Chordoid Glioma in the Third Ventricle

- Case Report -

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Chordoid glioma of the central nervous system is a recently recognized tumor. It is necessary to be differentiated with the craniopharyngioma or other suprasellar masses due to its clinical and radiologic similarities.

The authors report a patient of 47-year-old man with a chordoid glioma in the third ventricle who underwent total removal of the tumor. Preoperatively, the patient showed hypersomnia, memory disturbance, bitemporal hemianopsia, drowsy mental status, and striking weight gain (25Kg) during the last 5-6 months. MRI showed huge homogeneous enhancing mass (5×5×6cm), locating in the sellae extending to the third ventricle and corpus callosum. Tumor was removed via interhemispheric transcallosal interforniceal approach. Histopathologic finding was chordoid astrocytoma characteristically showing GFAP (+), EMA (−), Collagen type Ⅳ (−), ki-67 (−), p53 (−) in immunohistochemical stain and epithelioid cells with eosinophilic cytoplasm, mucinous matrix, high cellularity, no mitosis in H & E stain. Postoperatively, a patient recovered normal mental status, sleep pattern, and vision. Postoperative fractionated radiation therapy was done.

KEY WORDS : Chordoid glioma · Third ventricle · Hypothalamus · Suprasellar region.

Introduction

Chordoid glioma is a rare intracranial tumor. Since Brat, et al have firstly mentioned on a chordoid glioma in 1998, about 20 cases of chordoid glioma have been reported until 2000[1,2]. This tumor is likely to be arised in the hypothalamus, the third ventricle, and the suprasellar area. In computed tomography and magnetic resonance (MR) image, a chordoid glioma is shown by oval, well demarcating tumor with strong homogeneous contrast enhancement. It usually combines the symmetric vasogenic edema surrounding the tumor by mass effect. Histopathological findings are characterized by reminiscent of chordoma or chordoid meningioma, and its avid staining with glial fibrillary acidic protein. Clinically, the symptoms are similar to those of a craniopharyngioma and other suprasellar tumors. Since the chordoid glioma locates in the region of the anterior third ventricle and hypothalamus, total removal is difficult. And the tumor has a tendency of regrowing from the remaining tumor after incomplete removal. The radiotherapy is not effective so that the prognosis is unfavorable. Therefore, it has been insisted that a chordoid glioma should be aware as another distinctive intracranial tumor and also be diagnosed differentially from the common tumors arising from the third ventricle and the hypothalamus.

We report a case of chordoid glioma locating in the third ventricle showing distinctive clinical, radiological, and histopathological features.

Case

A 47-year-old man presented hypersomnia, memory disturbance and striking weight gain during 5-6 months before admission. On neurologic examination drowsy mental status and bitemporal hemianopsia were noted. MR image (Siemens 1.5 tesler Magnetom Vision, T1WI, T2WI, Gd-enhancing T1WI) showed well circumscribed mass located in the suprasellar area, third ventricle and hypothalamus. The tumor size was measured by 5×5×6cm. In T1WI, the mass was well demarcated and heterogeneous mixed with iso-and hypointense

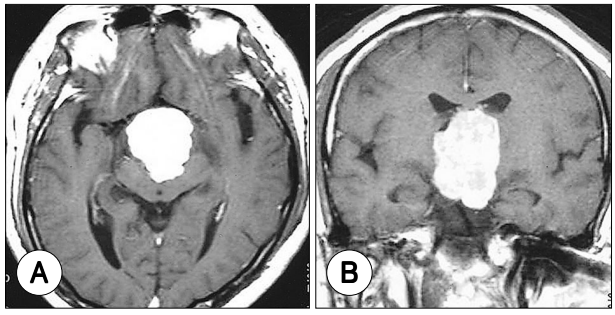

Fig. 1. Preoperative enhanced magnetic resonance axial(A) and coronal(B) images showing huge mass with homogeneous enhancement locating in the sellar and suprasellar area extending to the hypothalamus and corpus callosum.

