

Chordoid Glioma : an Uncommon Tumor of the Third Ventricle

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Chordoid glioma is an uncommon low-grade tumor of the third ventricle with histologic features of a chordoma and immunolabeling for glial fibrillary acid protein. We present a rare case of a patient with a chordoid glioma of the third ventricle and review the literature regarding this tumor's clinical, radiological and pathologic aspects.

KEY WORDS : Chordoid glioma · Third ventricle · Low-grade tumor.

Introduction

Chordoid glioma is a recently recognized rare brain tumor located exclusively in the third ventricle¹⁾. This unusual tumor is currently considered a glial neoplasm (WHO grade II) of uncertain histogenesis with distinct clinicopathological features¹⁻¹⁰⁾. It should be differentiated from other suprasellar and anterior third ventricular tumors due to unique disease entity.

We report a rare case of a 25-year-old woman with a chordoid glioma of the third ventricle and present a review of the literature in an attempt to clarify the clinical entity.

Case Report

A 25-year-old woman presented with amenorrhea for 2 years. Recently, she had developed decreased visual acuity and bitemporal hemianopsia for 3 months before admission. Other neurological examination was unremarkable, but

endocrinological studies showed hypoadrenalism and hypogonadism.

Preoperative brain magnetic resonance imaging (MRI) revealed a 3.5 × 3.5 × 3.5 cm sized round mass with iso-signal on both T1 and T2 weighted images within the third ventricle.

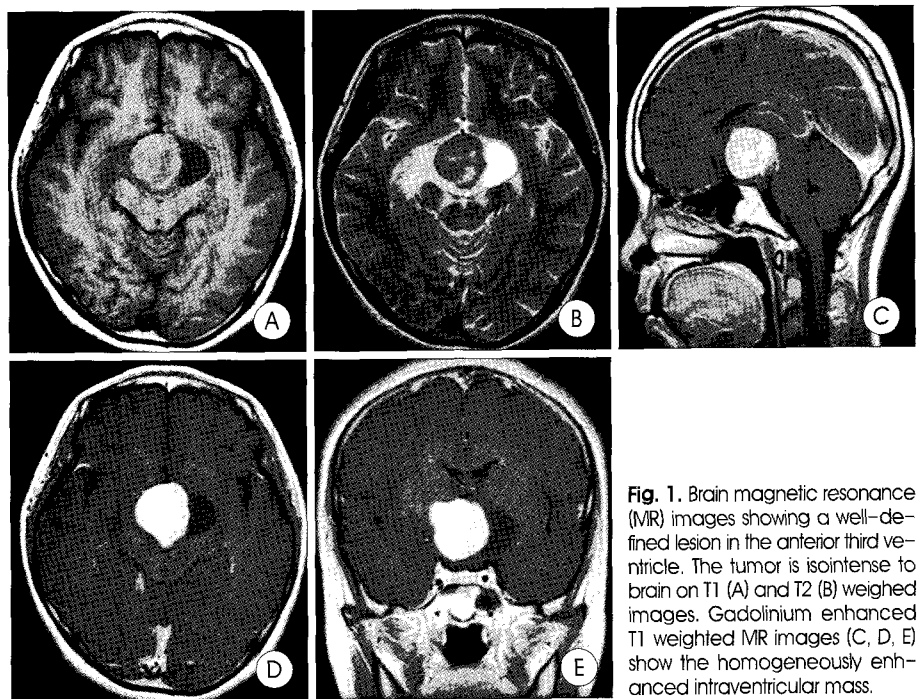


Fig. 1. Brain magnetic resonance (MR) images showing a well-defined lesion in the anterior third ventricle. The tumor is isointense to brain on T1 (A) and T2 (B) weighed images. Gadolinium enhanced T1 weighted MR images (C, D, E) show the homogeneously enhanced intraventricular mass.

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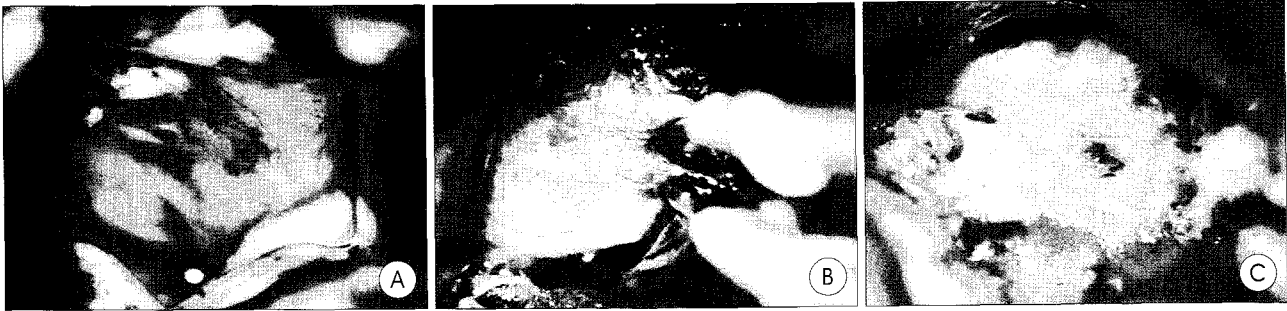


