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Ectopic cortical anaplastic ependymoma: An unusual case report and literature review

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ABSTRACT

Ependymona occasionally occurs outside the ventricular structures, which is called ectopic ependymona (EE), while pure cortex location is uncommon. However, cortical anaplastic ependymoma (CE) is rare, especially in children. There were only four primary CEs, which is located in the superficial cortex, were reported the age of the patient under 12 years old. The present case is a 20-month-old boy presenting with simple partial seizure was treated in our department. Cranial magnetic resonance imaging (MRI) revealed a fronto-parietal lobe mass of more than 50 mm in diameter with mixed signal intensity. Total removal of the mass lesion was performed without any neurological deficit. Pathological examination of the excised tumor were consistent with anaplastic ependymoma (AE). The patient had a good recovery after his surgical resection. Radiotherapy and chemotherapy were not taken into account in view of his age, the favorable site and the complete resection. The management of this unusual tumor is summarized in this paper.

1. Introduction

Ependymomas are common in the fourth ventricle in children. Less than one-third are found in the supratentorial region. Although it is a kind of lesion derived from the ventricular system, ependymonas occasionally may appear outside the ventricular structures, representing a rare group of ectopic ependymonas (EEs). Anaplastic ependymoma (AE) has been recognized as World Health Organization (WHO) grade III and defined on the basis of their pathological features. According to my knowledge, only four cases of cortical anaplastic ependymomas (CAEs) under 12 years old were reported in English literature (Table 1). Here we report one case of CAE in a only 20-month-old boy. The management of this unusual tumor is summarized in this paper.

2. Case presentation

A 20-month-old boy was referred to our department with intermittent seizures for more than one month. He experienced complex partial seizures, at a frequency of three to four per week. The physical examination was normal. Magnetic resonance imaging (MRI) of the brain with and without contrast exhibited a mass that was $5.2 \times 4.1 \times 2.7$ cm in size at the convexity of right-sided fronto-parietal lobe with cystic and solid appearance, and remarkably peritumoural edema. The tumor (mixed T1 and T2 intensity signal, fluid-attenuated inversion recovery (FLAIR): isointense and hyperintense) had heterogeneous enhancement after gadolinium injection (Fig. 1). No additional tumor was observed in the central nervous system. The patient continued to have seizures to medication.

A right fronto-parietal craniotomy was performed with total microsurgical tumor removal and without any neurological deficit. The tumor was vascular, well-circumscribed, grayish-red in color, and a cortical solid and cystic mass without connections to the dura or ventricle. Histologically, the tumor was hypercellular, with cellular and nuclear pleomorphism and perivascular pseudorosettes (Fig. 2A,B). Necrosis and calcifications were apparent in the cut surfaces. Immunohistochemical findings illustrated that the neoplastic cells were positive for glial fibrillary acidic protein (GFAP), vimentin and epithelial membrane antigen (EMA) (Fig. 2C–E). The proliferation index was approximately 25% by Ki-67

Abbreviations: EE, ectopic ependymona; CE, cortical ependymoma; AE, anaplastic ependymoma; CAE, cortical anaplastic ependymoma; WHO, World Health Organization; HE, hematoxylin and eosin stain; MRI, magnetic resonance imaging; GFAP, glial fibrillary acidic protein; EMA, epithelial membrane antigen; Lt, left; Rt, right; RT, radiotherapy; GTR, gross total resection.

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Table 1
All reported cases of cortical anaplastic ependymomas under 12 years.

First author	Sex/age (years)	Location	Presentation	Treatment	Followup (M)
Alexiou GA	F/10	Lt. frontal	Headache, seizures	GTR + RT	?
Lehman NL	F/1	Rt. frontal	Seizures	GTR	48
Van Gompel JJ	M/12	Rt. parietal	Seizures	GTR + RT	101
Z Liu	M/2	Lt. frontal	weakness	GTR	48
Present case	M/1.7	Rt. frontoparietal	Seizures	GTR	6

Lt: left; Rt: right; RT: radiotherapy; GTR: gross total resection; M: months.

(Fig. 2F) staining. The pathological examination of the excised tumor were consistent with AE (WHO grade III). The boy had a favorable outcome after surgery and continued to regularly take antiepileptic drugs. There is no any adjuvant chemotherapy or radiotherapy was given considering a gross total resection and his age. There has been no radiologic evidence of residual lesion and distant metastasis on the 6-month follow-up MRI (Fig. 3).

