Tentorial Traversal by Ependymoblastoma

We present a case in which MR showed unusual erosion of a supratentorial tumor through the tentorium.

Case Report

An otherwise normal 28-month-old boy had a 2-month history of intermittent vomiting and a 1-week history of headaches. Neurologic examination was normal except for a right extensor plantar response.

Axial CT scan (Fig. 1A) showed a large primarily intraventricular mass occupying the posterior left cerebral hemisphere. Directly caudad to this, a similar tumor was present in the left cerebellar hemisphere displacing the brainstem and fourth ventricle (Fig. 1B). No tumor was seen in the tentorial hiatus. Extensive patchy areas of increased density scattered throughout the tumor were interpreted as either blood or calcification. Coronal MR images (Fig. 1C) showed traversal of the supratentorial tumor through the tentorium into the left cerebellar hemisphere. Scattered throughout the tumor were large irregular areas of hemorrhage that were identified by increased signal intensity on T1 sequences. The areas of increased signal corresponded to the areas of increased attenuation on CT scans.

At surgery, a large invasive necrotic tumor of ventricular origin was resected partially. It extended through a large hole in the tentorium to invade the cerebellum.

Microscopically, the tumor was densely cellular and composed of uniform cells with hyperchromatic nuclei and, frequently, mitotic figures. Scattered small central-lumen rosettes (true rosettes) and tubules with multiple layers of nuclei were present. Occasionally, gliovascular structures were seen in which blood vessels were surrounded by radiating processes of tumor cells that had nuclei in the antipodal position. No calcium was seen. Pathologic diagnosis was ependymoblastoma.

Chemotherapy was attempted without success. The patient died 3 months after presentation.

Discussion

Ependymoblastoma is a rare neoplasm of the CNS that occurs in young children. It has histologic features of a primitive, densely cellular neuroepithelial tumor with numerous ependymal rosettes. Approximately 75% of ependymoblastomas are supratentorial, and most are large (3-11 cm). Although these tumors would be expected to be intra- or paraventricular (as in our case), most have been separate from the ventricular wall and presumably have arisen from ectopic cells committed to ependymal differentiation. These tumors share with other primitive neuroectodermal tumors the propensity for leptomeningeal seeding. The median survival is approximately 1 year [1].

The unique feature of this case is the demonstration of tentorial traversal. Primitive neuroectodermal tumors such as retinoblastoma or olfactory neuroblastoma may erode through bone to invade the base of the brain. Although other primitive neuroectodermal tumors and malignant gliomas may invade the dura and calvarium, traversal of the tentorium by an intraaxial tumor is rare.

The CT and MR findings in our case are otherwise similar to those of other primitive neuroectodermal tumors [2, 3]. The differential diagnosis includes ependymoma, astrocytoma, and choroid plexus papilloma or carcinoma [4].

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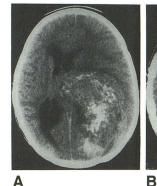
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Fig. 1.-Tentorial traversal by ependymoblastoma

A, Unenhanced CT scan shows a large heterogeneous supratentorial tumor conforming to shape of a dilated left lateral ventricle. Surrounding edema is minimal.

B, Contrast-enhanced CT scan shows mass invading left side of posterior fossa.

C, Coronal T1-weighted MR image, 600/20/2, shows tumor extending through a defect in left tentorium cerebelli. A large tumor vein (arrow) appears as a tubular area of signal void.





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