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Case Report

Intraparenchymal frontal lobe ependymoma without rosettes: a case report

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Abstract: Ependymomas, which are typically in the supratentorial or within the spinal cord in adults, are believed to arise from radial glial cell stem cells. They can occur throughout the neural axis, usually in close proximity to the ventricles or central canal. While the parietal lobe and temporal occipital region are common locations for supratentorial ependymomas, we present a rare case of an entirely intraparenchymal frontal lobe tumor, remote from the ventricular surface. Imaging revealed a cyst and solid cerebellar mass near the lateral ventricle. Single-voxel hydrogen proton magnetic resonance spectroscopy detected choline increasing and the N-Acetyl-L-aspartic acid decreasing. Histological features did not display the common diagnostic rosettes-morphous of clustered malignant cells.

Keywords: Intraparenchymal, ependymoma, frontal lobe, magnetic resonance spectroscopy

Introduction

Intracranial ependymomas represent about 2% of all intracranial tumors in adult patients. Imaging often reveals a well-demarcated cystic and solid mass with micro cystic components [1]. MRI of supratentorial ependymomas presents typically with high-signal on T1- and T2-weight image [2]. The histological features did not display the common diagnostic rosettes-morphous of clustered malignant cells.

Case report

A 41-year-old women presented in the emergency room with dizziness, intermittent headache, and nausea for 1 month. She had never experienced similar episodes in the past, and there were no other symptoms. There was no history of allergies, drug use, or severe heart or pulmonary disease. There was no significant family history. Physical examination revealed no pathological sign. There was no cranial nerve palsy, hemiparesis, or other neurological signs.

MRI demonstrated an incidental right-sided frontal lobe mass. Dedicated MRI of the brain showed a 47 mm solid mass in the right frontal lobe, resulting in slight distortion of the lateral ventricle but without effacement or hydrocephalus. The solid nodular component demonstrated enhancement (**Figure 1**).

Single-voxel hydrogen proton magnetic resonance spectroscopy (SVS) demonstrated a decreased N-Acetyl-L-aspartic acid (NAA) peak and increased choline (Cho) peak and creatine (Cr and Cr2) peaks (**Figure 2**), with the rate of NAA/Cr=0.34, Cho/Cr=3.18 and Cho/NAA=9.27 in the lesion. Combined with MRI, it was presumed to be a malignant tumor and most possibility a glioma.

Intraoperative exploration discovered that the mass originated from the right insular lobe, with an apparent boundary with the frontal lobe and temporal lobe. There were plenty of nourishing blood vessels between the tumor and periphery brain tissue. The character of the lesion was tenacious near the lateral cleft, while soft in other parts.

Histological section of the tumor tissue stained with hematoxylin and eosin supported the diagnosis of ependymoma with immunohistochem-

