

A Rare Extra-Axial Midline Tentorial Adult Medulloblastoma with Dural-Tail Sign Mimicking a Meningioma

Abstract

Medulloblastomas are categorized as the World Health Organization Grade IV neoplasms. Only 33 cases have been reported of extra-axial, mostly in the cerebellar pontine angle and lateral cerebellar hemisphere, medulloblastomas in the current literature. Our study showcases the first case of an extremely rare presentation of an extra-axial midline tentorial adult medulloblastoma with the dural-tail sign mimicking a meningioma. To achieve the best possible outcome, a high index of suspicion for medulloblastoma is critical especially in young patient with an atypical posterior fossa mass as treatment regimens drastically different between a medulloblastoma and a meningioma.

Keywords: Adult medulloblastoma, dural-tail sign, meningioma, sonic hedgehog

Introduction

Medulloblastoma, a term originally phrased by Harvey Cushing and Percival Bailey in early 1920s, is the most common malignant pediatric brain tumor.^[1] It accounts for approximately 20% of all pediatric brain tumors.^[1,2] Bimodal incidence is noted between 3 and 4 years of age, and between 8 and 10 years of age.^[2,3] Up to 30% of the total 500 medulloblastoma cases diagnosed yearly are in the adult population.^[2-4] However, this represents <1% of all adult primary brain tumors.^[5,6] 75% of medulloblastomas arise from the cerebellar vermis and have a tendency to infiltrate the 4th ventricle; however cerebellar hemisphere is a more common site of origin for these tumors in adults.^[2,5] As described by Furtado *et al.*, the hemispheric location of medulloblastoma and its proximity to dura gives it the dural-tail sign partly due to tumor infiltration.^[5] Wilms *et al.* and Detwiler *et al.* initially described the dural enhancement and a tail tapering away from the tumor on contrast magnetic resonance (MR) images pertaining to meningiomas.^[5,7,8] Among the tumors that present with dural-tail enhancement, medulloblastomas are very rare. Only 33 cases have been reported of extra-axial, mostly in the cerebellar pontine angle (CPA) and lateral cerebellar hemisphere, medulloblastomas in the

current literature.^[5,9,10] Here, we present the first report of an extra-axial midline tentorial adult medulloblastoma with the dural-tail sign mimicking a meningioma.

Case Report

The patient is a 29-year-old male with no significant medical or surgical history who presented to the emergency room with a syncopal episode and headache for the last 2 weeks. The neurological exam was normal. The MR image of the brain with and without contrast revealed an enhanced extra-axial midline tentorial mass with the tentorial dural-tail sign [Figure 1]. The patient underwent stealth-guided suboccipital craniectomy and excision of the mass. Postoperative course was unremarkable and MR of the entire spine was negative for the drop metastasis. Pathology demonstrated medulloblastoma Grade IV, desmoplastic variant, and belonging to the sonic hedgehog (SHH) subgroup. The patient underwent radiation and adjuvant chemotherapies consisting of vincristine, cisplatin, and cyclophosphamide.

Discussion

Medulloblastomas are categorized as the World Health Organization grade IV neoplasms within the embryonal neuroepithelial tumors.^[2,11] These tumors often present in the midline but there have

**Ninh Ba Doan,
Mohit Patel,
Ha Son Nguyen,
Karl Janich,
Andrew Montoure,
Saman Shabani,
Michael Gelsomino**

*Department of Neurosurgery,
Medical College of Wisconsin,
Milwaukee, Wisconsin, 53226,
USA*

Address for correspondence:

*Dr. Ninh Ba Doan,
Department of Neurosurgery,
Medical College of Wisconsin,
9200 W. Wisconsin Ave,
Milwaukee, Wisconsin, 53226,
USA.*

E-mail: ndoan@mcw.edu

Access this article online

Website: www.asianjns.org

DOI: 10.4103/1793-5482.228563

Quick Response Code:



How to cite this article: Doan NB, Patel M, Nguyen HS, Janich K, Montoure A, Shabani S, *et al.* A rare extra-axial midline tentorial adult medulloblastoma with dural-tail sign mimicking a meningioma. *Asian J Neurosurg* 2018;13:475-7.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

