

Chordoid Glioma with Psychosis: Case Report

Samuel Estronza, MD; Fanor M. Saavedra, MD; Orlando De Jesus, MD;
Emil A. Pastrana, MD

Objective: A rare, low-grade tumor found in the hypothalamus and anterior third ventricle, a chordoid glioma presents a challenge to neurosurgeons: Its successful resection is complicated by its inconvenient location.

Case Description: A 42-year-old male patient presented with a 1-year history of major depressive disorder, with psychotic features associated with generalized tonic-clonic seizures. Brain magnetic resonance imaging (MRI) with contrast revealed a suprasellar mass extending into the third ventricle. To resect the lesion, an interhemispheric transcallosal transventricular subfornical approach was used. Pathology revealed a chordoid glioma. The patient had a complicated post-operative period that included the development of neurogenic diabetes insipidus, followed by intractable hyponatremia and death (caused by malignant brain edema).

Conclusions: A chordoid glioma is a rare neoplasm that, in 2000, was incorporated into the World Health Organization (WHO) classification of central nervous system tumors. Due to its rarity, it is seldom considered in the differential diagnosis of suprasellar masses. Moreover, its unusual presentation and difficult location present a challenge for surgical and medical management. [*PR Health Sci J* 2018;37:174-176]

Key words: Chordoid glioma, Hypothalamic, Diabetes insipidus, Psychosis

In 1993 the World Health Organization (WHO) ratified a comprehensive classification of neoplasms affecting the central nervous system. The classification of brain tumors is based on the premise that each type of tumor results from the abnormal growth of a specific cell type and that said tumor's behavior correlates with the basic cell type. Tumor classification dictates the choice of therapy and predicts prognosis. A chordoid glioma is a rare low-grade tumor described for the first time in 1998 as a distinct clinical-pathological entity and classified as a WHO grade II glioma (1). Patients with this neoplasm typically present with nonspecific symptoms of obstructive hydrocephalus, including nausea and vomiting. Additional symptoms, including visual disturbances, endocrine imbalances, and dysautonomia, have been reported (2). The histopathological features of a chordoid glioma are consistent with those of a slowly growing neoplasm. Gross total surgical resection would be curative due to the inherent pathological behavior of the neoplasm. However, due to its anatomical location, this tumor's prognosis is comparatively poor. To our knowledge this is the first report of a patient with a chordoid glioma presenting with psychosis.

Case Report

History and Examination

A 42-year-old, right-handed male was referred to our service after a generalized tonic-clonic seizure following a 1-year treatment for acute-onset major depressive disorder with

psychosis. On evaluation he was found confused, agitated, and oriented only in person; in addition, he was behaving aggressively towards family members and claimed to be hearing voices. His cranial nerves were normal, and there was no evidence of weakness. His laboratory examination was notable for hyponatremia. A computed tomographic (CT) scan showed a large suprasellar hyperdense mass extending upward into the third ventricle. Magnetic resonance imaging (MRI) revealed a 2.5 x 4.5 x 5.0-centimeter mass centered at the hypothalamus, with extension into the third ventricle (Figure 1). On T1-weighted images, the mass was isointense to the brain; it was hyperintense on T2-weighted images. It showed marked enhancing with a gadolinium injection. There was a small cystic area at the inferior portion of the tumor.

Operation

A subtotal tumor resection was performed through a right parasagittal craniotomy using an interhemispheric transcallosal transventricular subfornical approach. The tumor was rubbery and firmly attached to the lateral and inferior structures.

Neurosurgery Section, Department of Surgery, University of Puerto Rico Medical Sciences Campus, San Juan, PR

The author/s has/have no conflict/s of interest to disclose.