Fig. 2. Intraoperative photographs demonstrating the posterior free surface of the tumor in the third ventricle before resection (A), lateral surface faced to ventricular wall (B), and tumor origin site on the ventricular floor after subtotal resection of mass (C).

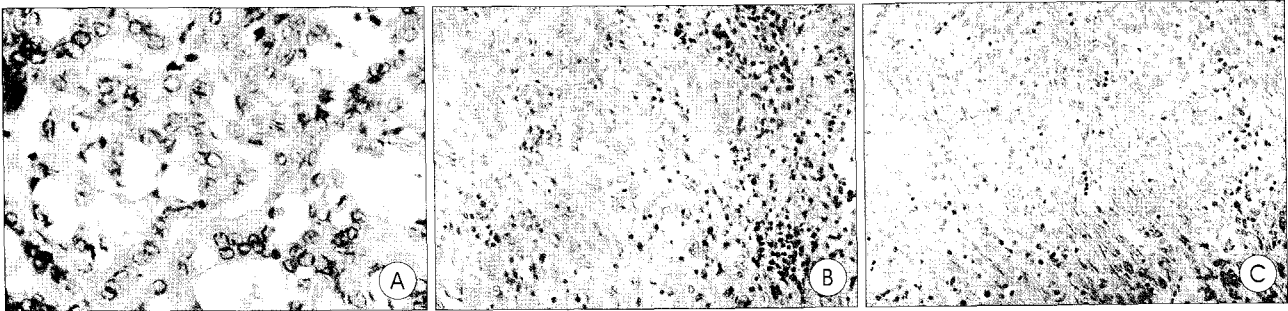


Fig. 3. Photomicrographs of the tumor specimen. A : Neoplasm is composed of epithelioid cells which revealed round to oval nuclei with abundant cytoplasm (H & E, original magnification $\times 400$). B : Tumor cells show a sheet or cord like arrangement. A heavy infiltration of lymphoplasmic cells and lymphofollicles is seen in the peripheral portion ($\times 200$). C : Immunohistochemical study demonstrating diffuse cytoplasmic reactivity for glial fibrillary acid protein (GFAP) (GFAP $\times 200$).

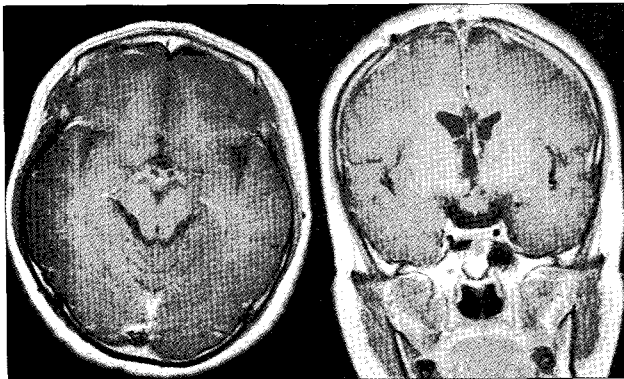


Fig. 4. Postoperative enhanced magnetic resonance imaging demonstrating no evidence of residual mass after 13-month follow-up.

ricle (Fig. 1). The mass showed well-circumscribed, homogeneous enhancement after gadolinium administration. Additionally, a cystic component and mild peri-lesional edema also were noted.

The mass was removed gross totally via anterior transcallosal transchoroidal approach. The tumor had a lobulated surface and a firm consistency like a meningioma (Fig. 2). It was smooth and yellowish in color (similar to the ependymal surface). The dissection plane between the mass and ventricular wall was easily defined. We found severe adhesion at the ventricle's floor, however, which was considered as an origin site. The tumor's adhesion required meticulous microdissection in order to obtain a gross total resection.

Pathological examination found the neoplasm to be composed of epithelioid cells which revealed round to oval nuclei with abundant cytoplasm. The tumor's cells showed a sheet or cord like arrangement and a heavy infiltration of lymphoplasmic cells and lymphofollicles was seen in the peripheral portion. Part of the tumor's cells showed a tubular or alveolar structure. Dense fibrosis with hyalinization was also noted. On immunohistochemical studies, tumor cells showed glial fibrillary acid protein (GFAP) (+), CD34 (+), vimentin (+), epithelial membrane antigen (EMA) (weak +), S-100 (-), CD68 (+) and P53 (-). Mitotic figures were absent. They demonstrated the feature of the chordoid glioma (Fig. 3).

