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Case Report

Diagnosis of intracranial embryonal carcinoma by cerebrospinal fluid cytology: a case report

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Abstract: Background: The incidence of primary intracranial germ cell tumors (GCTs) is relatively low comparing to other ones. Embryonal carcinoma (EC) is an especially rare subtype and the diagnosis presents to be a challenge. Few cases have been reported. Case presentation: We report a case of intracranial EC located in the temporal lobe with malignant tumor cells occasionally detected by the cytology of cerebrospinal fluid (CSF). The pathology confirmed the diagnosis after the patient underwent tumor resection. Conclusion: This is the first report about one case of intracranial primary EC located in the temporal lobe. It is also the first report of tumor cells of EC detected in the CSF.

Keywords: Embryonal carcinoma, temporal lobe, cerebrospinal fluid cytology

Introduction

The incidence of intracranial germ cell tumors (GCTs) is rare, accounting for 0.5% of primary brain tumors [1]. Tada (1997) has reported GCTs accounted for 4.9% of the 2284 cases of intracranial tumors in Japanese, germinoma accounted for 70.5% and teratoma 13.4%, while other malignant GCTs including EC were more rare [2]. Primary intracranial GCTs are encountered predominantly in the pineal and suprasellar regions, secondly in the basal ganglia and thalamus (76-90% of cases) [3]. However, reports of malignant GCTs especially the EC involving the lobes of brain, are extremely rare.

Because of the potential morbidity associated with the procedures targeting the pineal region or other deep brain tissues for biopsy or surgical treatment, detection of tumor markers of CSF is still an important complementary method for the diagnosis of intracranial GCTS [4-6]. Elevated levels of tumor markers such as alpha1-fetoprotein (AFP), and beta-human chorionic gonadotropin (β-hCG), are commonly applied [7].

The cytology of CSF is a useful diagnostic procedure in the evaluation of patients with neurologic disorders [8]. However, primary central nervous system (CNS) tumors are only occasionally encountered in the CSF because of their frequent location within the CNS parenchyma. Therefore, there are only a few reports dealing specifically with the cytology of primary CNS tumors [8-12]. We found the tumor cells of intracranial EC in the CSF and then confirmed by pathology after the surgery, which is the first case.

Case presentation

A 19-year-old boy without prior health problems suffered from drowsiness for 13 days, and had a headache mainly located in the right temporal region for 8 days before admission. Neurological examination was normal.

MRI scan revealed a 2.5×2.5 cm mass in the right temporal lobe accompanied with deformation of lateral ventricles, hypointense on T1-weighted sequences (**Figure 1**), mild hyperintense on T2-weighted sequences, and a strong contrast enhancement with obviously

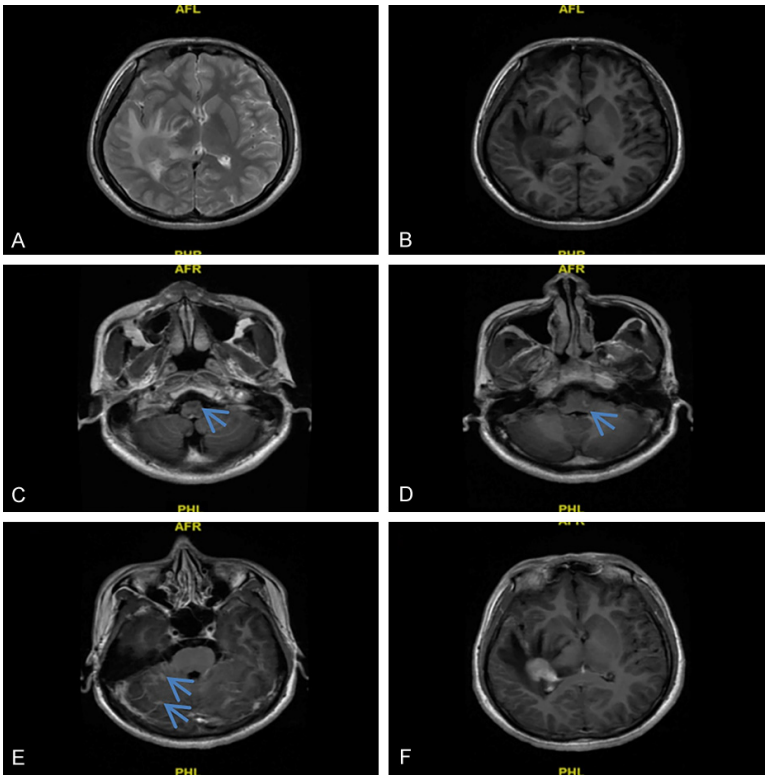

Figure 1. There is a mass lesion in the right temporal-occipital junction region, with a slightly high signal intensity on T2-weighted MRI (A) and slightly low signal intensity on T 1 (B). The lesion showed obvious inhomogeneous enhancement on contrast-enhanced T1-weighted (F), with the presence of peritumoral edema and enhanced multiple lesions in brain stem and cerebellar (C-E, blue arrow).

