Medulloblastoma

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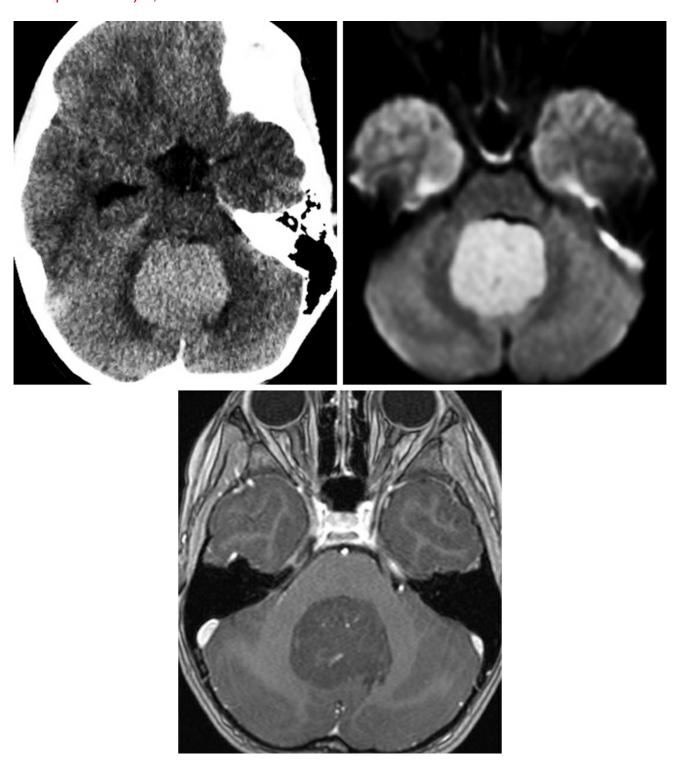
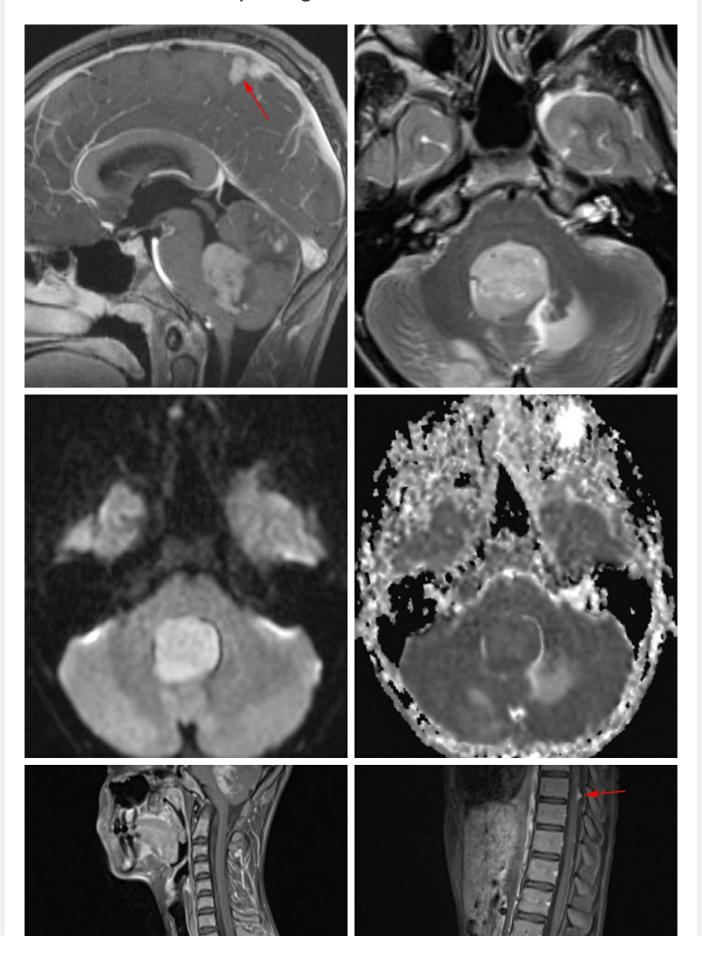


Figure 1: (Top Left) The hyperdensity of this medulloblastoma on CT imaging is typical and representative of the tumor's hypercellularity. (Top Right) Likewise, DWI shows restricted diffusion, also representing

hypercellularity. (Bottom) The patient's lack of significant enhancement on this T1-weighted postcontrast image is most typical of group 4 medulloblastomas. The intraventricular location is also typical, with these lesions sometimes visibly arising from the fourth ventricular roof.