The patient was recovered fully without significant neurological deficits except transient diabetes insipidus (DI). Her visual field defect had recovered within 3 months after the operation as well. However, endocrine abnormalities were not recovered and adrenal hormone replacement continued. A postoperative enhanced MRI demonstrated no evidence of residual mass after her 13-month follow-up (Fig. 4).

Discussion

Chordoid glioma is a rare low-grade tumor in the third ventricle first characterized as a distinct clinicopathologic entity in a 1998 report by Brat et al.¹⁾ It is a unique lesion distinguished by its topography, stereotypic histology and

female predilection. To our knowledge, about only 32 cases of chordoid glioma have been reported¹⁻¹⁰. Its clinical features are related to the compression of the hypothalamus (amenorrhea, DI), to optic chiasm (homonymous hemianopsia) in the vicinity of the third ventricle, or to the obstruction of CSF circulation (hydrocephalus). The scarcity of reported cases and their short follow-up periods, as well as their location within the third ventricle with frequent attachment to hypothalamic and suprasellar structures, make it difficult to reliably predict prognosis.

The radiologic findings of the chordoid glioma are similar to those previously reported. On MRI, chordoid gliomas are usually ovoid in shape and are located in the third ventricular-suprasellar region. They are well demarcated, usually solid (some lesions have a cystic component), and almost always homogeneously enhancing. The radiologic differential diagnosis includes: craniopharyngioma, ependymoma, optic and hypothalamic pilocytic astrocytoma, choroid plexus papilloma, and meningioma^{2,4,8}.

The nomenclature of a chordoid glioma reflects the facts that this neoplasm has chordoma-like histologic features while immunolabeling for GFAP. Histological findings in the reported cases consisted of a neoplasm with scarce or no infiltrative component, with abundant lymphoplasmacytic infiltrates with Russell bodies and epithelioid cells arranged in cords with a mucinous matrix, without nuclear pleomorphism or mitotic figures. Cells were intensely and diffusely immunoreactive with GFAP in every case, and slightly and focally reactive with EMA in half of the cases. This new tumor did not conform to any existing gliomal histopathologic classification system, leading Brat et al.¹ to propose that chordoid glioma was a separate pathologic entity. While the glial nature of chordoid gliomas seems settled, their histogenesis is unclear. Comparative genomic hybridization analysis demonstrated that chordoid gliomas lack the chromosomal and genetic abnormalities commonly found in diffuse astrocytomas⁷. Some ultrastructural studies suggest that chordoid gliomas may be of ependymocytic lineage or at least capable of differentiating along ependymal lines^{3,5}. Another possibility is that the tumor arises from the hypothalamus⁶. That possibility is supported in that most lesions adhere tightly to the hypothalamus during surgery, such as in our case, resulting in a difficult resection, although the tumor is well circumscribed. This neoplasm's histopathologic differential diagnosis has been performed in comparison to many different tumors: with ependymoma and pilocytic astrocytoma due to its low cytological malignancy and low infiltrative capacity together with its solid appearance, and with chordoid meningioma and chordoma due to the presence of epithelioid cells¹. Paraganglioma and pituitary adenoma were other proposed diagnoses¹⁰.

Gross total resection alone has rendered patients disease-free for reported postoperative intervals of over five years and appears to be the treatment of choice, even if it may result in an incomplete resection (50% of reported cases)². No recurrences have been reported up to date in cases of macroscopically complete tumor removal. Surgical exploration of these lesions, however, may be difficult and hazardous due to their tendency to adhere to critical neighboring structures. Because of their location and the difficulty of obtaining complete surgical resection without the patient suffering severe hypothalamic symptoms, prognoses in spite of the limited number of cases have been usually poor. Chemotherapy and radiation therapy have been unsuccessful in the few cases in which these modes of therapy have been applied. Brat et al.¹ reported that two partially removed tumors showed regrowth despite radiation therapy. However, stereotactic radiosurgery seems to be promising. No regrowth has been reported up to 3.5 years after gamma-knife radiosurgery^{7,9}. Nevertheless, the benefit of radiosurgery in the treatment of the partially excised chordoid glioma is uncertain because of the small number of patients treated.

Conclusion

The authors report a case of chordoid glioma of the third ventricle. This neoplasm is a recently described, unique pathologic entity that has been added to the WHO glioma classification. The anatomic location of chordoid glioma and the frequent adherence to the hypothalamus does not easily allow a complete surgical excision in the majority of cases. Long term follow-up and further study are needed to obtain accurate diagnosis, prognosis, and effect of treatment of this tumor.

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