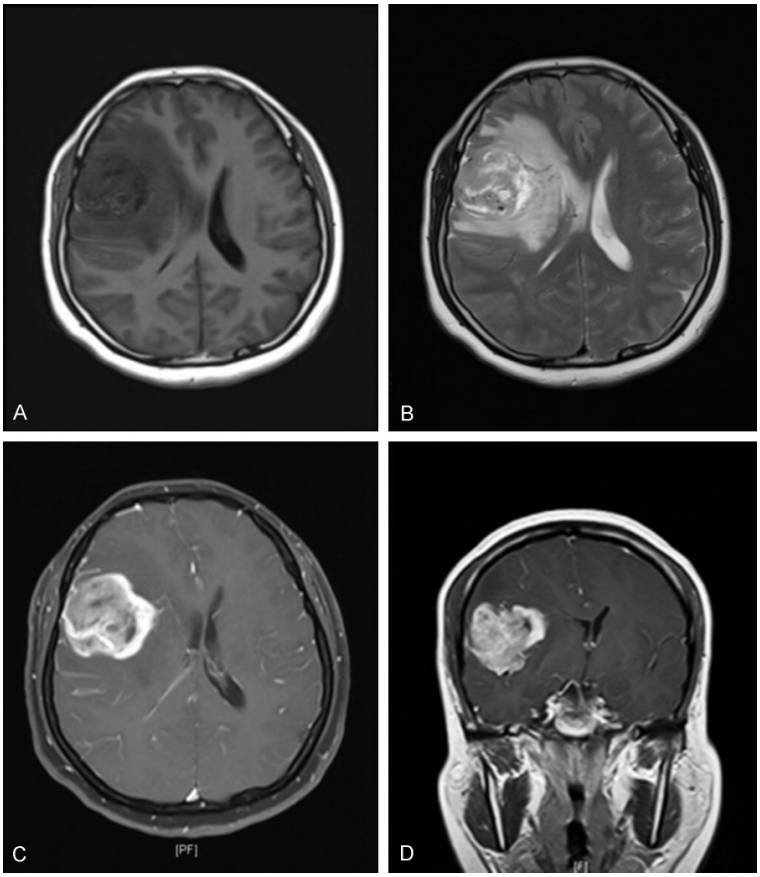

Figure 1. Preoperative MRI scan revealing a large intra-axial cystic and solid mass in the right frontal lobe producing mass effect on the ipsilateral lateral ventricle and midline, and intense contrast enhancement of the solid part of the tumor and cystic wall. A. T1-weighted imaging showing the lesion is isointense to grey matter. B. T2-weighted imaging showing the lesion consists of mixed high-intense signal. C. T1-weighted post-contrast imaging showing heterogeneous enhancement of the mass with central non-enhancing regions. D. Coronal section of T1-weighted post-contrast imaging showing the mass was remote from the ipsilateral lateral ventricular.

istry supporting of glial fibrillary acidic protein (GFAP) and S-100 positive, synaptophysin (Syn) partially positive, CD34 vessels positive, Ki-67 focally positive, as well as neurofilament protein (NF), progesterone receptor (PR) and epithelial membrane antigen (EMA) negative (**Figure 3**).

Discussion

Ependymomas, which are typically in the supratentorial or within the spinal cord in adults, are believed to arise from radial glial cell stem cells. Furthermore, glial stem cells divided ependymal cells rests remaining in the brain parenchyma and form intraparenchyma ependymomas, which have a predilection for the frontal, temporal, or parietal lobe [1]. The clinical presentation depends on the location to present with focal neurologic deficits, headache, and seizures. The frontal lobe lesion was described in 2 of 34 cases in early research [2]. Intraparenchymal ependymomas are homogenous granular, solid masses with associated cystic elements, necrotic foci, or hemorrhage. They have definite ependymal morphology on histopathology.

The solid portion of ependymoma on T1-weighted images is heterogeneous in intensity and isointense to hyper-intense on T2-weighted images. Hemispheric perilesional edema results in a mass effect, midline shift, and hydrocephalus disproportionate to the size of tumor as noted in the intraparenchymal type. MRS with short TE has been described in a series as showing increased glutamine and glutamate [3]. In a single institution study, spectroscopy at long TE=144 showed reduced NAA, Cho/Cr ratio of 4-7 and a Lac peak at 1.3 ppm which is suggestive of anaerobic metabolism [4]. Although MRS does not present with any characteristic marker in short TE, it may aid to differentiate Grade II from Grade III and in cases of recurrence [1].

Ependymomas are generally divided into three histological grades. The myxopapillary ependymomas and subependymomas are classified as Grade I tumors; the cellular, papillary, tanycytic, and clear cell subtypes are classified as Grade II tumors. The anaplastic variant is considered as a Grade III tumor. Perivascular pseudorosettes or true rosettes, where ependymal cells cluster around an empty lumen as though forming a small ventricle or spinal canal, in moderately cellular tumours are one diagnostic characters of intraparenchymal ependymomas [5]. However, the absence of these rosettes does not exclude the diagnosis as they are only