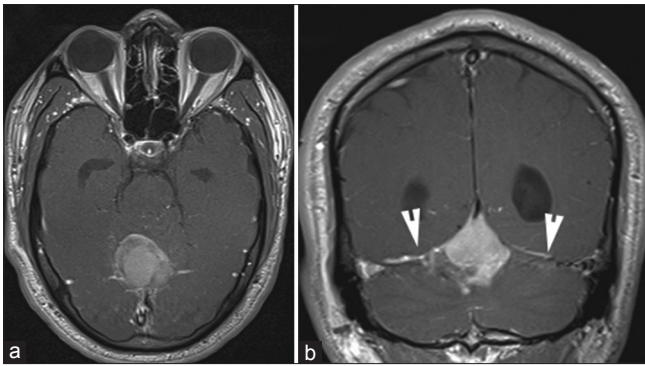


Figure 1: Magnetic resonance of the brain with contrast the axial plane (a) and the coronal plane (b) demonstrated an extra-axial midline tentorial mass with the dural-tail signs, which are represented by arrow heads

been rare case reports of them being present extra-axially. However, most of these extra-axial medulloblastomas are located at the CPA and lateral cerebellar hemisphere.^[9,12-14] Medulloblastomas are divided into several variants, such as the classical and desmoplastic/nodular types. Classical variant histology is the most commonly encountered medulloblastoma in practice; there is a presence of small, round to oval hyperchromatic nuclei and minimal cytoplasm.^[2,15-17] In desmoplastic/nodular variant, there is a presence of nodular, reticulin-poor islands of neurocytic differentiation surrounded by mitotically active cells.^[2,15-17] In addition, the desmoplastic histology is much more common occurrence in the adult population than in the pediatric population with 71% of them occurring in the lateral cerebellar hemisphere compared to 12.5% of the classical medulloblastoma variant.^[5,18,19] This lateral preference is partly explained by the fact that medulloblastomas arise from germinal cells anywhere along their migratory pathway, and these normally progress in a lateral direction.^[9,20]

In this report, we present the first report of an extra-axial midline tentorial adult medulloblastoma with the dural-tail sign [Figure 1]. Our patient was diagnosed with the desmoplastic/nodular variant of medulloblastoma because the morphology was consistent with that of a cerebellar embryonal tumor and medulloblastoma. In addition, molecular analysis for the tumor placed it in the SHH molecular subgroup. More interestingly, previous studies of the SHH molecular subgroup of medulloblastomas were noted to be the only subgroup among all different types in which the tumors were located within the cerebellar hemispheres.^[2,21] This would be the first reported case of SHH medulloblastoma that is not located in the cerebellar hemisphere. Furthermore, our patient's tumor presented with the dural-tail sign, which is most commonly encountered in meningiomas [Figure 1b]. However, the dural-tail sign has also been reported in various tumors such as glioblastoma multiforme, metastases, and lymphoma.^[5,7,8,22] There have only been two case reports describing the dural-tail presentation in a hemispheric medulloblastoma.^[5,8] Detwiler

et al. initially reported the presence of dural-tail in a hemispheric medulloblastoma, and attributed the dural changes that occur due to the tumor vicinity, angiogenesis, or inflammation as the possible cause for the dural-tail sign.^[5,8]

The treatment protocols for adults with medulloblastomas are based on the pediatric protocols.^[23] The standard involves surgical resection, radiation therapy, and adjuvant chemotherapy. A protocol, similar to the one that our patient is on, consisting of phase I and II of postradiation chemotherapy of vincristine and cyclophosphamide with or without the addition of cisplatin has been shown to have a 71–78% 5 years overall and progression free survival.^[2,23]

Conclusion

Our study showcases the first case of an extremely rare presentation of an extra-axial midline tentorial adult medulloblastoma with the dural-tail sign mimicking a meningioma. To achieve the optimal outcome, a high index of suspicion for medulloblastoma is crucial especially in young patient with an atypical posterior fossa mass as treatment regimens drastically different between a medulloblastoma and a meningioma.

Financial support and sponsorship

We would like to thank the Musella Foundation for its support.

Conflicts of interest

There are no conflicts of interest.

