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Desmoplastic Medulloblastoma: MR Findings

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Summary: In three cases of desmoplastic medulloblastoma, MR findings were varied. We report the unusual appearance of this tumor in two children and one adult.

Index terms: Medulloblastoma; Brain neoplasms, magnetic resonance

The desmoplastic variant of medulloblastoma is typically seen in adolescents or young adults and tends to favor one of the cerebellar hemispheres. We describe a spectrum of magnetic resonance (MR) imaging findings in three patients with intracranial desmoplastic medulloblastoma.

Case Reports

Case 1

A 33-year-old woman had a 1-month history of double vision and occipital headaches. A right-sided sixth cranial nerve palsy and cerebellar signs were present. MR imaging revealed a 4.2×3.8 -cm craniocaudal, midline, predominantly intraaxial posterior fossa mass invading the falx cerebelli, compressing the fourth ventricle, and causing tonsillar herniation. The mass was isointense with brain, enhanced nonhomogeneously, and had multiple cystic components. A second, similar, intraaxial left-sided cerebellar mass was minimally enhancing, measured 2.5×1.2 cm, and had only a small amount of associated edema. There was no evidence of other central nervous system (CNS) or systemic metastatic lesions. At surgery, the midline mass was identified microscopically as a desmoplastic medulloblastoma (Fig 1).

Case 2

A 6½-year-old boy had a 1-year history of progressive, severe headaches, recently with photophobia and vomiting. Neurologic examination was nonfocal. An MR image of the brain revealed abnormal enhancement of the meninges over the cerebral convexities and of the tentorium and falx. There was mild to moderate enlargement of the third and lateral ventricles. CSF culture was negative and a

dural biopsy specimen showed fibrosis without tumor or infection. One year later, the patient was admitted with an unsteady gait, decreased vision, emesis, and left-sided hemiparesis. An MR image of the brain revealed progressive diffuse meningeal enhancement, a large right frontoparietal enhancing mass with leftward bowing of the falx, multiple intraaxial and extraaxial enhancing nodular lesions, and T2 hyperintensity in a periventricular, right lentiform nuclear and right frontal distribution (Fig 2). There was no evidence of other CNS or systemic metastatic lesions. Brain and meningeal biopsy samples subsequently showed desmoplastic medulloblastoma.

Case 3

A 5-year-old girl had a 2-week history of fatigue, progressive headache, nausea and vomiting, and papilledema. MR imaging of the brain revealed a nonhomogeneously enhancing mass on T1-weighted images that was nonhomogeneously isointense and hypointense on fast spin-echo T2-weighted images, with cystic components and an intraaxial and extraaxial appearance. It extended into the right foramen acusticus and jugular foramen from the right cerebellar hemisphere and the region of the foramen of Luschka (Fig 3). There was no evidence of other CNS or systemic metastatic lesions. Subsequent subtotal tumor resection revealed a desmoplastic medulloblastoma.

Discussion

Although Bailey and Cushing in 1925 established the medulloblastoma principally as a tumor of the posterior vermis in children, Cushing, in a more exhaustive survey of this material (1930), included in his survey of 61 cerebellar medulloblastomas nine that were laterally situated, eight of these in adults. This subgroup of tumors, subsequently designated *desmoplastic medulloblastoma*, is characterized by an early and extensive local infiltration of the overlying meninges, probably arising from the more superficial part of the cerebellar molecular layer,

