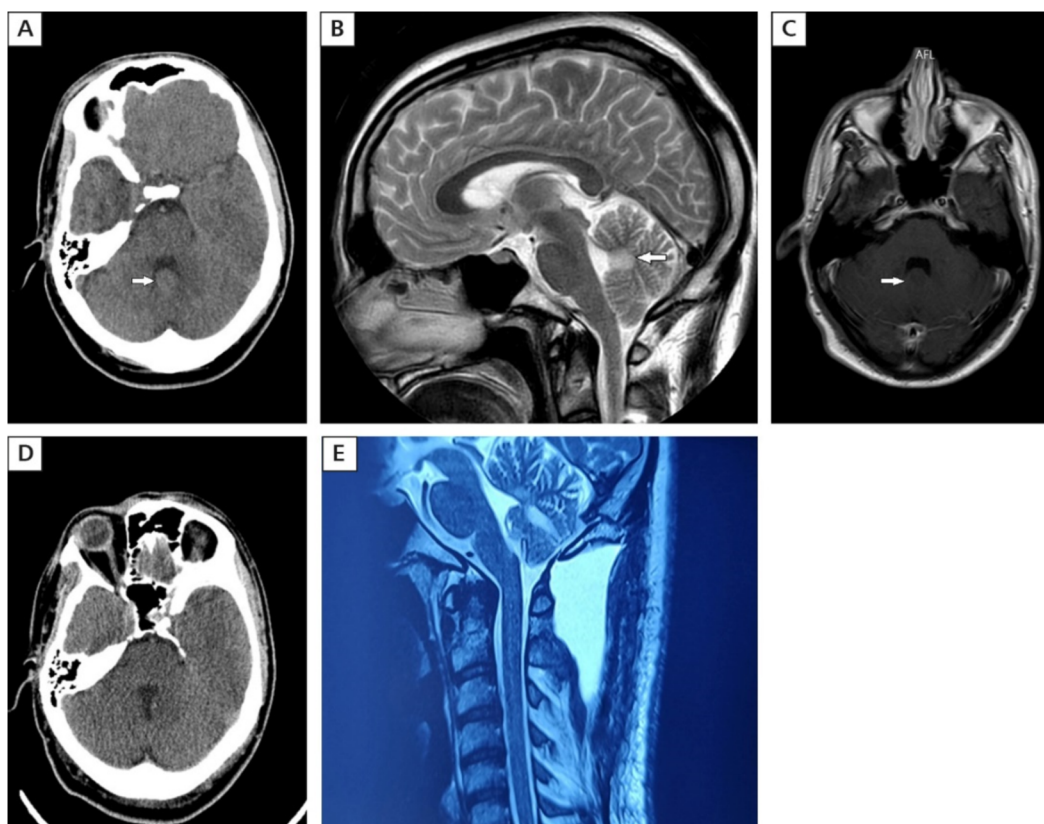


Figure 1. A–C. Pre-operative imaging. Axial brain computed tomography (CT)-scan without contrast injection (A), sagittal T₂ (B) and axial T₁ with gadolinium injection brain magnetic resonance imaging (MRI) (C). The lesion is mildly hyper-dense in CT-scan and signal change of anterior vermis, just behind the fastigium of the fourth ventricle, at the junction of its anterior lobe with the flocculonodular lobe is obvious; D–E. Post-operative imaging. Axial CT-scan and (B) sagittal T₂ MRI. The tract of telo-velar approach and elimination of signal change in vermis is obvious. A collection of cerebrospinal fluid (CSF) in craniocervical region due to dural leakage is evident but it was absorbed in follow up

Due to uncertain diagnosis and lack of another cause for the patient's nausea and vomiting, surgery was planned in a prone position with a sub-occipital craniotomy and telo-velar approach. The lesion had a soft consistency and was partly suctionable and was completely removed. After surgery, there were no new neurological deficits, and the patient's symptoms improved dramatically. Computed tomography (CT) showed adequate resection. Histopathological examination report was classic medulloblastoma, non-SHH and non-WNT (Fig. 2). On the post-operation imaging (Fig. 1D–E), there was no spinal spreading. The patient was referred for adjuvant therapy. We received a signed informed consent from the patient to use all his data and images in this publication.

Discussion

Medulloblastoma is a malignant embryonic tumor that has five histological variants. Sonic hedgehog is the most prevalent subtype in adults and is mostly seen in the lateral hemisphere of the cerebellum. Group 4 is the second most common subtype in adults and

has the worst prognosis [12]. The WNT has the lowest prevalence but the best prognosis and commonly occurs in the midline or the cerebellar peduncles. Group 3 is rare in adults [13].