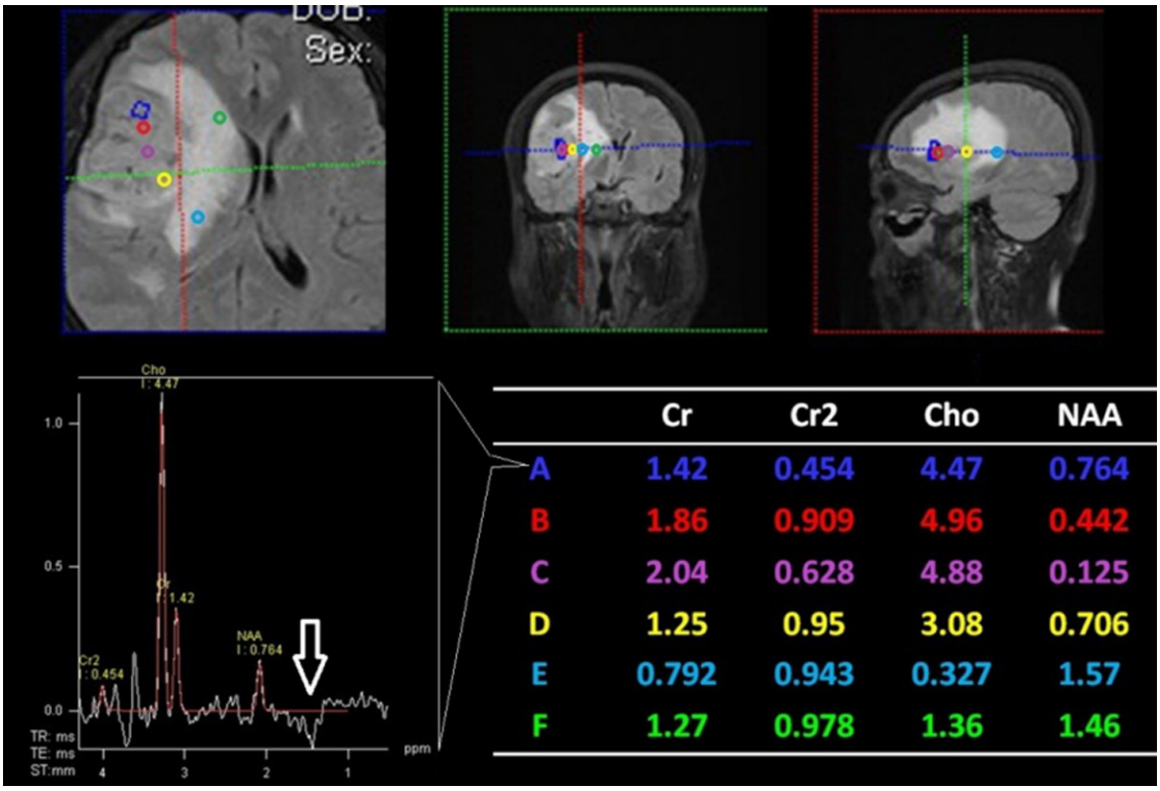

Figure 2. Single-voxel hydrogen proton magnetic resonance spectroscopy (SVS) of the lesion. From the inside (A-D) to the outside (E, F) of the tumor, the N-Acetyl-L-aspartic acid (NAA) peak obviously decreased, the choline (Cho) peak and creatine (Cr and Cr2) peak increased. From the A point, a tall Lactate (Lac) double-peak, marked with the white arrow, was exhibited from 1.2 ppm to 1.3 ppm.

seen in the minority of lesions. Furthermore, exhaustive workups with immunohistochemistry markers consisting of GFAP, S-100 and EMA are imperative for proper diagnosis. GFAP-positive cells in the fibrillary matrix, especially in the perivascular location, along with a cytoplasmic punctate and dot-like EMA positivity on IHC are the characteristics of ependymal [6].

Classically, the histological grade, site of the tumor, and age at diagnosis have remained the main prognostic factors in patients ependymoma [7, 8]. With increasing molecular biology research, better predictive and prognostic factors have been detected [9]. From the literature, many studies have identified HIC-1 methylation, 4.1B deletion, and 4.1R loss as common features in intracranial ependymoma. Supratentorial ependymoma is usually characterized by NOTCH-1 mutation and p75 expression. Although MEN1, TP53, and PTEN mutations are rarely reported in ependymoma, they may be related to poor prognosis, such as recurrence or metastasis. A new classification of ependymal tumors based on DNA methylation profiling was described in a study [10]. However, there is no specific marker that has been reported in intra-parenchymal ependymoma.