Address correspondence to: Emil A. Pastrana, MD, Neurosurgery Section, Department of Surgery, University of Puerto Rico, Medical Sciences Campus, P.O. Box 365067, San Juan, PR 00936. Email: emilapastrana@gmail.com

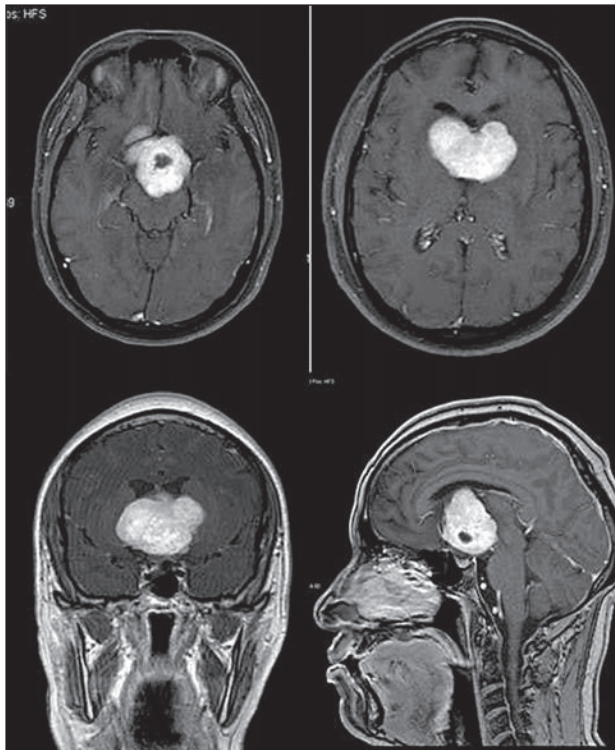


Figure 1. Pre-operative MRI, T1W sequence with contrast. A 2.5 x 4.5 x 5.0-centimeter mass centered at the hypothalamus, with extension into the third ventricle demonstrating marked enhancing with a gadolinium injection and associated cystic area at the inferior portion of the tumor.

Pathological findings

The histological sections showed moderate cellularity consisting of irregular clusters and cords of oval to polygonal epithelioid cells embedded in a mucinous and vacuolated stroma (Figure 2A). The tumor cells showed abundant eosinophilic cytoplasm, round to oval uniform nuclei with finely granular chromatin, and small nucleoli. Prominent interstitial lymphoplasmacytic infiltrates were noticeable, particularly at the tumor–brain interface, where they formed a well-defined thick rim; numerous Russell bodies which are uniquely seen in primary glial neoplasms were observed. Neither necrosis nor mitosis (either of which would indicate a high-grade tumor) was seen. Immunohistochemically, the tumor cells showed strong diffuse expression of glial fibrillary acidic protein (GFAP) (Figure 2B) and of vimentin. The tumor focally reactive for epithelial membrane antigen (EMA).

The histopathologic and immunohistochemical features were consistent with those of a chordoid glioma. The autopsy report revealed a congested brain with a loss of white matter and gray matter differentiation. The hypothalamic region and tuber cinereum were observed to have undergone ischemic changes.

Postoperative course

Postoperatively, the patient was alert and followed commands. Initially he developed a transient episode of diabetes insipidus

with hypernatremia, followed by severe and persistent hyponatremia. Due to his eventual decreased level of consciousness, he required mechanical ventilation. He was weaned from the ventilator but had sudden respiratory difficulty that required intubation and, failing to regain consciousness, rapidly progressed to a coma and death.

Discussion

A chordoid glioma is a rare neoplasm that, in 2000, was incorporated into the World Health Organization (WHO) classification of central nervous system tumors (2,3,4). Brat et al. (5) reported the first 8 cases in 1998. Only 55 cases have been reported in the literature (6,7,8). These tumors typically originate at the anterior aspect of the third ventricle, with attachment to hypothalamic and suprasellar structures. Carrasco et al. (9) suggested that they originate from the lamina terminalis and/or floor of the third ventricle. The histogenesis of this kind of tumor is still uncertain, but ultrastructural data indicate a possible derivation from ependymal cells (6,7,10), which may represent a subtype of ependymoma (7,10). The mean age of presentation is 46 years, with a female predominance of 2:1 (11).

Radiologically, these tumors are well circumscribed and located in the region of the hypothalamus/anterior third ventricle, with uniform and intense enhancement (12). They are hyperdense to gray matter on a CT scan, and isointense on a T1-weighted MRI, and slightly hyperintense on a T2-weighted MRI. Most tumors are solid, but in 25% of them, a small central cystic area is present (11,12). Calcifications have been reported in only 2 cases (13). Histologically, it is considered a low-grade tumor, with slow growth consisting of clusters and cords of epithelioid cells embedded in a mucinous matrix (4,11,13,14).

