Case Report

Chordoid Glioma of The Third Ventricle: A Case Report and Review of Literature

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Abstract
Chordoid glioma of the third ventricle is a newly described tumor. In the present study, we report a case of chordoid glioma and review various, previously published case series and individual case reports of chordoid glioma. With regard to the case report, the patient was a 53-year-old woman whose primary symptoms included decreased vision, a visual field deficit on the nasal side, and headache. CT and MRI demonstrated a tumor that was located in the supra-sellar region of the sella turcica. Microscopically, the tumor was composed of epithelioid cells in a mucinous background with abundant eosinophilic cytoplasm arranged in nests and strands. A mixed, chronic inflammatory infiltrate composed of lymphocytes and plasma cells was abundant in the peripheral portion of the tumor. Immunohistochemically, the tumor cells expressed GFAP and vimentin, whereas EMA was found to only label individual cells. Less than 1% of the cells were Ki-67 positive. The results demonstrate that chordoid gliomas of the third ventricle can be differentiated from other tumors based on clinical and histopathological features as well as immunohistochemical staining.

Keywords: Chordoid glioma, third ventricle, immunohistochemistry

Üçüncü Ventrikülün Kordoid Gliomu: Olgu Sunumu ve Literatür Derlemesi


Anahtar Kelimeler: Kordoid gliom, üçüncü ventrikül, immünohistokimya
INTRODUCTION

Chordoid glioma is a rare, low-grade brain neoplasm occurring in the third ventricular region, which predominantly occurs in middle-aged women. In 1995, Wanschitz described a 24-year-old woman with an isolated tumor in the third ventricle, and the tumor had the histological and immunohistochemical features of a notochord-like glioma. The investigators characterized the tumor as a type of meningioma that expressed GFAP. However, the immunohistochemical and ultrastructural studies demonstrated that the tumor cells originated from gliocytes rather than meningothelial cells. Brat first named this type of tumor “chordoid glioma” in 1998, and in 2000, the World Health Organization (WHO) formally designated it as chordoid glioma of the third ventricle. Since its first report, approximately 50 cases have been described in the English literature. In the present study, we report an additional case and summarize the clinical, morphological, and immunohistochemical features of the tumor. In addition, we discuss the differential diagnosis of chordoid glioma by reviewing the previous literature on this tumor type.

CASE PRESENTATION

Patient history

The patient was a previously healthy 53-year-old woman who complained of decreased vision, a visual field deficit on the nasal side and headache for one year. Magnetic resonance imaging (MRI) revealed an ovoid mass measuring 3.5×3.2×3 cm (Figure 1) in the supra-sellar region of the sella turcica. The mass appeared isointense with uniform contrast enhancement on the T1-weighted MR images, and the space-occupying effect was so significant that the optic chiasm was compressed and shifted. The ventricular system was also slightly compressed, resulting in hydrocephaly. On examination, the patient presented with a visual field defect on both nasal sides and hypopsia, but no other positive neurologic signs. Laboratory studies detected a normal level of serum hormones. The primary clinical diagnosis was craniopharyngioma. Intraoperatively, the tumor was located in the third ventricle with a clear boundary, and its total size was approximately 3.5×3.5×3 cm. The tumor tissue appeared gray-red in color and was soft with abundant vessels. Fortunately, the tumor was completely resected (Figure 2). Postoperatively, the patient was treated symptomatically without radio- or chemotherapy. After 18 months of follow-up, the patient was stable with no clinical events or signs of recurrence.

Pathology

Microscopically, the epithelioid cells were arranged in nests and strands with abundant eosinophilic cytoplasm in a mucinous background (Figure 3A). Tumor cells were generally orbicular-ovate or polygonal in shape, and the cytoplasm was eosinophilic with an obscure boundary. The cellular nuclei appeared uniformly round with stable chromatin; generally, nucleoli were visible, but evidence of karyokinesis was rare. No obvious necrosis or microvascular proliferation was observed in the tumor tissue, which was divided into lobules by filaments. In addition, there was abundant infiltration of lymphocytes and plasmocytes in the interstitial substance (Figure 3B).

Immunohistochemistry

The expression of GFAP and vimentin in the tumor cells was diffuse and strong (Figure 4); however, the expression of EMA was focal or scattered (Figure 5A). The CK, CD34, S-100, Syn and CgA markers were all nonreactive. The expression rate of Ki-67 was less than 1% (Figure 5B).
Figure 1: Well-demarcated, homogeneous enhancement in the solid area (arrows) (sagittal [A] and coronal [B] contrast-enhanced magnetic resonance images).

Figure 2: Postoperatively, MRI demonstrated that the tumor had been completely resected (sagittal [A] and coronal [B] contrast-enhanced magnetic resonance images).

