Medulloblastoma invading the transverse sinus

Case report

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Medulloblastoma is a highly malignant brain tumor of childhood. Although craniospinal dissemination within the subarachnoid space is common, invasion of the dural sinuses is rare. Here, the authors report on a 15-year-old girl who presented with a right cerebellar mass, obstructive hydrocephalus, and radiographic evidence of tumor invasion into the right transverse–sigmoid sinus junction. The patient underwent posterior fossa craniotomy, gross-total resection of the intraparenchymal component of the right cerebellar tumor, and coagulation of the tumor invading the transverse sinus. After pathological confirmation of anaplastic medulloblastoma, the patient underwent craniospinal radiation therapy and high-dose chemotherapy. At 2 years posttreatment, the child was neurologically intact with no radiographic evidence of residual disease or recurrence. The implications for disease prognosis and management are discussed. (http://thejns.org/doi/abs/10.3171/2013.7.PEDS1363)

Key Words • medulloblastoma • transverse sinus invasion • metastasis • oncology

Medulloblastoma is a malignant, invasive embryonal tumor of the cerebellum that accounts for 40% of all childhood posterior fossa tumors and 16%–25% of all pediatric CNS tumors. These lesions are significantly more common in boys than in girls, with an overall 5-year survival of 70%–80%.5,9

Although the majority of children present with isolated masses of the fourth ventricle or cerebellar hemispheres, leptomeningeal dissemination is seen on craniospinal MRI or at the time of surgery in about one-third of patients and is associated with a worse prognosis. Despite its well-known propensity to metastasize along CSF pathways, local invasion of posterior fossa structures, such as the dura mater and dural venous sinuses, is extremely rare and has yet to be reported in children with medulloblastoma.

Here, we report the rare case of a child with medulloblastoma with focal invasion of the transverse sinus. The cellular mechanisms underlying this finding as well as the implications for disease prognosis and management are discussed.

Case Report

History and Examination. A 15-year-old girl presented to our emergency room with a 2-week history of headaches, vomiting, and progressive ataxia. She also reported a 2-day history of double vision and slurred speech. Computed tomography scanning of the head demonstrated a hyperdense, 5.6 (transverse) × 5.0 (anteroposterior) × 3.4 (superoinferior)—cm mass centered in the right cerebellar hemisphere with effacement of the fourth ventricle and obstructive hydrocephalus. Magnetic resonance imaging confirmed the presence of a homogeneously enhancing tumor within the right cerebellar hemisphere, with invasion into the right transverse-sigmoid sinus junction (Fig. 1). Despite evidence of tumor invasion and compression, the transverse and sigmoid sinuses remained patent with no obvious thrombus seen on preoperative MRI.

Operation. The patient underwent posterior fossa craniotomy and gross-total resection of the intraparenchymal component of the right cerebellar tumor. During surgery, residual tumor invading the junction of the right transverse and sigmoid sinuses was noted and deemed unamenable to safe resection. This component was instead coagulated using bipolar cautery.

Pathological Findings. Histological examination of intraoperative specimens revealed a cellular neoplasm arranged in a sheetlike architecture with diffuse invasion of the surrounding cerebellum. The tumor cells showed moderate anaplasia with prominent nucleoli. Abundant mitotic figures, apoptotic bodies, and areas of necrosis were present. Tumor cells were immunopositive for BAF47 and immunonegative for nuclear β-catenin. Immunopositivity for p53 was observed in 25% of the cells. Neither nodularity nor desmoplasia was appreciated. Fluorescence in situ hybridization revealed amplification of MYCN. Final pathological diagnosis was consistent with classic medulloblastoma with anaplasia.

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**Postoperative Course.** Postoperative MRI revealed gross-total resection of the cerebellar intraparenchymal component with a small amount of enhancing tumor (<1.5 cm²) within the right transverse sinus (Fig. 2). Spinal MRI and lumbar puncture were negative for disseminated disease (M0, baseline). Postoperative clinical examination revealed normal cranial nerves, no significant motor or sensory dysfunction, mild dysdiadochokinesis, and mild truncal ataxia. The patient underwent radiation therapy, including 23.5 Gy to the craniospinal axis and a 55.8-Gy boost to the tumor bed, followed by 4 courses of high-dose chemotherapy with autologous stem cell transplant. At the 2-year follow-up, the patient had no neurological deficits (modified Rankin Scale Score 0) and no radiographic evidence of tumor recurrence (Fig. 3).

**Discussion**

Among pediatric brain tumors, medulloblastoma has a relatively high tendency toward CSF dissemination and, more rarely, extraneural spread. At diagnosis, 14%–43% of patients exhibit microscopic or nodular seeding in the subarachnoid space of the spine or brain.3,11 Spread outside the CNS is unusual in children, although not exceptional in adults,6,10 with bone and the abdominal cavity the most common extraneural sites involved.

Patients with medulloblastoma most commonly present with a posterior fossa mass and hydrocephalus. A minority of patients may present with diffuse leptomeningeal dissemination, which is known as “sugar coating” of the brain on postcontrast T1-weighted MRI. Odd presentations have been reported occasionally, including a skeletal...
Fig. 3. Two-year follow-up axial postcontrast T1-weighted MR image revealing no evidence of recurrent or residual tumor.

metastasis that was the initial presentation in a 21-year-old man. The patient in our case presented with direct invasion of one of the major dural venous sinuses, which is an unusual finding. To our knowledge, medulloblastoma with dural sinus invasion has not been reported previously.

In 1996 Eisen et al. mentioned that preoperative MRI is considered a good tool to predict dural, perineural, and venous sinus invasion of skull base tumors. In addition, postoperative MRI should be performed within 48–72 hours after surgery before gliosis and blood products obscure one’s ability to identify any residual tumor. In our case MRI showed this invasion both before and after surgery. In most cases of medulloblastoma, the surgeon should attempt to achieve gross-total or near-total resection, as the literature suggests a poorer prognosis in children with less complete resections. Nonetheless, surgical zeal should be tempered with appropriate caution to prevent morbidity and mortality, particularly in the face of unusual patterns of disease.

Our patient had no clinical evidence of leptomeningeal dissemination, the residual tumor was smaller than 1.5 cm², and the pathology was classic medulloblastoma with moderate anaplasia. She received radiotherapy in total of 23.5 Gy to the craniospinal axis and a 55.8-Gy boost to the tumor bed in addition to chemotherapy. If the size of the tumor remaining at the site of dural invasion had been larger than 1.5 cm², further resection or treatment as a high-risk medulloblastoma may have been required. In removing as much tumor as possible and coagulating the remnant, it was decided that the lesion could be treated as a standard-risk medulloblastoma.

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 to the study and manuscript preparation include the following: Conception and design: Nadi, Ahmad. Acquisition of data: Nadi, Khezri, Ahmad. Analysis and interpretation of data: Nadi, Bouffet. Drafting the article: Nadi, Khezri, Ellis, Rutka. 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: Taylor. Administrative/technical/material support: Ahmad. Study supervision: Taylor, Rutka.

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