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areas. In T2WI, it was also well circumscribed and consisted of mixed intensity with hyperintense and isointense lesion. In gadolinium enhancement image, the mass represented homogeneous enhancement with definite boundary(Fig. 1). The tumor displaced the hypothalamus anteriorly and corpus callosum superiorly.

The tumor was removed via interhemispheric transcallosal interforniceal approach. With resection of the corpus callosum, the whitish surface and hypervascular tumor was discovered. It was friable and sucked easily by Cavitating UltraSonic Aspirator(CUSA). Hypothalamus was displaced anteriorly to its normal position. Grossly total resection was performed. During surgery special precaution was paid to avoid the injury of the pituitary gland and stalk.

After the operation, the mental status recovered normal. However, the patient suffered from severe respiratory distress and intermittent apneic condition. Plain chest X-ray showed the findings of pulmonary edema. Tracheostomy and ventilatory support were done for the intensive care of respiratory distress which was normailized after 10 days. Transient left third nerve palsy and central diabetes incipidus were also normalized within three weeks. Hormonal dysfunction was not presented except decreased level of serum cortisol. Visual field defect was improved within 3 to 4 weeks.

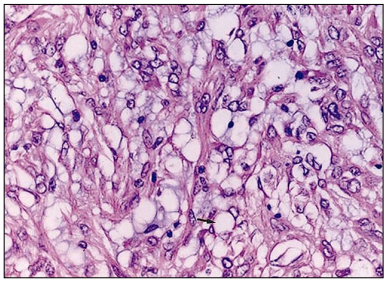

Fig. 2. The tumor cells showing multivesicular eosinophilic cytoplasm filled with mucinous material resembling chordoma. A black arrow indicates Russell body(H & E, ×120).

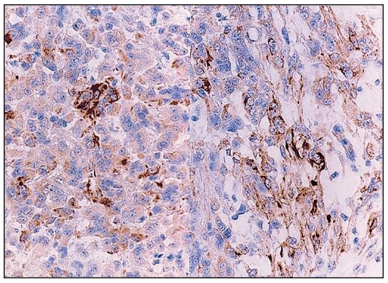

Fig. 3. Immunohistochemical stain for glial fibrillary acidic protein(GFAP)revealing strong positive in the chordoid tumor cells(×120).

Pathologically, it showed clusters and cord of oval-to-polygonal epithelioid, spindle, fibrillar cells with abundant eosinophilic cytoplasm in H-E stain(Fig. 2). Occasionally, vacuolated physaliferous cell was found. Their stroma consisted of an abundant, slightly basophilic and vacuolated extracellular mucin. Tumor showed high cellular density. However, it had no mitosis, necrosis and endothelial proliferations. Lymphoplasma cell invasion and Russell body were also noticed (Fig. 2). Immunohistochemistry of this tumor showed GFAP(+), EMA(−), Collagen type Ⅳ(−), ki67(−), and p53(−)(Fig. 3). All these distinctive radiolo-gical, clinicopathological, and immunohistochemical findings were similar to those of the reported cases in world literatures. So it was diagnosed as a chordoid glioma. Postoperative brain MRI revealed near total resection of the tumor in the third ventricle except a small portion of slight enhancement adjacent to the hypothalamus(Fig. 4). The fractionated radiation therapy(total 5980cGy) was done against to the remaining enhancing lesion. MRI in six months after surgery, tumor recurrence was not noticed(Fig. 5).

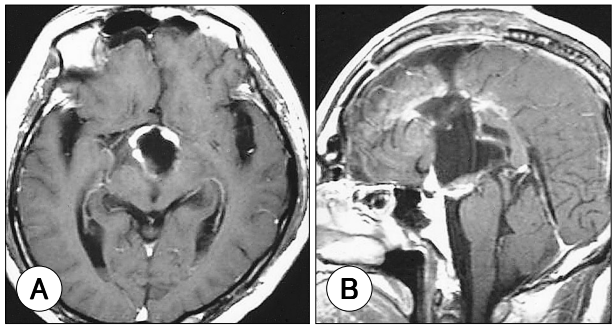

Fig. 4. Postoperative enhanced magnetic resonance axial(A)and sagittal(B) images after removal of the tumor showing small enhancing lesion adjacent to the hypothalamus.

