Pediatric Cerebellar Tumors

Emerging Imaging Techniques and Advances in Understanding of Genetic Features

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KEYWORDS

• Pediatric radiology • Neuroradiology • Brain tumor • Cerebellum • Medulloblastoma • Pilocytic astrocytoma • Ependymoma

KEY POINTS

- Cerebellar tumors are the most common solid neoplasms in children, with the 3 most common entities including pilocytic astrocytoma, medulloblastoma, and ependymoma.
- Diffusion-weighted/tensor imaging plays a key role in preoperative characterization of pediatric cerebellar tumors, with lower apparent diffusion coefficient values correlating with higher-grade tumors.
- Genetic characterization is resulting in new understanding of medulloblastoma.
- The previous 4 histologic categories are in the process of being supplanted with 4 genetic groupings, in particular based on analysis of the WNT and Sonic Hedgehog pathway genes.
- This genetic characterization has allowed therapeutic options targeted to the specific tumor and improved prediction of tumor aggressiveness compared with histologic categorization.

INTRODUCTION

Cerebellar tumors are among the most common central nervous system (CNS) neoplasms, not to mention solid neoplasms, in children.[1] These tumors include benign and malignant entities, tumors that have slow local spread, and others with leptomeningeal dissemination. Although there are a small number of tumor types that account for most of these tumors, recent work on the genetic origins of these lesions has created an understanding of a much broader landscape of tumors.[2] The newer understanding of genetic origins has the potential for targeted therapy, and imaging features that may help determine the tumor type (or even subtype) take on an increasingly critical role.

TUMOR TYPES

Pediatric cerebellar tumors most commonly involve four entities. Pilocytic astrocytomas (PAs), ependymomas, and medulloblastomas are the key players, with atypical teratoid rhabdoid tumor (ATRT)

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representing a high-grade embryonal tumor most common in the first year or two of life. Initial clinical presentation for all of these lesions is typically due to mass effect, including headaches, nausea and emesis, cranial neuropathies, and obstructive hydrocephalus.

PILOCYTIC ASTROCYTOMA

PAs, sometimes referred to as juvenile PAs, are benign neoplasms, classified as World Health Organization (WHO) grade I tumors. They account for more than two-thirds of all cerebellar astrocytomas and can be seen from birth to about 15 years of age. Other more aggressive astrocytomas, such as anaplastic astrocytomas and glioblastomas, are rare and usually seen in older children. There is an equal incidence of PA in boys and girls. PA of the cerebellum can arise from nearly any location in the cerebellum, including the hemispheres and vermis. PA of the cerebellum most classically has an imaging appearance of a cystic lesion with a mural nodule (Fig. 1). The solid portions of the tumor have a large volume of interstitial space with a high water content, which results in a somewhat hyperintense appearance on T2-weighted (T2W) imaging, and results in facilitated diffusion. The solid portion of PAs typically has diffusion characteristics of greater than 1300×10^{-6} mm^2/s (or 1.3×10^{-3} mm^2/s).[3] The solid portions will

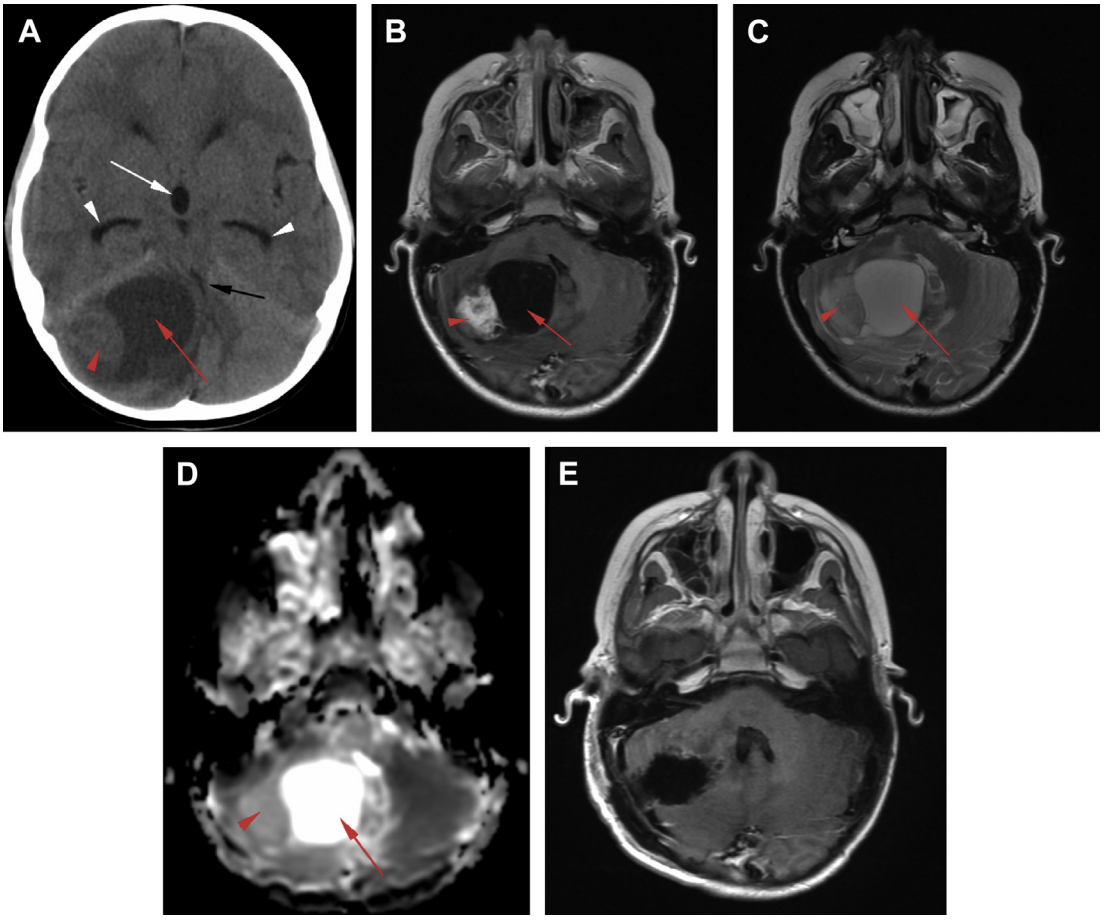

Fig. 1. PA. (*A*) Axial–computed tomography image in a 6-year-old boy shows a mass in the right cerebellar hemisphere that has a cystic component (*red arrow*) and a solid component (*red arrowhead*). There is near-complete effacement of the fourth ventricle (*black arrow*) with signs of obstructive hydrocephalus in the third ventricle (*white arrow*) and temporal horns of the lateral ventricles (*white arrowheads*). (*B*) Axial T1W + C image shows enhancement of the solid nodule (*red arrowhead*) but not of the cystic component (*red arrow*). (*C*) Axial T2W image shows intermediate hyperintense signal for the solid nodule (*red arrowhead*), suggesting a high water content, and fluidlike signal in the cystic component (*red arrow*). (*D*) ADC map shows facilitated diffusion in the solid nodule (*red arrowhead*), with diffusion characteristics of 1570×10^{-6} mm^2/s. The diffusion characteristics of the cystic component (*red arrow*) were greater than 3000×10^{-6} mm^2/s. (*E*) Axial T1W + C MR imaging after surgical resection shows gross total resection. This lesion was confirmed to be a PA. T1W + C, T1 + contrast.