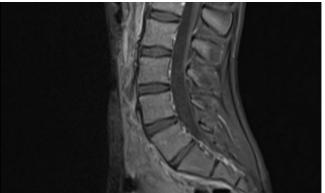


Figure 2: Sagittal T1 postcontrast (top left), axial T2-FS (top right), DWI (middle left), and ADC (middle right) images demonstrate an enhancing mass in the fourth ventricle with restricted diffusion (bright on DWI and dark on ADC) compatible with a hypercellular tumor. Multiple enhancing supratentorial and infratentorial nodules are noted along the surface of the cerebrum and cerebellum (top left), compatible with CSF dissemination of the tumor. (Bottom Left and Bottom Right) Sagittal T1 postcontrast images of the cervical, thoracic, and lumbar spine demonstrate enhancing nodules along the dorsal surface of the spinal cord compatible with drop metastases in this patient with aggressive medulloblastoma.

BASIC DESCRIPTION

- Malignant, invasive primitive neuroectodermal tumor usually arising within the posterior fossa
- Most common pediatric malignant posterior fossa tumor

PATHOLOGY

- WHO grade IV
- Divided into 4 molecular subtypes that originate in different locations:
 - Wingless (WNT): cerebellar peduncles, cerebellopontine angle
 (CPA) cistern
 - Sonic hedgehog (SHH): cerebellar hemispheres
 - o Group 3: fourth ventricle, midline
 - o Group 4: fourth ventricle, midline

• Divided into 4 distinct histologic subtypes: classic (most common), desmoplastic, large cell/anaplastic, and extensive nodularity

CLINICAL FEATURES

- Most commonly afflicts children aged 5 to 12 years; adult cases are rare
- Common presenting signs/symptoms are related to increased intracranial pressure/hydrocephalus
 - Headache, nausea, vomiting, altered mental status, papilledema, macrocephaly
- Treatment: surgical resection ± adjuvant chemoradiation
- Prognosis varies with molecular subtype and histology:
 - Wingless most favorable
 - Group 3 least favorable

IMAGING FEATURES

- General
 - Appearance varies with location/subtype
 - Solid, rounded fourth ventricular mass most common
 - ±Fourth ventricular effacement and distortion (in comparison with ependymoma, which often expands/enlarges the fourth ventricle)
 - Calcification and cysts occasionally present
 - Hemorrhage uncommon
 - Cerebrospinal fluid (CSF) dissemination common, including drop metastases within the spinal canal (15%–50%)
- CT
 - Fairly homogeneous, mildly hyperdense fourth ventricular mass
 - ±Hyperdense calcifications or hypodense cysts
 - Variable enhancement on contrast-enhanced CT imaging

(patchy or homogeneous)

- MRI
 - T1WI: hypointense
 - T2WI: isointense to hyperintense
 - FLAIR: improved delineation of tumor and CSF; minimal peritumoral edema
 - T1WI+C: majority show heterogeneous enhancement, although variable patterns exist; may see CSF dissemination throughout neuraxis
 - DWI: demonstrates DWI-bright, ADC-dark diffusion restriction
 - MRS: elevated Cho, decreased NAA, ± lactate peak

IMAGING RECOMMENDATIONS

 MRI without and with intravenous contrast including both brain and spine due to risk of CSF dissemination

For more information, please see the corresponding chapter in <u>Radiopaedia</u>.

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