Different approaches to the anterior third ventricle have been described; among them are the trans-cortical, the sub-frontal, and the interhemispheric transcallosal. All of them are associated with complications, such as hypothalamic or forniceal injury. The trans-cortical and sub-frontal approaches were not selected in the case under discussion because of the degree of brain parenchyma retraction that would have been needed to safely approach the lesion. The transcallosal interhemispheric approach offers the surgeon a corridor with minimal retraction.

Total resection is the treatment of choice (11). On many occasions in the past, this has not been possible without serious residual neurologic deficits (13). Thirty-two percent of past cases died in the perioperative period (11). Pulmonary embolism has been associated with several cases, but its relation to the disease remains unclear (2,4,10,11). The most frequent complication is hypothalamic dysfunction. In patients with subtotal resections, residual tumor has remained stable or grown slowly.

Conventional radiation therapy has little effect on any residual tumor (2,11,4). Radiosurgery has shown better results, with the residual tumor remaining stable in such cases (6,10,12,14).

We present a case of a chordoid glioma that deviated from the classical presentation. Our patient's tumor, located at the

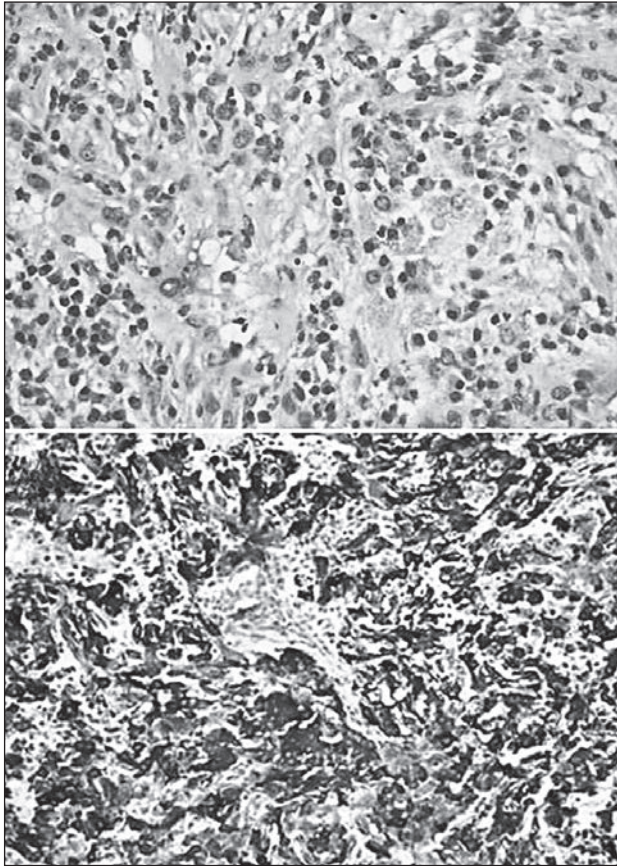


Figure 2. A) H&E of surgical specimen. Tumor cells showed abundant eosinophilic cytoplasm, round to oval uniform nuclei with finely granular chromatin and small nucleoli. B) Immunohistochemically, the tumor cells showed strong diffuse expression of glial fibrillary acidic protein and of vimentin.

anterior aspect of the third ventricle, was in close proximity to the posterior and lateral aspects of the hypothalamus. Experimental lesions produced in these areas in animals can produce a range of reactions, including rage, fear, aggression, and depression (15).

Resumen

Objetivo: El cordoide glioma es una neoplasia rara. Aunque es considerado una neoplasia de bajo grado, debido a su localización en el hipotálamo y la parte anterior del tercer ventrículo, este presenta un reto para neurocirujanos para el manejo adecuado con resección completa sin riesgo de morbilidad y mortalidad significativa en su fase post-operatoria. **Descripción de caso:** Un paciente masculino de 42 años de edad con historial médico pasado de desorden de depresión mayor con rasgos psicóticos presentó con convulsiones tónico-clónicas generalizadas. Resonancia magnética cerebral con contraste demostró una masa supraselar extendiendo hacia la parte anterior del tercer ventrículo. Para la resección de la masa, se hizo

un abordaje inter-hemisférico transcalsoso transventricular y subfornicial. La patología reveló un glioma cordoide. El paciente tuvo una fase post-operatoria complicada por el desarrollo de diabetes insípida, seguido por hiponatremia severa y refractaria. Finalmente falleció por edema cerebral maligno. **Conclusiones:** El glioma cordoide es una neoplasia rara incorporada en la clasificación de 2000 de la Organización Mundial de Salud (WHO por siglas en inglés). Raramente es considerado en el diagnóstico diferencial de masas supraselares. De todos modos, su presentación inusual y localización difícil presentan un reto para su manejo quirúrgico y/o médico.