Figure 3A: Epithelioid cells in nests in a mucinous ground substance (hematoxylin and eosin, original magnification ×40). B, Lymphocytic and plasmocytic infiltration (hematoxylin and eosin, original magnification ×40).
DISCUSSION

Clinically, chordoid glioma is a very rare tumor of the central nervous system. To date in the English literature, approximately 50 cases have been reported in individuals ranging from 12-71 years of age (mean 46 years). The tumor generally occurs in middle-aged women, and the male to female morbidity rate has been described as 1:1.9-2. Chordoid glioma is often located in the front of the third ventricle, which leads to the shifting of structures and symptoms of obstructive hydrocephalus, e.g., headache, nausea, and loss of coordination. At times, the tumor can dislocate the hypothalamus and optic chiasm inferiorly, leading to endocrine and visual disturbances. Moreover, the tumor may depress the medial temporal lobe resulting in psychological and memory disturbances. Based on imaging studies, a chordoid glioma appears ovoid and well circumscribed with a diameter of 2-4 cm and is located in the third ventricle immediately adjacent to the hypothalamus. The tumor shows intense and uniform enhancement with computerized tomography and magnetic resonance imaging.
imaging. Chordoid glioma has low-grade malignant behavior, seldom invading surrounding tissues. As a result, chordoid glioma is classified as a grade II based on WHO criteria.\(^6\)

**Histological characteristics**

In previously described cases of chordoid glioma, the tumor cells frequently demonstrated a pattern of differentiation consistent with gliomas, with rough fibriform processes.\(^1\) Other findings including reactive astrocytes, Rosenthal fibers, and patterns of chronic inflammatory cell invasion have been described in surrounding non-tumor tissues with Russell bodies. The other features have been reported in this case.

**Immunohistochemistry**

The majority of neoplastic cells revealed a strong and diffuse pattern of expression of vimentin and glial fibrillary acidic protein (GFAP), which, according to previous literature, is consistent with their glial cell origin. However, the patterns of expression with S-100 and EMA were not fixed and were focally positive, respectively. All tumors demonstrated immunoreactivity for the epidermal growth factor receptor and schwannomin/merlin, but not for TP53, Waf-1 or MDM2.\(^7\) The proliferation potential of chordoid glioma was similar to low-grade gliomas with zero or rare mitoses. The average labeling index of MIB-1 was very low and was reported to be 0-1.5\(^{1(1)}\) or < 5\(^{7}\)%

**Ultrastructure**

Tumor cells are round or spindle-shaped with irregular, polymorphic nuclei. Villi are scattered and short with no cilia or desmosomes. Frequently, scattered outer plates and a few semi-desmosome-like structures connecting the cellular membranes to the underlying basal lamina were seen. In general, intermediate junctions are frequently present. Expanded rough endoplasmic reticula were located around the nucleoli. In the middle zone of the cytoplasm, the majority of cellular organs were Golgi bodies, and at the outer zone, there was an abundance of mitochondria and secretory granules. The outermost layer of cytoplasm contained intermediate filaments arranged in parallel that appeared as concentric circle-like structures surrounding the nuclei.\(^2\)

**Histogenesis and molecular biology**

The subcommissural organ (SCO), which is located at the entrance of the aquaeductus mesencephali, is a glandular, organ-like structure composed of ependymocytes and subependymal cells. Cenacchi et al.\(^5\) first described the similarities in ultrastructure and molecular expression between chordoid gliomas and the SCO, as described above. They therefore suggested that chordoid gliomas are likely a subtype of ependymoma because the cells resemble the highly specialized ependyma of the subcommissural organ.\(^4\)

**Differential diagnosis**

Chordoid gliomas should first be distinguished from chordoid meningioma because both demonstrate eosinophilic, epithelioid tumor cells distributed in a myxoid matrix. Furthermore, both are infiltrated with lymphocytes and plasma cells. However, chordoid meningiomas are generally located around the supratentorial meninges. Histologically, chordoid meningiomas contain typical meningioma-like microscopic structures and usually demonstrate positive staining with EMA and negative staining with GFAP. Second, chordoid gliomas should be distinguished from the mucin-type papillary ependymoma because both express CK and vimentin. However, the mucin-type papillary ependymoma is most often situated at the cauda equina of the spinal cord rather than intracranially. Of note, while the histological features of the mucin-type papillary ependymoma include papillaries that are formed by macrofiber cells, which have fibers that use hyalinization to stretch to vessels, these
cells can also be found in chordoid gliomas.

**Therapy and prognosis**

Chordoid gliomas are located along the third ventricle, the hypothalamus, and in various structures superior to the saddle, which makes complete resection of these tumors very difficult. Approximately half of patients will suffer or die from tumor recurrence after subtotal resection. One study(5) evaluated 35 patients with a follow-up period between 6 and 68 months (means 22.5 months). Total resection was possible in 10 cases. Among these patients, 6/10 had no recurrence, 3/10 died from postoperative complications, and 1/10 was lost to follow-up. Among the 25 patients who underwent sub-total resection, 6/25 had no recurrence, 4/25 had tumor recurrence, 5/25 died and 10/25 were lost to follow-up.

Although chordoid glioma is regarded as a low-grade malignant tumor, its biological behavior is not encouraging. Due to the limited number of documented cases, the utility of adjuvant therapy after total resection is still unknown. In addition, therapeutic methods and results require further study.

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