In our case, the patient was a young man with low-intensity changes on magnetic resonance imaging (MRI) that were confined to the vermis without edema or enhancement, which ruled out a diagnosis of medulloblastoma. Also, according to the previously formulated concept that there is a correlation between location and histopathology of medulloblastoma, it seemed that the WNT was a more probable subtype in this case, but the histopathological exam showed that the tumor was non-SHH and non-WNT.

In our patient, we were in doubt whether he should be operated on, but due to persistent nausea and vomiting that did not respond to any medical treatment (metoclopramide, ondansetron, aprepitant, etc.) and lack of a definitive diagnosis for the changes in the vermis, we opted for surgery. It seems that the decision to operate on this patient was the best choice at that time because this surgery sped up the diagnosis, and he started chemo-radiotherapy sooner. Delay in

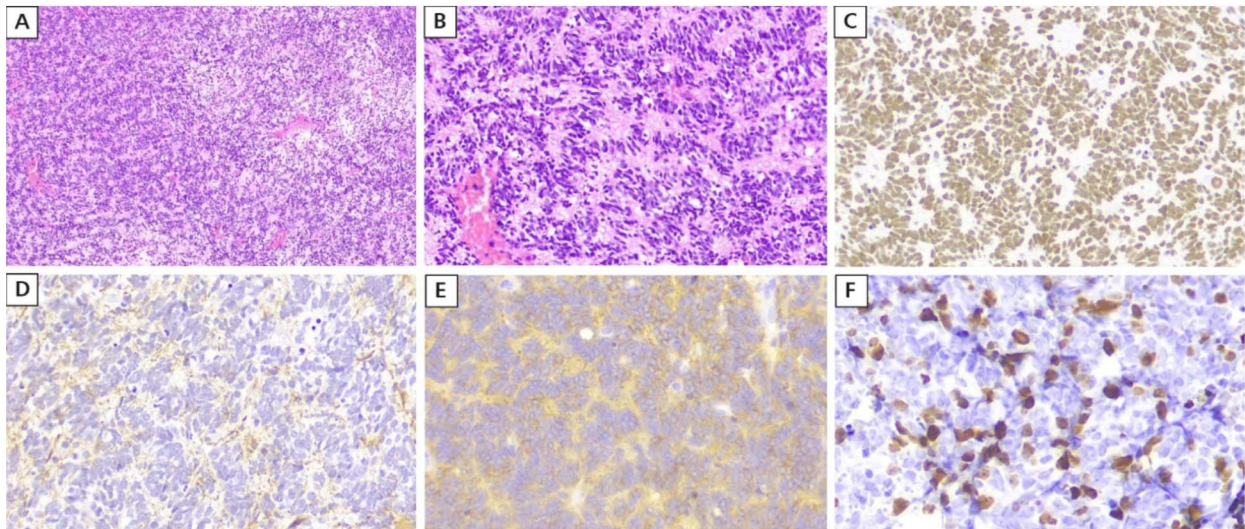


Figure 2. **A.** Classic medulloblastoma histology, comprising cellular sheet of small poorly differentiated embryonic cells in a scanty neuropil stroma (hematoxylin and eosin, $\times 40$); **B.** Frequent Homer-Wright rosette structures (hematoxylin and eosin, $\times 200$); **C.** Immunostaining for INI1 showed intact nuclear staining [INI1 immunohistochemistry (IHC), $\times 200$]; **D.** GFAP IHC showed no cytoplasmic reactivity ($\times 200$); **E.** The tumor cells are diffusely positive for synaptophysin IHC ($\times 200$); **F.** High proliferation index of activity of the tumor cells (Ki67 IHC, $\times 200$)

the treatment of medulloblastoma worsens prognosis, thus knowing tumor pathology and its immunohistochemically (IHC) properties can help in determining the treatment plan and informing the patient about the prognosis.

Conclusions

In the presence of posterior fossa lesions, medulloblastoma should always be kept in mind as one of the potential diagnoses, and a suspicion of such a diagnosis can expedite the identification and improve the prognosis.

Article Information and Declarations

Ethics statement

All data and images are allowed to be published by getting a signed informed consent form from the patient.

Author contributions

M.P.: conceptualization; H.G.: drafting and image production, correspondence; A.F.: data collection; A.S.: pathologic interpretation.

Funding

None.

Acknowledgements

None.

Conflict of interest

Authors declare no conflict of interest.

Supplementary material

None.

