Advanced Magnetic Resonance Imaging Findings of Anaplastic Papillary Glioneuronal Tumor

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ABSTRACT

We report a case of double brain tumors with anaplastic papillary glioneuronal tumor (PGNT) and atypical meningioma. PGNT typically has a good outcome and is regarded as a benign tumor. However, few cases of aggressive or malignant PGNT have been reported occasionally. To the author’s knowledge, this is the first report of advanced MRI features of PGNT. The MR perfusion, diffusion, susceptibility and spectroscopy findings may reflect the aggressive clinical course of this patient.

Keywords: brain, papillary glioneuronal tumor, MR, perfusion

INTRODUCTION

Papillary glioneuronal tumor was first described in 1998 and more than 90 cases have been reported [1], and it was designated as WHO grade I tumor in 2007 because of its benign clinical course. However, PGNT with poorer prognosis and aggressive nature have rarely been reported in the literature, and these tumors were considered by some authors to be a different disease entity [2-10]. Advanced MR techniques such as perfusion, diffusion, susceptibility, spectroscopy imaging may provide physiologic or biologic information, thus may help further characterize the lesions. In this paper, we describe a case of brain tumors with anaplastic PGNT and atypical meningioma. We aimed to report the advanced imaging features, which may reflect the aggressive nature of a PGNT.

CASE REPORT

A 37-year-old woman presented with right facial palsy in 2013. She also suffered from left temporo-parietal headache and slurred speech. Occasionally involuntary right facial twitching was also noted for 2 years. She denied recent trauma and past medical history. Physical exam found central facial palsy on right side and slurred speech, without other motor or sensory deficits.

Brain CT of the patient revealed two tumors, one in left frontal region and another one in left parietal lobe. Brain MRI was then performed. The left frontal region tumor measuring 5.6 × 3.5 cm was solid, extra-axial, with strongly heterogeneous enhancement, dural tail and adjacent hyperostosis, which is suggestive of a meningioma. The left parietal lobe tumor measuring 4.9 × 3.8 cm was solid, intra-axial and had perifocal edema and compression of the left lateral ventricle, resulting in midline shift to right side. It displayed heterogeneous signal intensity on T1WI (Fig. 1a), T2WI (Fig. 1b), T2-FLAIR (Fig. 1c) and post-contrast T1WI (Fig. 1d). Digital subtraction angiography showed a hypervascular frontal region tumor and a hypovascular parietal lobe tumor. Advanced MRI of the parietal lobe tumor demonstrated mixed signal intensity on
Advanced MRI findings of anaplastic papillary glioneuronal tumor

Figure 1

Figure 1. Conventional MR images of the anaplastic PGNT. T1WI a. T2WI b. and T2-FLAIR c. images show a heterogeneous tumor in left parietal lobe with perifocal edema and compression of the left lateral ventricle, resulting in midline shift to the right side. Post-gadolinium T1WI b. shows the heterogeneously enhancing tumor. Hyperostosis of the left-sided skull induced by the atypical meningioma was also noted.

DwI (Fig. 2a) and ADC (Fig. 2b), a central hypointensity on SWI (Fig. 2c), and focal high relative CBV (rCBV) on perfusion MR (Fig. 2d). MR spectroscopy (Fig. 2e) found high Cho/Cr ratio and low NAA, which are characteristics of a high-grade tumor. DTI tractography (Fig. 2f) showed the tumor lateral to the left corticospinal tract. Preoperative differential diagnosis of this tumor included high-grade glioma and brain metastases.

The patient underwent a left fronto-temporo-parietal craniotomy and total resection of the left frontal region tumor. One month later, a second operation with navigation-assisted craniotomy was arranged for the left parietal lobe tumor. A grayish tumor behind the somatosensory cortex was removed under guidance of Cavitron Ultrasonic Aspirator (CUSA).

Pathological study of the left frontal region tumor confirmed an atypical meningioma. The left parietal lobe tumor (Fig. 3) was characterized by papillary architecture, nuclear atypia, and increased mitotic activity. The results of immunohistochemical study of the parietal lobe tumor were as follows: GFAP (+), IDH-1 (+), Neu-N (+/-), synaptophysin (+), Ki-67 18%, representing a mixture of neuronal and glial components and a high proliferative index. The combined findings are diagnostic of an anaplastic papillary glioneuronal tumor.

