

## 2. Infratentorial Brain Neoplasms

Medulloblastomas, which occur most often in children, are the most common infratentorial neoplasm (and the most common malignant pediatric brain tumor). 75% originate in the cerebellar vermis and protrude into the fourth ventricle (Fig. 2.1 A, B). Medulloblastomas are highly malignant, commonly spreading via the CSF. Thus, lumbar puncture and contrast-enhanced MR imaging of the entire neural axis are essential. Metastasis outside the CNS is rare, most commonly involving the bone marrow. Treatment consists of some combination of resection, craniospinal radiotherapy, and chemotherapy. Like most brain tumors, medulloblastomas have low SI on T1WI (white arrow, Fig 2.1 A). On T2WI, their appearance varies from isointense to mildly hyperintense relative to brain parenchyma. Medulloblastomas show restricted diffusion, a point of distinction from astrocytomas. Intense, heterogenous contrast enhancement (Fig 2.1 B, black arrow) is typical, although some lesions demonstrate no or only patchy enhancement. Medulloblastomas tend to extend into the 4<sup>th</sup> ventricle, leading to obstructive hydrocephalus, reflected in part by ventricular enlargement in Figure 2.1 A. The increase in ventricular pressure leads to a

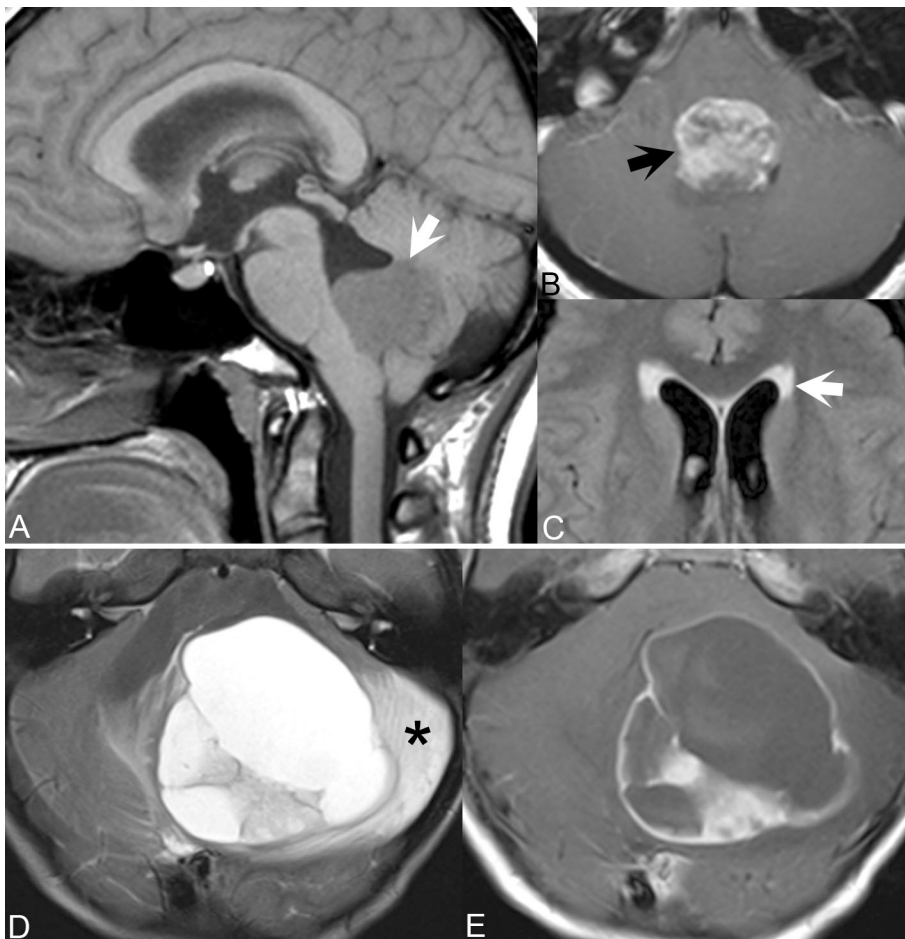


Fig. 2.1

thin uniform band of abnormal high SI present on T2WI in the periventricular white matter (Fig 2.1 C, white arrow)—due to increased interstitial fluid. Spectroscopy reveals a marked elevation in the ratio of choline to N-acetyl-aspartate (NAA)—a finding typical of malignant tumors—and taurine. A medulloblastoma may be difficult to distinguish from an ependymoma on imaging. However, ependymomas are softer ‘plastic’ tumors and tend to accommodate to the shape of the ventricle, often also squeezing through the foramina (Magendie and/or Luschka), while medulloblastomas tend to deform the 4<sup>th</sup> ventricle (Fig 2.1 A).

Cerebellar astrocytomas closely follow medulloblastomas, in incidence, being the second most common posterior fossa neoplasm. A midline, vermian mass exhibiting restricted diffusion favors medulloblastoma while a hemispheric cerebellar mass favors astrocytoma. Pilocytic astrocytomas (PAs) are WHO Grade I astrocytomas and comprise the majority of cerebellar astrocytomas. These tumors carry a good prognosis (90% survival after 10 years) and high rate of surgical cure. They are associated with neurofibromatosis type 1, in which disease the lesions most commonly occur along the optic pathway. PAs tends to be round, cystic lesions (Fig. 2.1 D, E) with a solid component (or nodule). The tumor in Figure 2.1 D, E is dominated by a cystic component which characteristically appears similar in SI to CSF on T1WI and T2WI, but can be differentiated from such on FLAIR scans. Lack of high SI intensity on T2WI argues against PA. Because of elevated protein content, the cystic components will appear bright on FLAIR images. Unlike medulloblastomas, both the cystic and solid components of a PA typically demonstrate increased apparent diffusion coefficient (ADC) values. Though the nidus of the tumor in Figure 2.1 D-E is clearly seen on T2WI, this may not always be the case. Thus, every cystic cerebellar lesion warrants further evaluation with intravenous contrast administration to distinguish a PA from a purely benign fluid collection. The tumor nidus will invariably enhance, and the cyst rim may as well (Fig. 2.1 E). Though benign, PAs can be associated with substantial vasogenic edema (Fig 2.1 D, asterisk). Non-PA cerebellar astrocytomas tend to be solid, well-differentiated but infiltrating, and thus associated with a poorer prognosis. The latter lesions often fail to enhance, thus further distinguishing them from PAs.

Other brainstem and posterior fossa tumors include hemangioblastoma, choroid plexus papilloma and diffuse midline glioma (brainstem glioma). Hemangioblastomas are WHO grade I tumors, which occur both sporadically and in von Hippel-Lindau disease. An enhancing mural nodule with a nonenhancing cystic component, which may be confused with the cysts of PA, is characteristic. Hemangioblastomas tend to be smaller and affect an older (>15 years of age) population than PA. Choroid plexus papillomas are strongly enhancing, lobulated (cauliflower-like) intraventricular lesions, with the most common

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locations being the lateral (usually atrium) and 4<sup>th</sup> ventricles. Diffuse midline gliomas (which include tumors previously referred to as diffuse intrinsic pontine gliomas), found in the thalamus, brainstem and spinal cord, are expansile tumors with high SI on T2WI and FLAIR and little to no enhancement. These carry a poor prognosis, with a 2-year survival less than 10%.