3. Discussion

Ependymomas are neuroectodermal tumors arising from the ependymal lining of the cerebral ventricles or the central canal of the spinal cord [1]. The majority of ependymomas in children emerge in the infratentorial region. Supratentorial ependymomas lie in the third or lateral ventricle. Cortical ependymoma (CE) which has been speculated to be EE originating from embryonic remnants of ependymal tissue and primarily located in the cerebral cortex, is relatively rare [2]. CAE, as in our patient, is extremely infrequent [3]. There are about 40 patients with CE reported in the literature [4]. To date, less than 18 cases of AE reported are situated in cerebral cortex, representing uncommon cases of ectopic lesions, While there total of 5 cases (including present case) in children under 12 years old, are CE.

Epilepsy as the presenting symptom has been considered a good prognosis, which makes early discovery of brain tumor possible. Furthermore, it appears to be easily caused by benign tumors [5]. Though epilepsy in our patient without symptoms of raised intracranial pressure and focal signs, was only clinical manifestation, the neoplasm diagnosed as AE was malignant. The CAE has relatively typical radiographical traits. These lesions on MRI usually appear iso-to hypointense on T1 and hyperintense T2 -weighted images whereas image in our case is mixed signal, and have relatively distinct margins [3,6]. Brain edema is unremarkable in the 4 cases (Table 1), which is different from our case. Necrosis and calcification in most tumors with variable enhancement are almost visible. EEs are generally small in size [7]. However, the CAE in our case is more than 5 cm in the maximum diameter and positioned in two lobes of the brain surface, unlike these four reported cases with CEA confined to one lobe.

No definite consensus exists regarding the treatment of these lesions [8]. CEs are more amenable to gross total resection than infratentorial and ventricle ones [3]. They have an indolent behavior and are not prone to local recurrence [7]. However, it is not clear if their indolent course counts on an earlier finding due to cortical involvement, their accessibility in addition to resectability, or their intrinsic pathobiology [9]. The cortical mass in our patient who did not go through any following therapy after surgery, was totally removed. The need for adjuvant radiotherapy for patients with supratentorial extraventicular ependymomas is a controversial issue. Early studies stressed the potential role of radiotherapy in WHO grade III ependymomas [7]. Niazi et al. held the kind of viewpoint that all patients with WHO grade III ependymoma required radiotherapy [10]. Postoperative radiotherapy should be executed in those patients with residual tumor and AEs [11]. On the other hand, prophylactic whole-spinal axis irradiation offers little utility in patients with localized supratentorial ependymomas, regardless of the tumor grade [12]. Radiation therapy, as in our patient, was not carried out in consideration of his age, the extent of excision and the favorable location. As the effect of chemotherapy in the treatment of ependymomas is concerned, it plays a limited role [13]. No clinical study is in favor of the evidence that these tumors respond to chemotherapy.

The grade of the tumor and extent of surgical resection have been regarded as important prognostic predictors of survival, because patients with gross total resection tend to have better outcome than those with subtotal resection [12]. A number of prognostic factors for ependymomas have been described in the literature, including patient age, tumor location, histology and extent of resection [14]. Only tumor total resection is considered as a reliable prognostic factor for predicting long survival time [1].

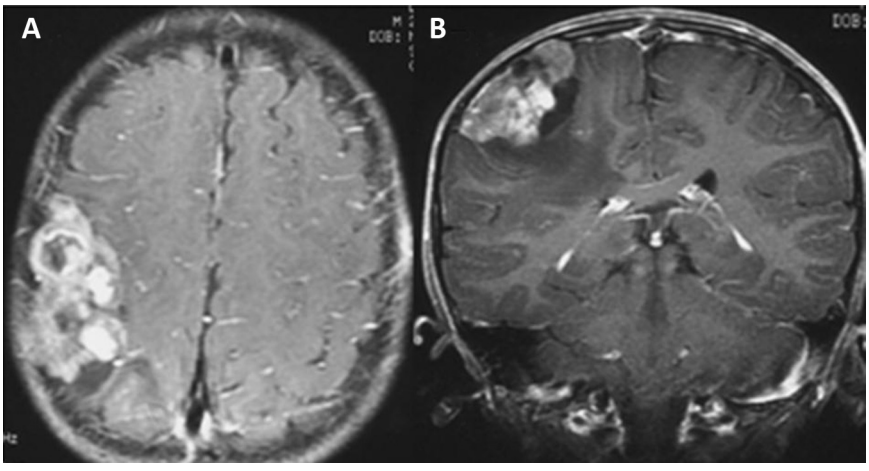

Fig. 1. Preoperative contrast-enhanced axial (A) and coronal (B) T1-weighted image showing a mass in the right fronto -parietal lobe with mixed signal intensity.