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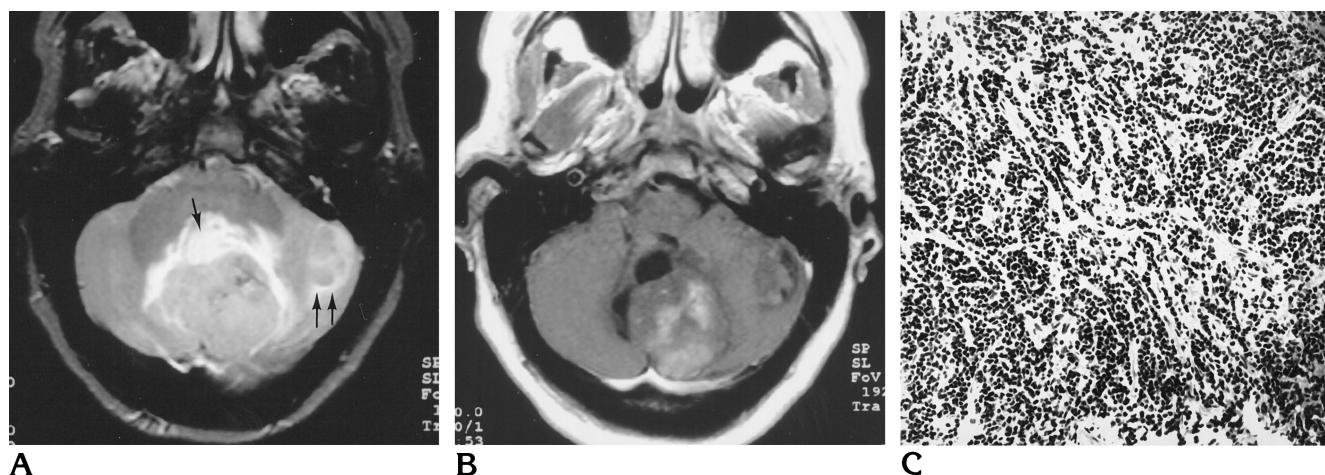


Fig 1. Case 1: 33-year-old woman with double vision and occipital headaches for 1 month.
 A, Axial T2-weighted (2400/90/1 [repetition time/echo time/excitations]) conventional spin-echo MR image shows a large isointense midline posterior fossa mass with a ventral cystic component (*single arrow*) and mild surrounding edema. A smaller left hemispheric intraaxial tumor mass is present (*double arrows*).
 B, Contrast-enhanced (0.1 mmol/kg gadoteridol) T1-weighted (600/15/1) axial MR image shows nonhomogeneous enhancement of the larger mass with minimal enhancement of the smaller mass (desmoplastic medulloblastoma).
 C, Uniformly small cells without significant nuclear pleomorphism and virtually absent cytoplasm arranged in lines or clusters on a reticulin-rich background (hematoxylin-eosin, original magnification $\times 325$).

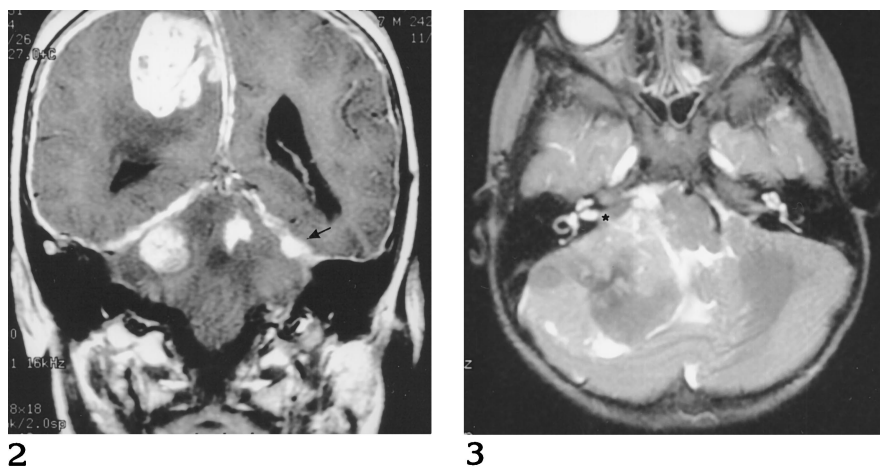


Fig 2. Case 2: Contrast-enhanced (0.1 mmol/kg) T1-weighted (500/18/1) coronal MR image of a 6½-year-old boy at 1-year follow-up shows increased diffuse meningeal enhancement with a large enhancing right frontoparietal intraaxial mass bowing the falx, bilateral intraaxial cerebellar masses, and a left tentorial-based nodule (*arrow*) (desmoplastic medulloblastoma).

