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Case Report

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J Korean Neurosurg Soc 42 : 224-227, 2007

Chordoid Glioma of the Third Ventricle with Unusual MRI Features

Chordoid glioma is a rare primary brain tumor of the third ventricle, and was recently characterized as a novel tumor entity. Typical radiological features of chordoid gliomas include; a solid, round-to-ovoid, well-circumscribed, contrast-enhancing mass of the hypothalamus and anterior third ventricle. Despite being classified as WHO grade 2, the third ventricular chordoid glioma has been reported to have a poor clinical outcome because of its anatomical location. The authors report a case of chordoid glioma of the third ventricle in a 48-year-old man with unusual radiological features of cystic component and ill defined circumscription.

KEY WORDS : Chordoid glioma · Third ventricle · Gamma knife · Cystic · Radiological feature.

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INTRODUCTION

Chordoid glioma is a rare low-grade tumor of the third ventricle, and was firstly reported as a distinct clinico-pathological entity in 1998 by Brat et al.[1]. Less than 35 cases of chordoid glioma have been reported in the English literatures[1-7,9]. According to their investigations, tumors tend to occur mainly in the third ventricle in middle-aged women[1]. The majority of chordoid gliomas arise in the hypothalamic, suprasellar, and third ventricular region, and the common radiological features of chordoid glioma have been described as solid, round-to-ovoid, well-circumscribed, contrast-enhancing masses of the hypothalamus and anterior third ventricle[5]. Here, the authors describe a case of chordoid glioma of the third ventricle, which presented as an isolated third ventriculomegaly with mainly cystic mass.

CASE REPORT

A 48-year-old man presented with progressive headache, memory impairment, and gait disturbance in December 2002. Magnetic Resonance Image (MRI) showed dilated third and lateral ventricles without any focal mass lesion (Fig. 1). The patient underwent a ventriculoperitoneal shunt under the impression of obstructive hydrocephalus in December 2002 at another hospital. He was referred to our institution for aggravated symptoms, presenting with intermittent diplopia and recent memory impairment in July 2004. MRI showed an enhancing mass in the midbrain, aqueduct of Sylvius, and multifocal enhancing lesions at the wall of the 3rd ventricle and the foramen of Monro (Fig. 2). The isolated ventriculomegaly of the third ventricle showed progression. Under the radiological diagnosis of high grade glioma of the tectum with ventricular seeding, an endoscopic third ventriculostomy and biopsy were performed in September 2004.

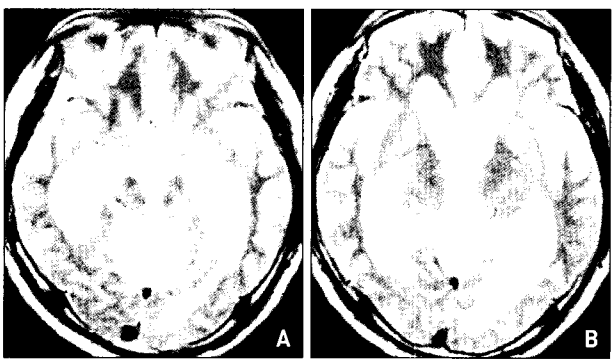

Fig. 1. T2 axial magnetic resonance (MR) images (A, B) show isolated ventriculomegaly of the third ventricle. No other MR images were available. At another hospital, a ventriculoperitoneal shunt was performed under the impression of obstructive hydrocephalus.

• Received : May 10, 2007
• Accepted : August 8, 2007
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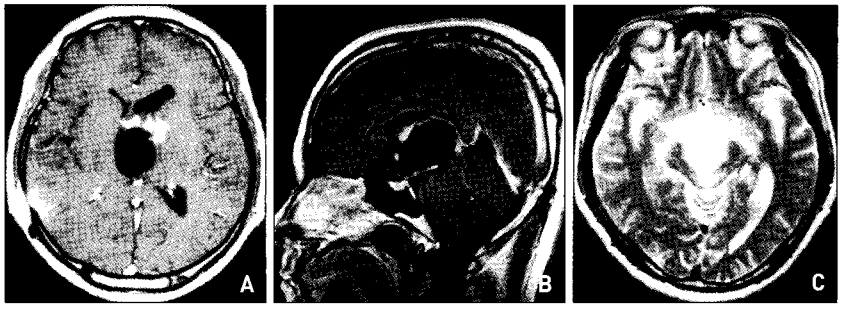

Fig. 2. T1 enhanced axial (A), sagittal (B), and T2 axial (C) magnetic resonance images revealing an enhancing mass in the midbrain and aqueduct of Sylvius, and around the foramen of Monro and left basal ganglia. Ventriculomegaly and multifocal enhancement at the wall of the third ventricle are also evident.