After operation, the patient was treated with concurrent chemoradiotherapy (Temozolomide; 6000 cGy in 30 fx). Despite the aggressive management, follow-up MRI 13 months after surgery disclosed two tiny nodules in left parietal lobe and temporal lobe, suggestive of recurrent tumors.

DISCUSSION

Papillary glioneuronal tumor was first described in 1998 [11]. After that, more than 90 cases were reported in the literature [1]. Overall, the male to female ratio is about 1.3, and the mean age is 31.6-year-old [12]. All of these PGNTs were in supratentorial region, with predominantly periventricular location [1, 12]. The common symptoms of these patients are headache, seizure, nausea/ vomiting, visual disturbance, and even no symptoms [12].

Although PGNT was classified as benign tumor, some authors have noted that PGNT can be clinically aggressive or pathologically malignant. Ishizawa et al. reported the first case of PGNT with progressive clinical course in 2006 [2]. Vaquero and Coca suggested the existence of “atypical PGNT” and reported a case of PGNT with mitotic activity and a high proliferative index (Ki-67 15%) in 2007 [3]. “Anaplastic PGNT” or “aggressive PGNT” was then used to describe cases with aggressive clinical course or with high proliferative index [4, 5]. To date, about 10 cases of anaplastic PGNT have been reported in the literature [2-10].

We report the first case of double brain tumors of anaplastic PGNT and atypical meningioma. Both conventional MR images and advanced MR images of the aggressive PGNT are presented. The diagnosis of anaplastic PGNT is based on typical pathological findings of papillary architecture, mixture of neuronal and glial components, and a high proliferative index (Ki-67: 18%). According to the literature, radiological appearance of PGNT is variable, including 4 major categories: cystic mass with mural
Advanced MR images of the anaplastic PGNT. DWI a. and ADC b. display a mixed iso- and hypointense tumor on DWI compared to gray matter. SWI c. shows a central hypointensity. Perfusion MR d. shows focal areas of high rCBV. MR spectroscopy e. discloses high Cho/Cr ratio and low NAA, which are characteristics of a high-grade tumor. DTI tractography f. demonstrates the tumor lateral to the left corticospinal tract.

In our case, conventional MRI shows heterogeneous appearance, perifocal edema and mass effect can be seen in various degrees [13]. Hemorrhage may occur that can be misinterpreted as cavernous hemangioma [14].

In our case, conventional MRI shows heterogeneous appearance, perifocal edema and mass effect of the tumor. On DWI, the PGNT was mixed iso- and hypointense compared to gray matter, suggestive of no diffusion restriction of the tumor. Other DWI studies of PGNT displayed hypointense or mixed hypo- and isointense tumors, without difference between benign and anaplastic PGNT [5, 15]. On SWI sequence of our case, signal drop was seen in central part of the tumor, indicating hemorrhagic PGNT that have been reported in the literature [14].

On MR spectroscopy of our case, increase in Cho/Cr level was noted in the central part of PGNT. There are no data of MRS in PGNT, however, high Cho/Cr ratio determining the degree of malignancy and predicting outcome in intracranial astrocytoma [16, 17]. Thus the MRS finding in our case is typical of a high-grade tumor. On perfusion MR, our case revealed focal high rCBV, which means higher vascularity resulting from angiogenesis. Previous studies in glioma conclude that rCBV value is predictive of outcome [18]. This is to say, perfusion MR finding of PGNT in our case is also characteristic of an aggressive tumor.
Our patient received surgical removal of the two tumors, and followed by intensive management with combined chemotherapy and radiotherapy. However, early recurrence still occurred 13 months after surgery. So, the information provided by advanced MRI in our case may indicate aggressive clinical course, and double brain tumors may result in poorer prognosis additionally. Until now, high proliferative index is a potential indicator of poor prognosis in PGNT, but only 50% (9 of 18 cases) PGNT with Ki-67 more than 5% progress or recur eventually [1]. In order to find clear recommendation of follow-up and treatment, more cases and further study are required to confirm the relationship between advanced MRI findings and clinical outcome of PGNT.

CONCLUSIONS

In conclusion, the present case is the first reported double brain tumors of anaplastic PGNT and atypical meningioma. The advanced MR findings and the presentation may reflect the aggressive nature of the PGNT we reported. Further studies are required to establish the role of advanced MRI in predicting outcome of PGNT.

REFERENCES