References

1. Banan R, Hartmann C. The new WHO 2016 classification of brain tumors-what neurosurgeons need to know. *Acta Neurochir (Wien)* 2017;159:403–418.
2. Buccoliero AM, Caldarella A, Gallina P, Di Lorenzo N, Taddei A, Taddei GL. Chordoid glioma: clinicopathologic profile and differential diagnosis of an uncommon tumor. *Arch Pathol Lab Med* 2004;128:141–145.
3. Kawasaki K, Kohno M, Inenaga C, Sato A, Hondo H, Miwa A, Fujii Y, et al. Chordoid glioma of the third ventricle: a report of two cases, one with ultrastructural findings. *Neuropathol* 2009;29:85–90.
4. Pomper MG, Pässe TJ, Burger PC, Scheithauer BW, Brat DJ. Chordoid glioma: a neoplasm unique to the hypothalamus and anterior third ventricle. *AJNR Am J Neuroradiol* 2001;22:464–469.
5. Brat DJ, Scheithauer BW, Staugaitis SM, Cortez SC, Brecher K, Burger PC. Third ventricular chordoid glioma: a distinct clinicopathologic entity. *J Neuropathol Exp Neurol* 1998;57:283–290.
6. Cenacchi G, Roncaroli F, Cerasoli F, Ficarra G, Merli GA, Giangaspero F. Chordoid Glioma of the third ventricle: an ultrastructural study of three cases with histogenetic hypothesis. *Am J Surg Pathol* 2001;25:401–405.
7. Jain D, Sharma MC, Sarkar C, Suri V, Rishi A, Garg A, Vaishya S. Chordoid glioma: report of two rare examples with unusual features. *Acta Neurochir (Wien)* 2008;150:295–300.
8. Vanhauwaert DJ, Clement F, Van Dorpe J, Deruytter MJ. Chordoid glioma of the third ventricle. *Acta Neurochir (Wien)* 2008;150:1183–1191.
9. Carrasco R, Pascual JM, Reina T, Nieto S, Linera J, Sola RG. Chordoid glioma of the third ventricle attached to the optic chiasm. Successful removal through a trans-lamina terminalis approach. *Clin Neurol Neurosurg* 2008;110:828–833.
10. Reifenberger G, Weber T, Weber RG, Wolter M, Brandis A, Kuchelmeister K, et al. Chordoid glioma of the third ventricle: immunohistochemical and molecular genetic characterization of a novel tumor entity. *Brain Pathol* 1999;9:617–626.
11. Tomani H, Kamehiro M, Oguchi M, Higashi K, Yamamoto I, Njima T, et al. Chordoid glioma of the third ventricle: CT and MR findings. *J Comput Assist Tomogr* 2001;24:336–338.
12. Dziurzynski K, Delashaw JB, Gultekin SH, Yedinak CG, Fleseriu M. Diabetes insipidus, panhypopituitarism, and severe mental status deterioration in a patient with chordoid glioma: case report and literature review. *Endocr Pract* 2009;15:240–245.
13. Iwami K, Arima T, Oooka F, Fukumoto M, Takagi T, Takayasu M. Chordoid glioma with calcification and neurofilament expression: case report and review of the literature. *Surg Neurol* 2009;71:115–120.
14. Sangoi AR, Dulai MS, Beck AH, Brat DJ, Vogel H. Distinguishing chordoid meningiomas from their histologic mimics: an immunohistochemical evaluation. *Am J Surg Pathol* 2009;33:669–681.
15. Plum F, Uiter RV. Nonendocrine diseases and disorders of the hypothalamus. In: Reichlin S, Baldessarini RJ, Martin JB, eds. *The Hypothalamus*. New York, NY: Raven Press; 1978:415–473.