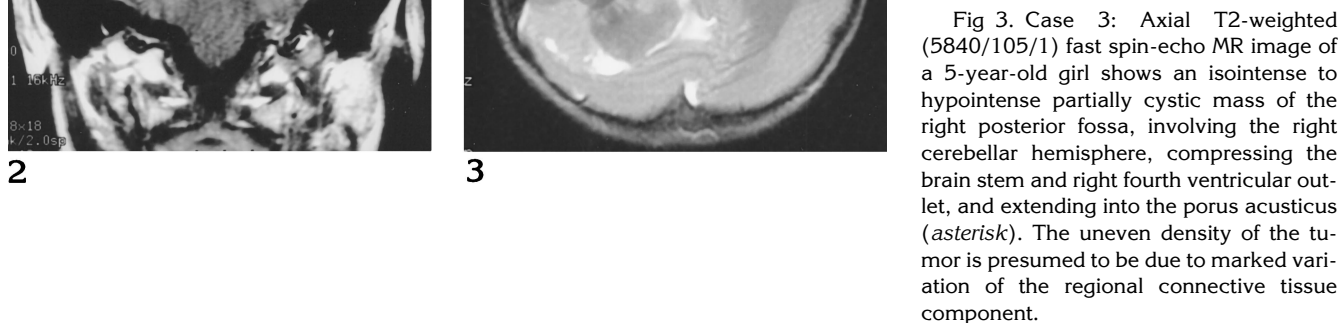


Fig 3. Case 3: Axial T2-weighted (5840/105/1) fast spin-echo MR image of a 5-year-old girl shows an isointense to hypointense partially cystic mass of the right posterior fossa, involving the right cerebellar hemisphere, compressing the brain stem and right fourth ventricular outlet, and extending into the porus acusticus (*asterisk*). The uneven density of the tumor is presumed to be due to marked variation of the regional connective tissue component.

and by a variable amount of reticulin fiber formation (1). More recent series have confirmed that desmoplastic medulloblastoma tends to occur more often in the lateral cerebellar hemisphere than does the classic medulloblastoma (71% versus 12.5%). Its mean age of onset of 17 years also stands in contrast to that of classic medulloblastoma (9 years in the same study) (2). The term desmoplasia in the pathologic classification of tumors implies the presence of a sizable component of collagenous connective

tissue within a neoplasm. If the entire tumor is composed of connective tissue, it is called a *desmoid tumor* (as in the abdomen) or a *fibroma/fibrosarcoma* (elsewhere in the body). In desmoplastic medulloblastoma, a variable but appreciable amount of reactive connective tissue is present in addition to neoplastic medulloblastoma cells. The medulloblastoma cells form nodules or linear trabeculae separated by bands of collagen visible in reticulin stain. In addition, lightly stained, reticulin-free islands of

tumor may also be seen in continuation with a more compact linear cellular arrangement of reticulin-rich tumor.

We found an example of an MR image of this tumor showing a large multicystic mass of the right posterior fossa invading and compressing the brain stem and extending superiorly into the quadrigeminal cistern (3), reminiscent of the findings in our case 3 (Fig 3). In this 5-year-old girl, our initial differential diagnosis included ependymoma, because of the characteristic findings of cyst formation, extraaxial infiltration, and a location in part adjacent to the foramen of Luschka (4). The relative isointensity on T2-weighted images is consistent with both ependymoma and medulloblastoma (primitive neuroectodermal tumor), and the latter was the other preoperative consideration. The more striking low-signal component of the tumor on T2-weighted images in case 3 relative to that in case 1 reflects a greater degree of desmoplasia histologically (Fig 3).

The imaging findings and clinical course in case 2 were the most atypical in our series. Perhaps the only consistent explanation comes from early histologic descriptions of desmoplastic medulloblastoma in which the bulk of the tumor is actually extracerebellar, associated with a predominantly meningeal direction of growth originating in the superficial part of the cerebellar molecular layer (1). It is therefore postulated that the initial diffuse meningeal and later intraaxial and subependymal foci of tumor reflect this phenomenon. The differential diagnosis of tumors associated with desmoplasia includes desmoplastic infantile ganglioglioma (a supratentorial neoplasm arising in infancy), superficial dural astrocytoma, and gliofibroma (3). Pleomorphic xanthoastrocytoma characteristically involves the meninges and can produce a desmoplastic reaction, although this is not typical (5). MR findings of large series have been reported only with pleomorphic xanthoastrocytomas (6).

In our three cases, the findings in case 1 are

most typical of previously reported desmoplastic medulloblastoma, particularly the patient's age and the lateral cerebellar hemispheric location of one tumor focus. The multicentricity of the tumor masses is still atypical (2). The differential diagnosis is principally that of metastatic tumor, especially adenocarcinoma.

There is disagreement about the prognostic significance of histopathology in medulloblastoma. Early and more recent studies that have included both adults and children found improved survival in patients with desmoplastic medulloblastoma (7, 8). Other series, mostly of adult patients, reported no significant difference in the outcome of patients with desmoplastic versus classic medulloblastoma (9, 10).

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