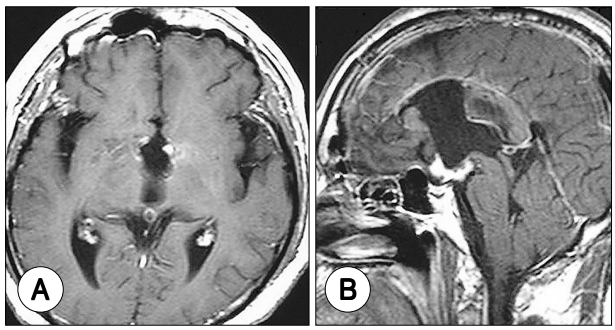

Fig. 5. Post-irradiation enhanced magnetic resonance axial(A) and sagittal(B) images in 6 months after removal of the tumor.

Discussion

Chordoid glioma is a very rare and recently recognized primary intracranial tumor. Even though this tumor has predominance in the female between the ages of 40th and 70th as 12 cases out of 18 reported cases were female in the literature, author's case was a 47 year old male. Clinical symptoms are variable including obstructive hydrocephalus, memory disturbance, hypersomnia, visual field defect, ataxia, headache, psychologic disorder, and thyroid dysfunction due to the tumors arising in the third ventricle, hypothalamus, and suprasellar

tumors. In our case, hypersomnia, memory disturbance, visual field defect, and striking weight gain were observed. The radiologic features include a large mass which showed in the suprasellar area, homogeneous contrast enhancement, but no calcifications and rare cystic component[1,8]. Therefore, craniopharyngioma, pituitary macroadenoma, metastatic tumor, meningioma, pilocytic astrocytoma of the optic nerve and hypothalamus, and aneurysm must be included in the differential diagnosis[7,11]. In a standpoints of differential diagnosis, craniopharyngioma presents calcification and cystic formation, and distinctively solid or nodular enhancement. Pituitary macroadenoma enlarge the sella, giving the "J shape configuration" and erode the bone. Pituitary macroadenoma frequently extends to the suprasellar region displaying the "snowman appearance".In metastasis, symptoms are noticed even in smaller tumor. And early cavernous sinus involvement by a small pituitary mass and/or hypothalamic/infundibular involvement are demonstrated. An edematous response in adjacent portions of the brain is also seen. In our case, the tumor was very large extending to the suparasellar region, third ventricle and hypothalamus. Our case also demonstrated well demarcation from the surrounding tissues and homogeneous enhancement. Rarely Rathke's cleft cyst, hamartoma of the tuber cinereum and granulomatous diseases such as sarcoidosis, tuberculosis and eosinophilic granuloma also must be considered in the differential diagnosis[7].