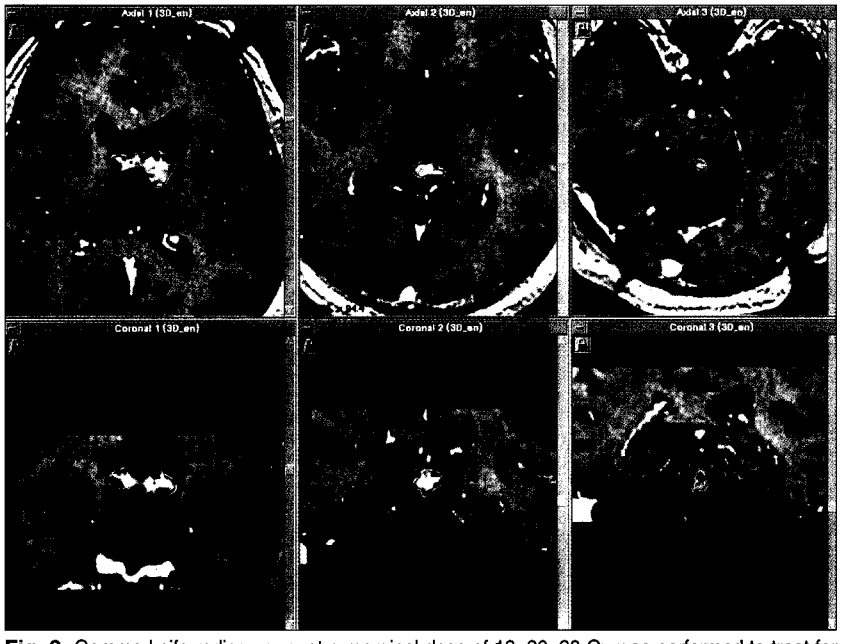

Fig. 3. Gamma knife radiosurgery at a marginal dose of 18, 20, 20 Gy was performed to treat for three remnant lesions, respectively.

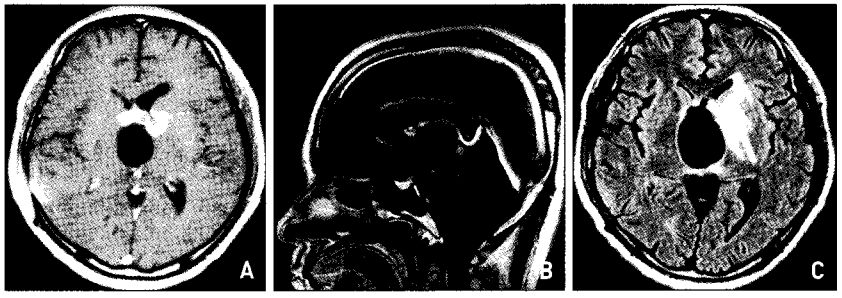

Fig. 4. Eight months after gamma knife radiosurgery, a regrowing enhancing tumor is visualized in the foramen of Monro extending to the left medial basal ganglia and posterior portion of the third ventricle near the pineal area and associated with a large cystic dilatation of the third ventricle on T1 enhanced axial (A), sagittal (B), and FLAIR axial (C) follow-up magnetic resonance images.

The lesion was located near the foramen of Monro and was yellowish and tough in consistency. The pathologic diagnosis was chordoid glioma of the third ventricle. Gamma knife radiosurgery at a marginal dose of 18-20 Gy was performed to treat remnant tumor (Fig. 3). Eight months after radiosurgery the patient suffered from hydrocephalus-like symptoms and follow-up MRI showed an enhanced tumor in the foramen of Monro extending to the left medial basal ganglia and around the right foramen of Monro. In addition, a large cystic dilatation of the third ventricle was observed, a possible feature of trapping (Fig. 4). In June 2005, the gross total removal of tumor was done through the transcallosal approach to the 3rd ventricle.