References

1. Qin Q, Huang D, Jiang Y. Survival difference between brainstem and cerebellum medulloblastoma: the surveillance, epidemiology, and end results-based study. *Medicine (Baltimore)*. 2020; 99(41): e22366, doi: [10.1097/MD.00000000000022366](https://doi.org/10.1097/MD.00000000000022366), indexed in Pubmed: 33031272.
2. Juraschka K, Taylor MD. Medulloblastoma in the age of molecular subgroups: a review. *J Neurosurg Pediatr*. 2019; 24(4): 353–363, doi: [10.3171/2019.5.PEDS18381](https://doi.org/10.3171/2019.5.PEDS18381), indexed in Pubmed: 31574483.
3. Louis DN, Perry A, Wesseling P, et al. The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. *Acta Neuropathol*. 2016; 131(6): 803–820, doi: [10.1007/s00401-016-1545-1](https://doi.org/10.1007/s00401-016-1545-1), indexed in Pubmed: 27157931.
4. Mrinal B, Hriday H, Basanta K, et al. A Clinical Study of medulloblastoma in Correlation with Molecular Biology and Histological Variations in Childhood medulloblastoma in Northeast India. *Indian Journal of Neurosurgery*. 2019; 8: 155–160.
5. Louis DN, Ohgaki H, Wiestler OD, et al. The 2007 WHO classification of tumours of the central nervous system. *Acta Neuropathol*. 2007; 114(2): 97–109, doi: [10.1007/s00401-007-0243-4](https://doi.org/10.1007/s00401-007-0243-4), indexed in Pubmed: 17618441.
6. Kool M, Korshunov A, Remke M, et al. Molecular subgroups of medulloblastoma: an international meta-analysis of transcriptome, genetic aberrations, and clinical data of WNT, SHH, Group 3, and Group 4 medulloblastomas. *Acta Neuropathol*. 2012; 123(4): 473–484, doi: [10.1007/s00401-012-0958-8](https://doi.org/10.1007/s00401-012-0958-8), indexed in Pubmed: 22358457.
7. Waite KA, Cioffi G, Kruchko C, et al. Aligning the Central Brain Tumor Registry of the United States (CBTRUS) histology groupings with current definitions. *Neurooncol Pract*. 2022; 9(4): 317–327, doi: [10.1093/nop/npac025](https://doi.org/10.1093/nop/npac025), indexed in Pubmed: 35859542.
8. Fellay CN, Frappaz D, Sunyach MP, et al. Medulloblastomas in adults: prognostic factors and lessons from paediatrics. *Curr Opin Neurol*. 2011; 24(6): 626–632, doi: [10.1097/WCO.0b013e32834cd4b1](https://doi.org/10.1097/WCO.0b013e32834cd4b1), indexed in Pubmed: 22027544.
9. Gajjar AJ, Robinson GW. Medulloblastoma-translating discoveries from the bench to the bedside. *Nat Rev Clin Oncol*. 2014; 11(12): 714–722, doi: [10.1038/nrclinonc.2014.181](https://doi.org/10.1038/nrclinonc.2014.181), indexed in Pubmed: 25348790.
10. Dangouloff-Ros V, Varlet P, Levy R, et al. Imaging features of medulloblastoma: Conventional imaging, diffusion-weighted imaging,

- perfusion-weighted imaging, and spectroscopy: From general features to subtypes and characteristics. *Neurochirurgie*. 2021; 67(1): 6–13, doi: [10.1016/j.neuchi.2017.10.003](https://doi.org/10.1016/j.neuchi.2017.10.003), indexed in Pubmed: [30170827](https://pubmed.ncbi.nlm.nih.gov/30170827/).
11. Zhang Na, Ouyang T, Kang H, et al. Adult medulloblastoma: clinical characters, prognostic factors, outcomes and patterns of relapse. *J Neurooncol*. 2015; 124(2): 255–264, doi: [10.1007/s11060-015-1833-y](https://doi.org/10.1007/s11060-015-1833-y), indexed in Pubmed: [26026861](https://pubmed.ncbi.nlm.nih.gov/26026861/).
 12. Zhao Fu, Ohgaki H, Xu L, et al. Molecular subgroups of adult medulloblastoma: a long-term single-institution study. *Neuro Oncol*. 2016; 18(7): 982–990, doi: [10.1093/neuonc/now050](https://doi.org/10.1093/neuonc/now050), indexed in Pubmed: [27106407](https://pubmed.ncbi.nlm.nih.gov/27106407/).
 13. Shonka N, Brandes A, De Gr. Adult medulloblastoma, from spongioblastoma cerebelli to the present day: a review of treatment and the integration of molecular markers. *Oncology*. 2012; 26: 1083–1091.