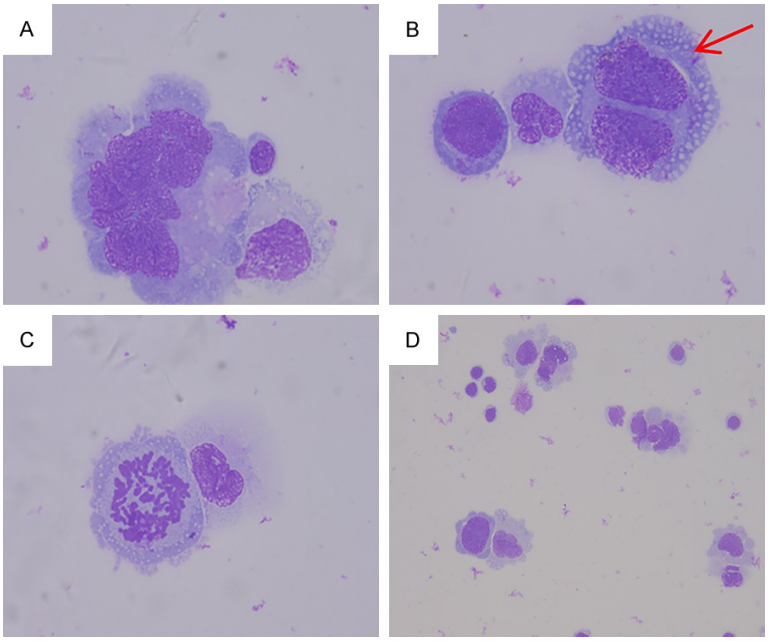

Figure 2. A. The cells were large in size with abundant cytoplasm protruding from the membrane. The nucleus was obviously atypical and stained deeply. B. The neoplastic cells had double nucleoli with biphasic cytoplasm of which peripheral staining was deeper comparatively. C. The mitosis was easily observed with abundant biphasic cytoplasm. D. Neoplastic cells were obviously polymorphous.

edema adjacent. Multiple mild-enhancing lesions were found in the brainstem and cerebellum. There was no evidence of tumor within the pineal, suprasellar region, and the basal ganglion.

The lumbar puncture showed a yellow color fluid and a fluid pressure of over 330 mm H_2O. The analysis of CSF included: white blood cells 90×10^6/L, mononuclear cells 81×10^6/L, multinuclear cell 8×10^6/L, glucose 0.1 mmol/L, chloride 112 mmol/L, and proteins 5.37 g/L.

The tumor marker β-HCG was 0.8 mIU/L in blood and 8 mIU/L respectively, The AFP were 9 mIU/L in blood and 3.5 mIU/L respectively.

After performing the cytology of CSF, we found that the malignant cells were detected (**Figure 2**).

Then, the patient had a transtemporal craniotomy for tumor resection and the tumor pathology revealed EC (**Figure 3**). The situation of the patient was getting worse and he died about one month after the surgery.

Discussion

Intracranial GCTs have classically been divided into two histologic groups: germinomas and non-germinomatous germ cell tumors (NGGCTs) [13]. EC belongs to NGGCTs and is mainly found in the pineal and suprasellar regions. A few

