Ependymoblastoma: CT and MRI Demonstration

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Ependymoblastoma is a rare and highly malignant brain tumor considered to be a subtype of primitive neuroectodermal tumors. We presented a case of ependymoblastoma in a 14-year-old girl. CT and MRI showed a large, supratentorial, paraventricular tumor with cystic and irregularly enhancing solid components. Foci of intratumoral hemorrhage and absence of peritumoral edema were also demonstrated. We claimed that ependymoblastoma should be included in the differential diagnosis of a well-circumscribed, large, supratentorial tumor without perifocal edema in a child or young adult similar to this case.

Key words: Computed tomography, Ependymoblastoma, Magnetic resonance image

Ependymoblastoma was defined as a malignant glioma that differentiated with ependymal cell characteristics [1,2]. It is presumed to arise from periventricular neuroepithelial precursor cells. The term 'ependymoblast' implies an incompletely differentiated ependymal cell showing some phenotypic features of ependyma together with immature characteristics, such as a high nucleus to cytoplasmic ratio, dense chromatin and mitotic activity. A recent revision of the World Health Organization classification of central nervous system tumors [3] places ependymoblastoma in a group of primitive neuroectodermal tumor (PNET) with the capacity to differentiate into ependymal cells. Within this classification, ependymoblastoma has clear histologic characteristics, which allow its differentiation from medulloblastomas and other PNETs [3-5]. It can also be distinguished from ependymoma, a term referring to mature ependymal cells with anaplastic differentiation [1, 2]. Some cases in the literature conformed to this definition have been reported [1,2,4,6-9], but only a few of them had imaging findings [6,7,10,11]. We herein report another case of ependymoblastoma with an emphasis on the CT and MRI findings.

CASE REPORT

This 14-year-old girl had a 1-month history of intermittent headache, nausea and vomiting. The headache always attacked during the midnight. She also complained of progressive weakness and clumsiness of the right upper extremity. Neurologic examination showed mild right hemiparesis with the muscle power of the right upper extremities of grade 4. Positive pronator sign was noted in the right hand, and papilloedema was found in the bilateral eye grounds.
Cranial CT showed a large heterogeneous mass with cystic and irregular solid components in the left parietooccipital area (Fig. 1). MRI showed that the mass was seemingly well-defined and had a close proximity to the left lateral ventricle. Speckled areas of hyperintensity were seen in the solid component of the mass on T1-weighted images. No surrounding brain edema was noted. Following administration of Gadolinium, the lesion exhibited rim enhancement in its cystic component and heterogeneous enhancement in its solid counterpart (Fig. 2).

She underwent left parietal craniotomy and grossly total tumor resection. During surgery, the tumor was measured about 5x5x6 cm in size. Its cystic portion contained yellowish fluid while its solid counterpart was friable, gray, and greasy, with foci of hypervascularity. Some poorly capsulated suable necrotic tissue was noted. Microscopically, the tumor was densely cellular and composed of uniform cells with hyperchromatic nuclei. Some small central-lumen rosettes (true rosettes) were present (Fig. 3). Scattered areas of mitotic figures and hemorrhage were also noted. After the surgical intervention, she underwent radiotherapy and then chemotherapy. However, local tumor recurrence and spinal drop metastasis developed after 6 and 15 months respectively, and she finally died 23 months after presentation.

**DISCUSSION**

Ependymoblastoma is a special type of embryonal central nervous system neoplasm arising in young subjects. The chief histological characteristic of ependymoblastoma is the dense cellularity with distinctive and numerous ependymal rosettes. These rosettes form concentric cellular rings around small round lumina [1,2]. The histological differential diagnosis mainly includes anaplastic ependymoma and medulloblastoma. The former is characterized by the prominent perivascular pseudorosettes, while the latter is distinctive by its cerebellar location and is characterized by Homer-Wright rosettes which tumor cells cluster around a fibrinoid matrix.

