An Unusual Presentation of Dysembryoplastic Neuroepithelial Tumor as a Fourth Ventricular Mass

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Purpose
To present the histopathologic and radiographic features of a dysembryoplastic neuroepithelial tumor centered within the fourth ventricle.

Materials & Methods
Dysembryoplastic neuroepithelial tumors (DNTs) typically occur in children and young adults with pharmacoresistant complex partial seizures. Radiographically, DNTs are generally cortically based tumors, predominately supratentorial in location, most commonly within the temporal lobe, and are frequently associated with cortical dysplasia. To our knowledge, there are only four reports describing the histologic features of DNTs in the posterior fossa and no reports of fourth ventricular DNTs. The authors present a case in which a tumor occurred in the fourth ventricle of a 13-year-old girl presenting with a 3-week history of headaches. Unenhanced CT of the head demonstrated obstructive hydrocephalus as a result of a low density mass (H.U. 15) within the fourth ventricle. On MR imaging, the 4.0 cm diameter nonenhancing mass was centered within the fourth ventricle and extended superiorly into the quadrigeminal plate and superior cerebellar cisterns. The lesion was low in signal on T1, high in signal on FLAIR. The lesion was predominately high in signal on T2 with a "speckled" appearance and was normal to slightly increased in signal with diffusion. MR spectroscopy showed an elevated choline-to-creatine ratio, decreased NAA, and a possible small lactate peak.

Results
Histologically, the tumor was composed of predominately small round cells within a mucinous background, containing occasional random "floating" well-differentiated mature neurons within these mucin pools. The lesion had a multinodular architecture (though in some areas "diffuse"), involved cerebellar parenchyma, demonstrated the "specific glioneuronal element," forming linear arrays, and was associated with cerebellar cortical dysplasia (increased and ectopic granule cells in the molecular layer, focally forming small nodules; also some displaced and atypical Purkinji cells), all findings consistent with the "complex" form of DNT. The immunohistochemistry and special stains were also typical for DNT. The ultrastructural analysis showed primitive-appearing
cells with high nuclear-cytoplasmic ratio and abundant cell processes suggestive of oligodendroglial-type cells, but with cell processes containing microtubules and dense core neurosecretory granules. These features are also consistent with DNT.

**Conclusion**
The diagnosis of DNT should be included in the differential of nonenhancing infratentorial and intraventricular lesions which are low in signal on T1, high in signal on T2 with a paucity of peritumoral edema, and normal to slightly increased in signal on diffusion.

**References**