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Ependymoblastoma: MR presentation

A case report and review of the literature*

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Abstract. Ependymoblastoma is a rare and devastating tumor of childhood considered by most to be a subtype of primitive neuroectodermal tumors. We present the first detailed MRI description of this entity with a review of the limited pathologic and radiologic literature concerning this tumor.

In 1970, Rubinstein [1] redefined ependymoblastoma as a primitive malignant glioma that differentiated with ependymal cell characteristics. Few reported cases in the literature actually conform to this definition [1–7]. We believe this to be the first detailed MRI description of this rapidly fatal tumor.

Case report

The patient was a 21-month-old white male, previously in excellent health. His parents had noticed, approximately 3 weeks prior to admission, that the child was walking with difficulty. He was without any other symptoms and had up to then had a normal growth and development.

Physical examination revealed a well-nourished 21-month-old boy who was talkative and playful. Upon ambulation, his left leg was noticed to turn out and lag with increased flexion at the hip. Examination of the left lower extremity demonstrated decreased tone compared with the right, associated with a positive Babinski sign and a slight increase in the left patellar reflex without clonus. The rest of the physical examination was unremarkable. MRI of the entire spine was within normal limits.

The patient was noted within the week to have an increase in left lower extremity signs and new left upper extremity weakness. Evaluation was redirected towards the possibility of an upper motor lesion, and MRI of the head was performed.

MRI of the head demonstrated a well-defined mass in the right frontoparietal cortex measuring 4.1 cm in maximal diameter which abutted the falx medially and displaced the right lateral ventricle inferiorly. The mass demonstrated heterogeneous low signal intensity on T1-weighted images (Fig. 1 a). T2-weighted images (Fig. 1 b) demonstrated increased signal intensity with no observable vasogenic edema. Post-gadolinium T1-weighted images (Fig. 1 c) revealed mild central heterogeneous enhancement. The patient subsequently underwent surgical excision with removal of the bulk of the very friable and soft tumor.

Histologic examination revealed an extremely cellular malignant neoplasm composed of cells with round to ovoid nuclei and scant cytoplasm set in a finely fibrillar neuropil-type matrix. Nuclear chromatin was finely granular and evenly dispersed and nucleoli were generally absent or inconspicuous. Mitotic activity was very high and was accompanied by individual cell necrosis and larger scattered zones of necrosis.

The neoplasm had the general appearance of a primitive neuroectodermal tumor (PNET). However, in addition to Homer Wright rosettes and perivascular rosettes, the neoplastic cells formed scattered "true rosettes" of ependymal type with a clearly defined central canal (Fig. 2). The cells making up the true rosettes exhibited mitotic figures.

Immunohistochemical markers for glial fibrillary acidic protein, neuron-specific enolase, synaptophysin, and neurofilament proteins were focally positive. Electron microscopic studies disclosed elaborate intercellular junctional complexes associated focally with cytoplasmic bodies consistent with blepharoplasts. These ultrastructural features are consistent with ependymal differentiation. The pathologic diagnosis was ependymoblastoma.

Discussion

Rubinstein [1, 2] has defined the ependymoblastoma as a special type of embryonal central nervous system neoplasm arising in young subjects and in which the cytologic features of a primitive, highly cellular neuroepithelial tumor are associated with the presence of numerous ependymal rosettes. A recent revision of the World Health Organization classification of central nervous system tumors [8] places ependymoblastoma in a group of PNETs with the capacity to differentiate along one or more cell lines, in this case with ependymal differentiation (WHO histological typing = 1.9.3).

* The opinions or assertions contained herein are the private views of the authors and are not to be construed as official or reflecting the views of the Department of the Army or the Department of Defense.

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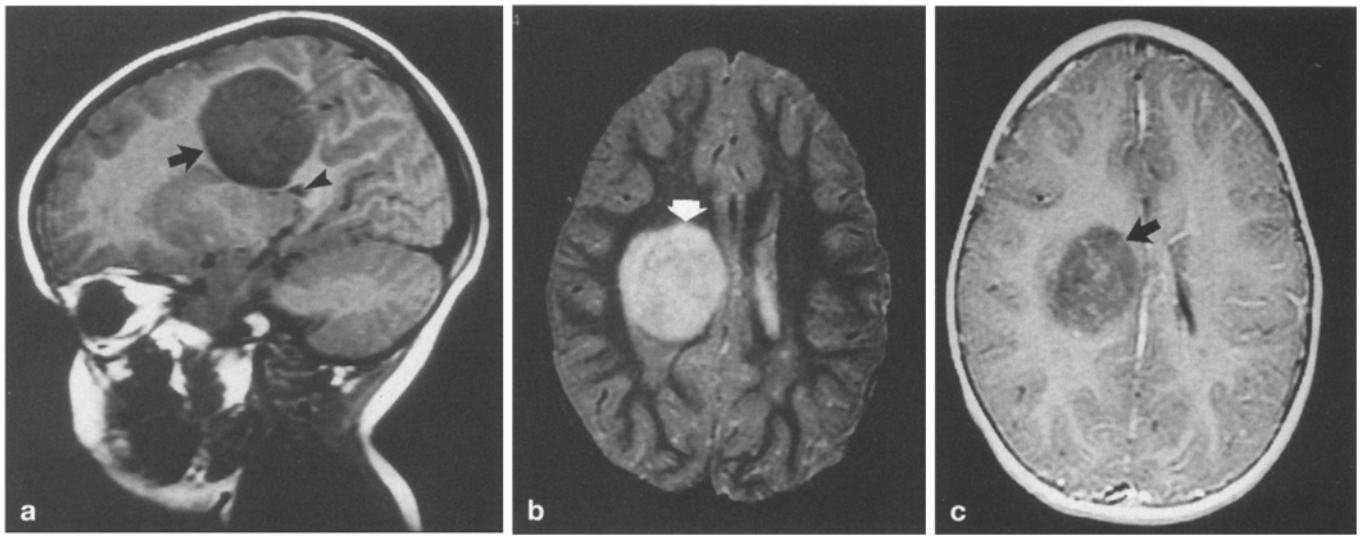