The tumor was adherent to the anterior and posterior aspect of the 3rd ventricle. The pathologic examination revealed that the tumor tissue was composed of ovoid to polygonal cells with eosinophilic cytoplasm. Neoplastic cells exhibited increased cellularity, cellular pleomorphism with a chordoma-like arrangement and focal lymphoplasmacytic infiltration. Neither anaplastic features nor necrotic foci were observed (Fig. 5). There were some similarities with pilocytic astrocytoma. However, the lymphoplasmocellular infiltration on the myxoid background of epithelioid cell could be differential point from pilocytic astrocytoma. An immunohistochemical study demonstrated diffuse reactivity for glial fibrillary acidic protein (GFAP) and S-100. No tumoral labeling was observed for epithelial membrane antigen (EMA). The Ki-67 index was 5% of neoplastic nuclei (Fig. 6). The patient had no additional neurologic deficits postoperatively and remained free of disease at his last imaging follow up in February 2007 (Fig. 7). No complication related to hypothalamic dysfunction was seen.

DISCUSSION

Chordoid glioma of the third ventricle is a rare tumor and its incidence has not been evaluated because it has only been recently described. It is a tumor of the middle aged and older adults though one case has been reported in a 12-year-old boy[2]. In general, the symptoms of this lesion include

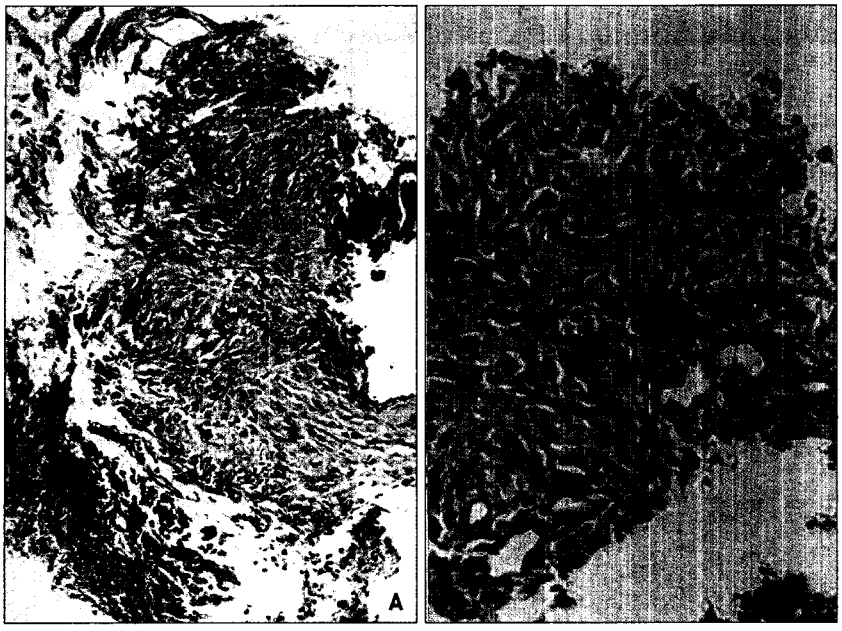

Fig. 5. Histological features (A, B) of the third ventricular mass, which show a cord-like arrangement of epithelioid tumor cells accompanied by lymphoplasmocellular infiltration (H & E, x400).

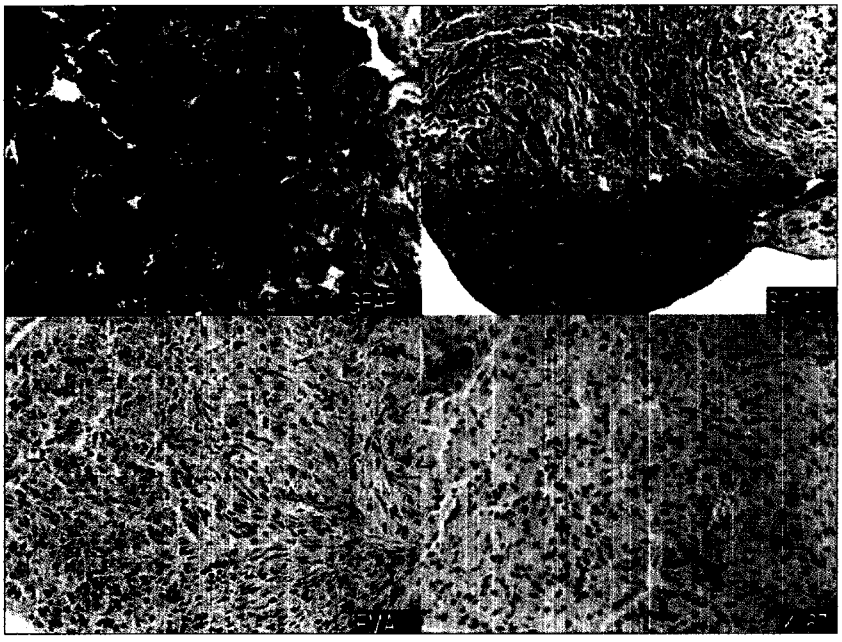

Fig. 6. Immunohistochemical study of the tumor showed that tumor cells are immunoreactive for glial fibrillary acidic protein and S-100, and focally positive for epithelial membrane antigen. The Ki-67 proliferating index is low at 5%.