In conclusion, supratentorially intra-parenchymal ependymomas are rare and have a side spectrum of clinical and radiologic phenotypes. MRS may differentiate the histological grade but not specific for diagnosis. Accurate preoperative diagnosis is difficult but important for deciding on the course of treatment, and the definitive diagnosis depends on the immunohistochemistry markers on the pathology examination. Favorable outcomes for supratentorially intra-parenchymal ependymomas can be achieved by total resection or assistant radiotherapy in incompletely excised patients.

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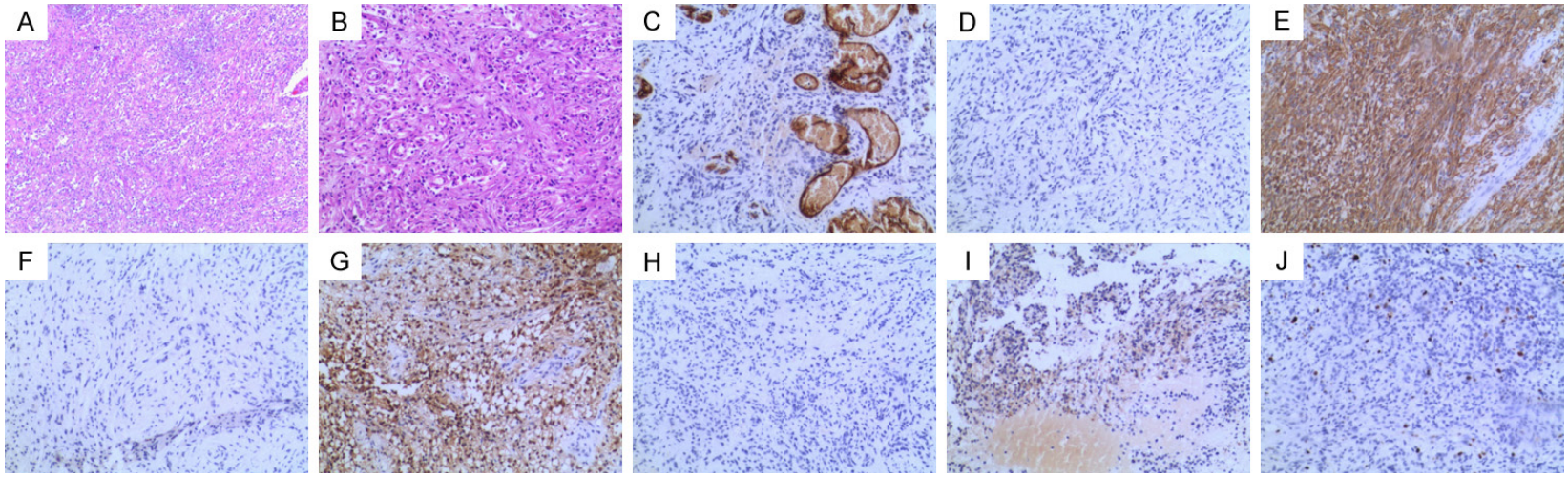

Figure 3. Tumor histological section staining. A. Staining with hematoxylin and eosin did not demonstrate the classic clustered cellular neoplastic proliferation with islands of high nuclear density with dense, as called perivascular pseudorosettes or true rosettes (original magnification 40); B. Staining with hematoxylin and eosin demonstrating in magnification 100; C. CD34 staining for tumor mature and normal vessels; D. Epithelial membrane antigen (EMA) staining negative indicated non-epithelium originated; E. Glial fibrillary acidic protein (GFAP) staining positive supported the origin of gliacyte; F. Neurofilament protein (NF) staining negative ruled out characteristics of neuroendocrine origin; G. S-100 protein positive supported the origin of neurocyte; H. Progesterone receptor (PR) negative indicated the non-hormonal dependent character; I. Synaptophysin (Syn) partially positive indicated neuroendocrine characteristics; J. Ki-67 focally positive indicated the inert characteristics.

Disclosure of conflict of interest

None.

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