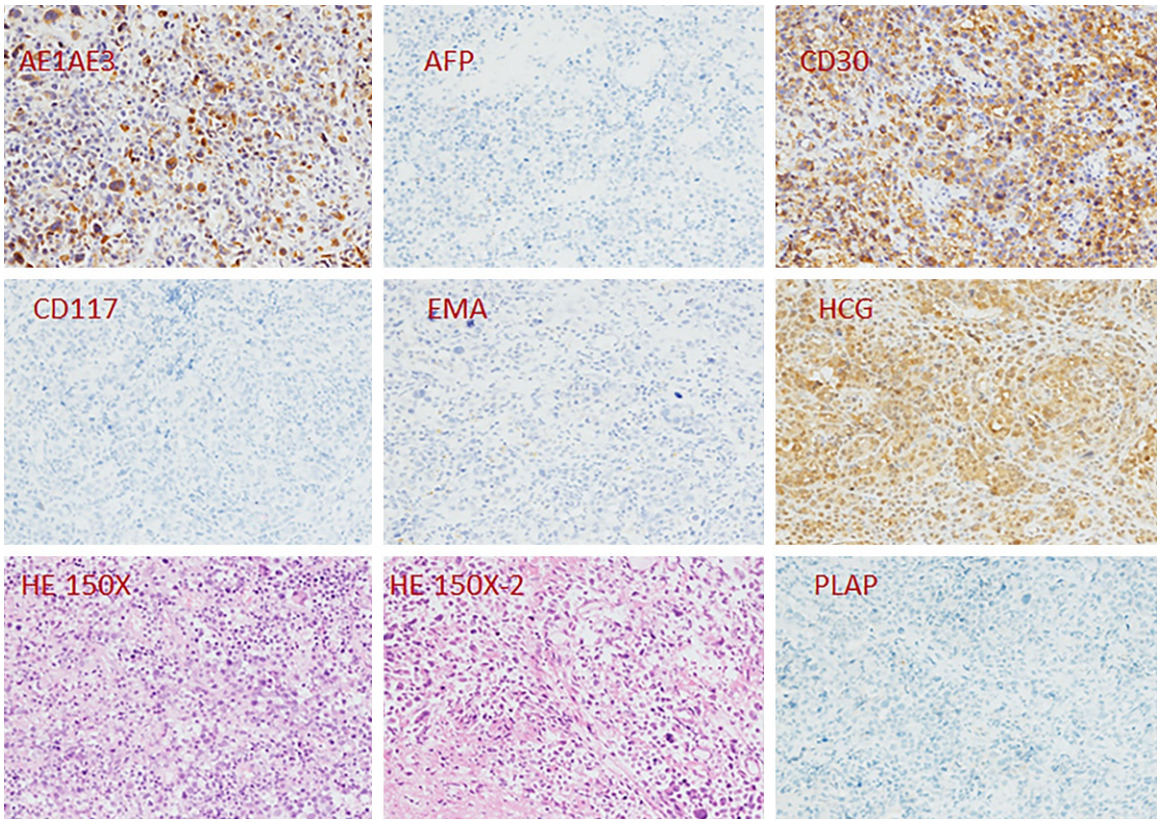

Figure 3. The slides show that the tumor cells grow in sheets or nests with Lamellar necrotic area, some of the tumor grows in solid. Most of the tumor contains polygonal cells, which have vesicular nuclei with coarse, basophilic chromatin and one or two prominent nucleoli. Most of the cell membranes are well defined and the cytoplasm is abundant and usually amphophilic. Mitotic figures and apoptotic bodies are numerous under the microscope. Syncytiotrophoblast cells are present in the solid area of the tumor. Lmmunohistochemistry: The tumour cells are typically positive for wide spectrum cytokeratin (AE1/AE3), and positive for CD30. It is negative for CD117, AFP, and PLAP. Epithelial membrane antigen is negative. The syncytiotrophoblastic giant cells are cytokeratin and HCG positive.

cases involved the third ventricle and basal ganglia have been reported [14-17]. Today we report a case of EC developing in the region of temporal lobe, which can be considered in the young patients' differential diagnosis of cerebrum masses.

Early diagnosis and treatment are the key to improve the curative effect and quality of life of patients with intracranial GCTs. It is pointed out that the earlier the diagnosis and intervention, the more significant the improvement of the disease [18]. However, the high fatality and mutilation rate associated with surgical biopsy and resection because of the location of the intracranial EC, made pathologic diagnosis unachievable.

EC may secrete some proteins into the blood and/or CSF, such as β-HCG and AFP, which can be detected as complements of the diagnosis. We also found high levels of β-HCG in CSF, which reminded us the diagnosis of GCTs. However. as an evidence for a definitive diagnosis, it is still insufficient.

Further analysis of CSF cytology with May-Grünwald-Giemsa (MGG) staining, we occasionally found the EC cells, which were confirmed by the pathology after the surgery. Cellular smears show large and variable neoplastic cells, with plenty cytoplasm even protruding from the surface of cell membranes. Aberrant nuclear morphology, such as hyperchromatic nuclei with irregular shapes signify the abnormal cell proliferation.

It is an encouraging sign because it was first reported EC cells can be observed from a non-operative means with no risks associated with

surgery. The specificity and sensitivity are certainly needed to be verified through more and more cases.

Overall, primary intracranial EC is rare with variable clinical manifestations. EC can be involved in cerebral lobe, which should be taken into account of differential diagnosis of intracranial tumors. If a lumbar puncture is allowed, CSF cytology is an effective approach to help diagnostic, guided treatment, and evaluated prognosis.

Disclosure of conflict of interest

None.

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