Fig. 1. a T1-weighted sagittal (SE, TE = 16, TR = 400) image demonstrates sharply marginated low-intensity mass *(arrow)* with displacement of the right lateral ventricle *(arrowhead)* and extension to the ventricular surface. **b** T2-weighted axial (SE, TE = 80, TR = 2016) image demonstrates increased signal intensity with heterogeneity of mass *(arrow)* and paucity of surrounding vasogenic edema. **c** Post gadolinium-DTPA T1-weighted axial (SE, TE = 11, TR = 366) image demonstrates mild central heterogeneous enhancement of the mass *(arrow)*

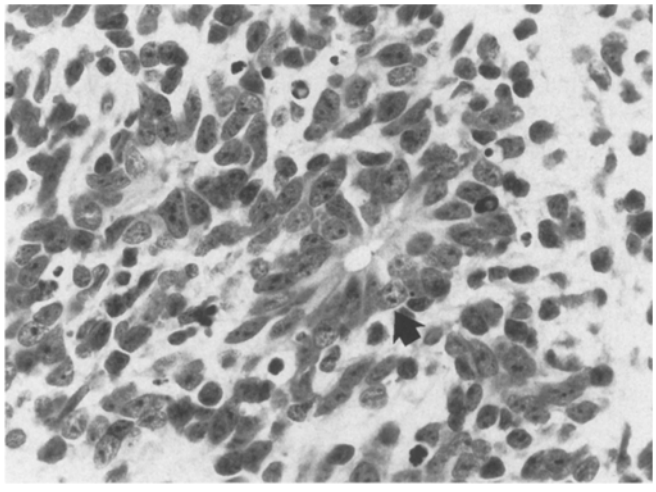

Fig. 2. "True rosette" *(arrow)* with central canal indicates ependymal differentiation. Hematoxylin-eosin, original negative magnification × 150

Within this classification, ependymoblastoma has clear cytogenetic and histologic characteristics which allow its differentiation from medulloblastomas and other PNETs [3, 8, 9]. Clear differentiation should be made between ependymoblastoma and malignant ependymoma, a term referring to mature ependymomas with severe anaplastic differentiation (often referred to as ependymoblastoma in the past) [1, 2].

In 1985, Mork and Rubinstein [2] reviewed the five cases in the literature and seven new cases. The age range at presentation was from birth to 36 years, with a median of 2 years, ten of 12 cases presenting in the first 5 years of life. Eight of the cases were supratentorial. Of these, one occupied the lateral ventricle and four were related to the ventricular ependymal lining. The rest, however, were clearly separate from the ependymal surfaces of the lateral ventricles. This is often explained by the existence of ectopic nests of cells committed to ependymal differentiation [2, 7]. Cruz-Sanchez et al. [3] report histopathologic findings on five separate ependymoblastomas, of which four were supratentorial; all were related to a ventricle.

These tumors are usually massive at the time of diagnosis (3–11 cm), yet well circumscribed [2]. The younger patients tend to have larger tumors. Monteferrante et al. [4] and Mork and Rubinstein [2] each report one case where the tumor crosses the tentorium. Occasionally, cystic changes may be identified within the tumor mass [2]. In their review, Mork and Rubinstein [2] found areas of necrosis in four cases and hemorrhagic foci in four.

Ten of 12 patients in the review by Mork and Rubinstein [2] demonstrated seeding of the leptomeninges, a feature shared with PNETs [10, 11], three with widespread deposits in the cerebrospinal pathways. One of these patients had pulmonary metastases, and Shyn et al. [6] recently reported a case with pulmonary as well as widespread cerebrospinal involvement. Ependymoblastomas display an extremely aggressive course with a mean survival of 1 year after surgery [2].

CT and MRI descriptions of ependymoblastoma are virtually nonexistent in the literature due to to its rarity and its only recent histologic distinction from other PNETs. Pigott et al. list CT findings of one ependymoblastoma [5], in a series of PNETs, as an isodense mass with heterogeneous enhancement, large mass effect, necrotic change with moderate calcification, minimal edema, and no areas of hemorrhage. These findings are very similar to those reported for the PNETs [4, 5, 10–13].

Monteferrante et al. [4] present MRI images of a case with large irregular areas of hemorrhage. Although a detailed description is not offered, the authors state

their MRI findings to be similar to those of Figueroa et al. [13] for four *undifferentiated* PNETs. As with the case presented herein, Figueroa et al. [13] describe heterogeneous gadolinium enhancement and increasing signal intensity from T1- to balanced to T2-weighted images. As this case demonstrates, necrosis, cystic change, and hemorrhage are histologically and radiographically not consistent findings.

In conclusion, although ependymoblastomas can be distinguished from PNETs histologically, there is no clear distinction by imaging criteria. A large, clearly demarcated supratentorial mass with remarkably minimal edema and heterogeneous enhancement in a young child should suggest this category of tumor and a further search of the CSF spaces by MRI. The differential diagnoses will include, in addition to PNET, other enlarging supratentorial tumors such as astrocytoma, oligodendroglioma, ependymoma, teratoma, and mesenchymal tumors. Close association with a ventricle may allow mention of ependymoblastoma as an entity; however, the absence of proximity to a ventricle does not exclude ependymoblastoma as a diagnosis.

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