Histopathologically, several unique features are observed. The tumors consists of clusters and cords of oval to polygonal epithelioid cells embedded in a mucinous stroma. The epithelioid cells have abundant eosinophilic cytoplasm, uniform nuclei with a delicate chromatin pattern, and inconspicuous nucleoli. A prominent lymphoplasmacytic infiltrate with numerous Russell bodies is usually seen. Low mitotic activity, no microvascular proliferation and necrosis are noticed. Reactive astrocytes, a number of Rosenthal fibers and chronic inflammatory cells exist due to mass effect by the tumor, but no infiltration into the brain parenchyme are observed. Immunohistochemical studies show strong positive to GFAP and vimentin and negative or weakly positive to epithelial membrane antigen(EMA), cytokeratin, S-100 protein, and ki-67(MIB-1)[8]. Expression of CD-34 and epidermal growth factor have been also noticed[9]. Ultrastructural findings of this tumor reveal round to spindle cells with focal projections resembling microvilli, abundant cytoplasmic intermediate filaments, scattered intermediate junctions, and focal basal lamina formation[1]. Even though more supportive features of ependymal derivation of this tumor, such as cilia and desmosomal junction, have not been reported, the presence of focal basal lamina formation and microvilli presume the possibile origin of this tumor from the ependymal cells[1]. Histopathological differential diagnosis includes chordoma, chordoid meningioma, atypical teratoid tumor, and rhabdoid tumor. On H & E stain, chordoid glioma generally lack of vacuolated physaliferous cells, mucinous cytoplasm and show no tendency for bone infiltration, which are typical features of chordomas[2,8]. In immunohistochemistry, chordoma shows NF(−), GFAP(−), 30% positive to vimentin, 80% positive to S-100 protein, and more positive to cytokeratin, EMA than chordoid glioma[3,6]. H & E stain for chordoid meningioma presents cellular whorl, psammoma body, and nuclear inclusion. Immunohistochemical studies for chordoid meningioma reveals NF(−), GFAP(−), S-100(−), CK(−), EMA(+), and vimentin(+). Chordoid lymphoplasmacyte-rich meningioma has the distinctive features of reactive lymphoid follicle with germinal center, and occur in adolescents(ages 8-19 years) associatd with systemic Castleman's syndrome[1,8]. In atypical teratoid tumor and rhabdoid tumor, immunohistochemical studies depict 38% positive of NF, 73% positive of GFAP, 40% positive of S-100, 100% positive of cytokeratin, EMA, and vimentin[10]. In our case, vacuolated physaliferous cells with mucinous cytoplasm was rarely seen in H & E stain, and GFAP(+), EMA(−), Collagen type Ⅳ(−), ki67(−), p53(−) were noticed in immunohistochemical stain, which are distinctive features of the chordoid glioma.

Regarding to the histogenesis of the chordoid glioma, Guido Reifenberger et al described that this tumor is not related to any intracranial tumors in the immunological and molecular biological aspects[8]. Another study presented the probability of the derivation of chordoid glioma from the subependymal tissues or the hypothalamic cells[7,9]. Others studied the possibility of the influence of the estrogen and progesterone receptors on the histogenesis of chordoid glioma in relation to the middle aged female preponderance but they did not confirm any relationship with the hormonal effects[11,12]. Still the histogenesis of this tumor has been remained unresolved. Author's case, however, seemed to arise from the hypothalamic cells due to broad adhesion to the hypothalamus.

On the prognostic analysis of the 18 reported cases in literature, seven patients(39%) were died. The causes of death were the tumor recurrence in one, medical complications in five, and undescribed reason in one[4]. The main medical complication was the respiratory failure. After surgery, the tumor was recurred in 12 patients due to incomplete tumor removal. However, the tumor did not show any regrowth even after incomplete removal in five patients. In our case, the patient had suffered from respiratory distress and pulmonary edema immediate after the surgery. Intensive care returned the patient to normal condition. And during the operation,

attention was given to preserve the hypothalamus and pituitary gland. Fractionated radiation therapy was done to residual tumor which adhered to hypothalamus.

Conclusion

Chordoid glioma arising in the third ventricle, suprasellar area, and hypothalamus is one of a rare intracranial tumor. Due to radiological similarities, differential diagnosis must include craniopharyngioma, pituitary adenoma, and metastatic tumor, meningioma, and pilocytic astrocytoma. It, however, represents a novel entity with distinctive clinicopathologic and immunohistochemical features. Even though it is pathologically low grade tumor, total extirpation is not easy due to difficult surgical approach. The prognosis of this tumor is unfavorable. For the sake of prognostic stratification and treatment, it is important to correctly identify this tumor, and additional cases and longer follow-up periods must be allowed for evaluation.

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