Case Report

A cystic anaplastic ependymoma mimicking a pilocytic astrocytoma

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A B S T R A C T
Ependymomas are grouped in 4 subtypes based on their cellular characteristics including subependymoma, myxopapillary ependymoma, classic ependymoma, and anaplastic ependymoma. Among those, anaplastic ependymoma are the most malignant and fast-growing tumors as grade III according to the fifth edition of the WHO classification of tumors of the central nervous system. Commonly, ependymoma is predominantly solid. The cystic ependymoma is very rare. In this paper, we aimed to introduce a 2-year-old male with cystic anaplastic ependymoma inside fourth ventricle which was misdiagnosed as a pilocytic astrocytoma. We recommend that the possibility of anaplastic ependymoma should be taken into account in every case of infratentorial cyst inside the fourth ventricle without mural nodule and with rim enhancement.

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Introduction

Ependymal cells that line the cerebral ventricles and the spinal cord's central canal are considered to be the source of ependymomas. Although they can develop anywhere along the neuraxis, they are most frequently detected in children's posterior fossa and adults' spinal cord. Cystic ependymomas are uncommon tumors of the central nervous system that can resemble other primary central nervous system tumors including pilocytic astrocytomas or hemangioblastomas in appearance. These factors make them frequently challenging to diagnose [1–3]. We describe a rare instance of cystic anaplastic ependymoma in a child which was misdiagnosed as a pilocytic astrocytoma.

Case presentation

A 2-year-old male, complaining of headache and vomiting for 1 month, was admitted to Children’s Hospital 2. The medical history of the patient was normal. The neurological examination was unremarkable, without focal neurological deficits, and laboratory tests were within normal limits. Then, the patient underwent a 1.5-Tesla brain MRI, with a contrast agent. Hydrocephalus in the supratentorial region was identified. On axial T2-weighted imaging, a hyperintense cystic mass, deriving from the fourth ventricle, was clearly observed, without surrounding vasogenic edema (Fig. 1). The right-to-left (RL), anterior-to-posterior (AP), and feet-to-head (FH) diameters of the mass were 36, 45, and 52 mm, respectively.
Neither ossification nor hemorrhage was detected within the mass, on susceptibility-weighted imaging. The mean apparent diffusion coefficient (ADC) of this mass was 0.37 (Fig. 2). On T1-weighted imaging, with contrast enhancement, the mass appeared heterogeneously enhanced with clearly rim enhancement (Fig. 3). On diffusion tensor imaging, the fractional anisotropy value of tumor was 0.33 (Fig. 4). On MRI spectroscopy, the choline/N-acetyl aspartate ratio was 0.48 (Fig. 5). Serological tumor markers were not assessed. The initial preoperative diagnosis, in this case, based on the clinical and radiological information, was determined to be pilocytic astrocytoma. The patient underwent a radical tumor eradication surgery and was diagnosed, histopathologically, as an anaplastic ependymoma. The patient was released after 2 weeks and proceeded to pursue adjuvant chemotherapy and radiotherapy at a separate oncological center.

Discussion

Pilocytic astrocytoma, which peaks between the ages of 5 and 15, is a low-grade brain neoplasm that frequently exhibits the characteristic radiological signs of a cyst with the mural nodule. The posterior fossa is where these tumors are most frequently discovered, and on T1-weighted imaging, the cyst content is typically mildly hyperintense to cerebrospinal fluid [4,5]. Pleomorphic xanthoastrocytoma (peak age 10-30 years), ganglioglioma (peak age 10-20 years), and desmoplastic infantile ganglioglioma are other tumors with cysts that resemble mural nodules (peak age 1-2 years). On T1-weighted MRI, the cystic component of these tumors, which are often located in the supratentorial compartment, is hypointense. Metastatic tumors can very infrequently also appear as a cyst with a mural nodule [4,5]. This is possible the first instance of infratentorial anaplastic ependymoma in a child that has been documented as a pure cyst with rim enhancement and without mural nodule. Some previous papers reported that two cases of anaplastic ependymoma were heterogenous masses with cyst and mural nodule [2,4].

Ependymomas account for 4% of all adult central nervous system tumors [2]. They often happen in the infratentorial compartment and are more frequent in youngsters. Astrocytoma, medulloblastoma, and tumors of the choroid plexus are among the possible diagnoses for malignancies in the posterior fossa [6,7]. Histopathological findings were marked hypercellularity, nuclear atypia and brisk mitotic activity. It may also have intramural or glomeruloid vascular pro-
liferation, pseudopalisading necrosis, perivascular rosettes. Discrete or infiltrative margin is common. Noted that nonpseudopalisading necrosis is not sufficient for diagnosis in otherwise low grade ependymoma [8,9].

Surgery is the first line of defense against anaplastic ependymomas, if at all possible. The purpose of surgery is to acquire tissue in order to identify the kind of tumor and to remove as much of the tumor as is feasible without increasing the patient’s symptoms. There is no conventional therapy for anaplastic ependymomas after surgery. Chemotherapy and/or radiation therapy may be used as adjuvant therapies [10].

Fig. 3 – Diffusion tensor imaging of the tumor showed the fractional anisotropy value of tumor was 0.33.

Fig. 4 – Axial (A) and sagittal (B) and T1-weighted images with contrast agent revealed the cystic tumor inside the fourth ventricle (arrow) with clearly rim enhancement.
Fig. 5 – Magnetic resonance spectroscopy of the tumor revealed that the Cho/NAA ratio was 0.48.

### Conclusion

To sum up, anaplastic ependymoma is a rare tumor that, in rare cases, appears on imaging as a solitary simple cyst without a mural nodule. In each instance of infratentorial cyst inside the fourth ventricle without mural nodule and with rim enhancement, the risk of anaplastic ependymoma should be considered.

### Data sharing statement

Not applicable.

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### Patient consent

Informed consent has been obtained from the family members of the patient included in this study.

### Ethics approval

This study has been approved by the hospital ethics committee (Ref: 352/ND2-CDT).

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