Ependymoblastoma occurs in young children including neonates. In the first and second year of life, the most common clinical manifestation is increased intracranial pressure and hydrocephalus. Focal neurological deficits are more common in older children. Males and female appear to be equally affected. In their review of 12 cases of ependymoblastoma, Mork and Rubinsten [2] found that the age ranged at presentation was from birth to 36 years, with a median of 2 years. Most tumors were well circumscribed and large at the time of diagnosis (3-11 cm). Eight of the cases were supratentorial. Of these, five merged with the ventricular ependymal lining. The rest, however, were clearly separated from the lateral ventricles, suggesting the existence of ectopic nests of cells committed to ependymal differentiation [2, 9].

Imaging descriptions of ependymoblastoma are rare in the literature due to its rarity and its only recent histologic distinction from other PNETs. The CT and MRI appearances of ependymoblastoma have been described as a large, heterogeneous, well-circumscribed supratentorial mass with no significant surrounding edema. Intratumoral calcification or hemorrhage may be found [6,7,10,11]. Such imaging appearances are similar to those of other PNETs [12-15]. Diagnostic possibilities also include other enhancing supratentorial tumors such as astrocytoma, oligodendroglioma, and ependymoma. In our case, most of the typical...
Figure 3. Photomicrography shows dense hyperchromatic cells with true rosettes around the canal structures (arrows). (Hematoxylin and eosin, X350)

Figure 2. a. Axial T1-weighted image (TR/TE = 366/16), b. axial T2-weighted image (3250/90), c. coronal T2-weighted image (3250/90), d. enhanced axial T1-weighted image (433/26), and e. enhanced coronal T1-weighted image (433/26) demonstrate a seemingly well-defined mass (arrows in a, b, c, d, and e) composing of a cystic component with rim enhancement and an irregular solid component with heterogeneous enhancement. Speckle areas of hyperintensity are noted in the solid portion of the mass on T1WI, corresponding to focal areas of hemorrhage (arrowhead). Note close proximity of the lesion to the left lateral ventricle and lack of edema in the adjacent brain parenchyma.

imaging findings of ependymoblastoma were well demonstrated. In particular, MRI clearly demonstrated internal hemorrhage and total absence of peritumoral vasogenic edema while CT was helpful to exclude the presence of intratumoral calcification. The internal hemorrhage is a common finding of malignant tumors and can lessen the likelihood of benign tumors such as pleomorphic xanthoastrocytoma. Other imaging findings, such as large size, supratentorial location relating to the lateral ventricle, heterogeneous enhancement, and lack of peritumoral edema in this young patient may give a clue to include ependymoblastoma in the differential diagnosis.

Ependymoblastoma displays an extremely aggressive course with rapid growth and widespread cerebrospinal dissemination. The
median survival is approximately 1 year [2]. Development of effective treatment protocols for ependymoblastoma is limited by its rarity and aggressive tumor behavior. The role of total resection, post-operative irradiation and adjuvant chemotherapy remains inconclusive. Further investigation with accumulation of cases is needed. Our patient underwent a gross total resection, postoperative radiotherapy and chemotherapy treatment, and died 23 months after presentation.

**REFERENCES**

室管膜胚細胞癌：電腦斷層攝影及磁振造影表現

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室管膜胚細胞癌為罕見及高度惡性之腦瘤，為原始神經表皮腫瘤之一種次類型。本文報告一名十四歲室管膜胚細胞癌患者，其電腦斷層攝影及磁振造影顯示一個大腦天幕上、腦室旁之腦瘤，內有囊腫成份及不規則顯影加強之固體成份，同時也顯示局部腫瘤出血及缺乏腫瘤旁水腫。我們認為當年青腦瘤病人有類似影像表現時，應把室管膜胚細胞癌列入鑑別診斷內。

關鍵詞：電腦斷層攝影，室管膜胚細胞癌，磁振造影