References

- Bailey P. Cushing medulloblastoma cerebelli: A common type of midcerebellar glioma of childhood. *Arch Neurol Psychiatry* 1925;14:192-224.
- Millard NE, De Braganca KC. Medulloblastoma. *J Child Neurol* 2015. pii: 0883073815600866.
- Smoll NR, Drummond KJ. The incidence of medulloblastomas and primitive neuroectodermal tumours in adults and children. *J Clin Neurosci* 2012;19:1541-4.
- McNeil DE, Coté TR, Clegg L, Rorke LB. Incidence and trends in pediatric malignancies medulloblastoma/primitive neuroectodermal tumor: A SEER update. *Surveillance Epidemiology and End Results. Med Pediatr Oncol* 2002;39:190-4.
- Furtado SV, Venkatesh PK, Dadlani R, Reddy K, Hegde AS. Adult medulloblastoma and the “dural-tail” sign: Rare mimic of a posterior petrous meningioma. *Clin Neurol Neurosurg* 2009;111:540-3.
- Menon G, Krishnakumar K, Nair S. Adult medulloblastoma: Clinical profile and treatment results of 18 patients. *J Clin Neurosci* 2008;15:122-6.
- Wilms G, Lammens M, Marchal G, Demaerel P, Verplancke J, Van Calenbergh F, *et al.* Prominent dural enhancement adjacent to nonmeningiomas malignant lesions on contrast-enhanced MR images. *AJNR Am J Neuroradiol* 1991;12:761-4.
- Detwiler PW, Henn JS, Porter RW, Lawton MT, White WL, Spetzler RF. Medulloblastoma presenting with tentorial

- “dural-tail” sign: Is the “dural-tail” sign specific for meningioma? *Skull Base Surg* 1998;8:233-6.
9. Chung EJ, Jeun SS. Extra-axial medulloblastoma in the cerebellar hemisphere. *J Korean Neurosurg Soc* 2014;55:362-4.
 10. Jaiswal AK, Mahapatra AK, Sharma MC. Cerebellopontine angle medulloblastoma. *J Clin Neurosci* 2004;11:42-5.
 11. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvet A, *et al.* The 2007 WHO classification of tumours of the central nervous system. *Acta Neuropathol* 2007;114:97-109.
 12. Brackmann DE, Bartels LJ. Rare tumors of the cerebellopontine angle. *Otolaryngol Head Neck Surg* 1980;88:555-9.
 13. Britton BH. Adult medulloblastoma: Neurotologic manifestations. *Ann Otol Rhinol Laryngol* 1975;84(3 Pt 1):364-7.
 14. House JL, Burt MR. Primary CNS tumors presenting as cerebellopontine angle tumors. *Am J Otol* 1985; Suppl:147-53.
 15. Sure U, Berghorn WJ, Bertalanffy H, Wakabayashi T, Yoshida J, Sugita K, *et al.* Staging, scoring and grading of medulloblastoma. A postoperative prognosis predicting system based on the cases of a single institute. *Acta Neurochir (Wien)* 1995;132:59-65.
 16. Giangaspero F, Perilongo G, Fondelli MP, Brisigotti M, Carollo C, Burnelli R, *et al.* Medulloblastoma with extensive nodularity: A variant with favorable prognosis. *J Neurosurg* 1999;91:971-7.
 17. McLendon RE, Friedman HS, Fuchs HE, Kun LE, Bigner SH. Diagnostic markers in paediatric medulloblastoma: A Paediatric Oncology Group Study. *Histopathology* 1999;34:154-62.
 18. Carrie C, Lasset C, Alapetite C, Haie-Meder C, Hoffstetter S, Demaille MC, *et al.* Multivariate analysis of prognostic factors in adult patients with medulloblastoma. Retrospective study of 156 patients. *Cancer* 1994;74:2352-60.
 19. Levy RA, Blaivas M, Muraszko K, Robertson PL. Desmoplastic medulloblastoma: MR findings. *AJNR Am J Neuroradiol* 1997;18:1364-6.
 20. Kadin ME, Rubinstein LJ, Nelson JS. Neonatal cerebellar medulloblastoma originating from the fetal external granular layer. *J Neuropathol Exp Neurol* 1970;29:583-600.
 21. Perreault S, Ramaswamy V, Achrol AS, Chao K, Liu TT, Shih D, *et al.* MRI surrogates for molecular subgroups of medulloblastoma. *AJNR Am J Neuroradiol* 2014;35:1263-9.
 22. Guermazi A, Lafitte F, Miaux Y, Adem C, Bonneville JF, Chiras J. The dural tail sign – Beyond meningioma. *Clin Radiol* 2005;60:171-88.
 23. Jakacki RI, Burger PC, Zhou T, Holmes EJ, Kocak M, Onar A, *et al.* Outcome of children with metastatic medulloblastoma treated with carboplatin during craniospinal radiotherapy: A Children’s Oncology Group Phase I/II study. *J Clin Oncol* 2012;30:2648-53.