headache, nausea, and vomiting due to hydrocephalus, visual field loss due to optic chiasm compression, endocrine abnormalities, and personality changes[1,2,4-7,9]. The immunophenotype of the chordoid glioma involves diffuse labeling for GFAP and CD34 with limited or no EMA reactivity, which distinguishes this neoplasm from metastatic carcinoma, chordoid or papillary variants of meningioma, epithelial chordoid plexus tumors, and intradural chordoma. Cases reported to date showed a remarkably uniform histologic appearance. Tumor tissues contain clusters and cords of epithelioid cells with eosinophilic cytoplasm and a prominent lymphoplasmocellular infiltrate. The stroma is mostly composed of basophilic, vacuolated mucin-like matrix, and as yet, no mitosis has been reported. Immunohistochemically, the tumor shows strong, diffuse reactivity for GFAP and vimentin[1,2,4,6,7,9]. Staining for EMA and cytokeratins have produced inconsistent results[1-3,6,9]. Reifenberger et al.[6] reported that the strong expression of CD34 with GFAP and of vimentin in chordoid gliomas represent a unique immunohistochemical pattern that facilitates the differential diagnosis of these neoplasms. The origins of chordoid glioma are not well understood. Pomper[5] speculated that the tumor arises from the hypothalamus because most lesions have shown tightly adherent to it. Cenacchi et al.[3] investigated the ultrastructural features of these neoplasms and were able to hypothesize a 'cell of origin' that might explain its regional specificity. Pathologic findings in our case were consistent with the previously reported cases.

Our case was unusual with respect to its radiological finding and response to treatment modalities. The lesion in our patient was a cystic and focally enhanced third ventricular mass and hydrocephalic symptoms recurred despite endoscopic third ventriculostomy. According to Reifenberger et al, no regrowth has been reported for up to 3.5 years after gamma knife radiosurgery[6,8] but gamma knife radiosurgery had little effect on our patient. No chemotherapy was administrated in our case. Gross total resection of the tumor alone has rendered our patient disease-free for over 2 years postoperatively. It is known that gross total resection of chordoid glioma is often difficult due to its location and adherence to surrounding structures. Moreover, some experiences of hypothalamic dysfunction[1,6,9] and thromboembolic events including pulmonary embolism[1,3,6] had

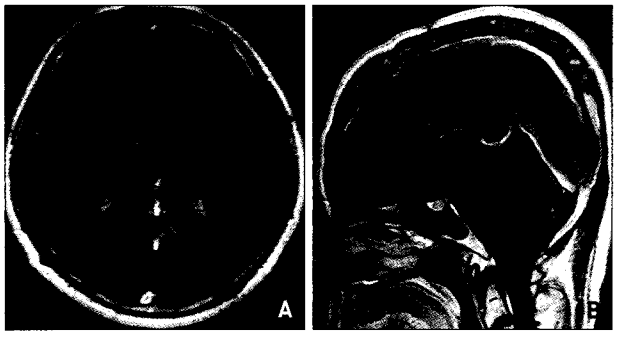

Fig. 7. No definite evidence of residual or recurrent tumors is observed on 2 year-postoperative T1 enhanced axial (A), and sagittal (B) magnetic resonance images.

been reported in association with the surgical treatment of chordoid glioma. Fortunately, the patient in this report was optimally managed by gross total resection without significant postoperative complication despite the recurrent hydrocephalic event, because the tumor was mainly cystic.

Although being classified as WHO grade 2, third ventricular chordoid glioma may have a poor clinical outcome because of its anatomical location. Limited follow-up studies suggest that as-full-as-possible resection favors a better outcome, which concurs with our experience. After more cases are reported, the long term outcome and management of these lesions will undoubtedly be further defined.

CONCLUSION

We present a rare case of chordoid glioma of the third ventricle with unusual cystic features. A diagnosis of chordoid glioma should be borne in mind for a cystic mass with multiple enhancing nodules and isolated ventriculomegaly of the third ventricle.

• Acknowledgement
This study was partially supported by grants from Seoul National University Hospital and the Korean Brain and Spinal Cord Research Foundation.

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