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Case Report

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

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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

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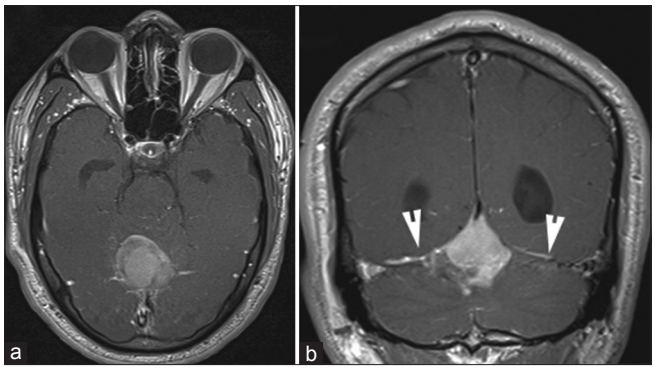

Figure 1: Magnetic resonance of the brain with contrast the axial plane (a) and the coronal plane (b) demonstrated an extra-axial midline tentorial mass with the dural-tail signs, which are represented by arrow heads

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.

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Conflicts of interest

There are no conflicts of interest.

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