Case Report

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Chordoid glioma of right lateral ventricle in young male: a case report

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Introduction and importance: Chordoid glioma is an extremely rare, slow-growing brain tumor. This report discusses the clinical presentation, imaging characteristics, surgical approach, histopathology, and prognosis. The review of literature provides an overview focusing on diagnostic challenges and treatment strategies.

Case presentation: A 21-year-old male presented with progressive blurring of vision, headache and intermittent convulsive seizures over a period of 2 months. Computerized tomography and magnetic resonance imaging revealed a mass in the right lateral ventricle, possibilities of central neurocytoma or ganglioglioma. Surgical resection was performed, and histopathological examination confirmed the diagnosis of chordoid glioma through a combination of histological and immunohistochemical analyses.

Clinical discussion: Chordoid glioma is a distinct clinicopathologic entity, recognized for its unique histological and immunohistochemical features. It is crucial to differentiate chordoid gliomas from other neoplasms due to differences in treatment approaches and prognosis.

Conclusion: Chordoid glioma, although rare, should be considered in the differential diagnosis of lateral ventricular tumors, especially in young adults presenting with visual and neurological symptoms. Early diagnosis and surgical intervention are crucial for improving outcomes.

Keywords: brain tumor, chordoid glioma, immunohistochemistry, lateral ventricle

Introduction

Chordoid glioma is an extremely rare, slow-growing brain tumor first described by Wanschitys *et al* in 1995 as a peculiar variant of meningioma[1]. Later in 1998, Brat *et al* first used the term chordoid glioma and defined it to be primarily located within the third ventricle and hypothalamus[2]. Despite its generally benign histological nature, the tumor's location often results in significant clinical challenges due to its proximity to vital neuroanatomical structures such as the hypothalamus and thalamus[3]. Chordoid gliomas account for a very small number of brain tumors, with fewer than 100 cases reported in the literature and very few cases in the lateral ventricle[4,5]. Although most commonly seen in adults, this tumor has a wide range of age distribution with cases reported from 15 to 75 years old[6].

HIGHLIGHTS

- Chordoid glioma is a distinct clinicopathologic entity, recognized for its unique histological and immunohistochemical features.
- We present a case of 21-year-old male who had a mass in the right lateral ventricle, possibilities of central neurocytoma or ganglioglioma. Surgical resection was performed, and histopathological examination confirmed the diagnosis through a combination of histological and immunohistochemical analyses.
- Chordoid glioma, although rare, should be considered in the differential diagnosis of lateral ventricular tumors, especially in young adults

Given its rarity, chordoid glioma is often misdiagnosed preoperatively as other ventricular tumors, such as subependymoma, meningiomas, ganglioglioma or hypothalamic gliomas, which can significantly affect the therapeutic approach and prognosis[7]. The unique combination of imaging characteristics and histopathological features are critical for diagnosis, making it essential to distinguish chordoid gliomas from other neoplasms in the differential diagnosis[8]. The origin of glioma in ventricular system is thought to be ependymal cells lining the ventricular cavity. In cases of choroid glioma, specialized cells called tanycytes and in third ventricle locations multipotent cells from Rathke's Cleft have been proposed as a cell of origin.

The clinical features of chordoid gliomas are predominantly secondary to their space-occupying nature and local compressive

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properties and occur with symptoms such as headache, visual defect, memory loss, and endocrinological dysfunction[9]. Despite the fact that surgical excision is the cornerstone of treatment, the rarity and typically low morbidity of the tumors have contributed to sparse information about their surgical outcomes. Although in most cases considered indolent, few reports of case series have described cases with aggressive behavior[10-12]. As part of an attempt to contribute to the limited literature, this case report documents the experience of an ultra-rare tumor within the lateral ventricle and contributes to expanding knowledge on its clinical and surgical considerations.

Case presentation

This is a case of a 21-year-old male with an otherwise unremarkable medical history who came complaining of a gradual decline in eyesight over 2 months, as well as recurrent seizures and headaches. He also had bouts of confusion along with issues with short-term memory and excessive sleep during the day. Upon testing the patient, he was found to be lethargic and needed external stimuli to stay awake, making him drowsy during the neurological exam. While his level of consciousness was impaired, the only notable finding on his cranial nerve exam was papilledema. In combination, his motor exam showed preserved strength but increased muscle tone, while sensory and cerebellum functions were normal. He did not have any obvious risk factors for having intra cranial lesions such as head injury, other neurologic abnormalities, or family history of cancers or substance abuse. Due to his concerning and rapidly worsening symptoms, a thorough work up was started right away.

Investigations

Initial hematological and biochemical investigations were within normal limits, ruling out systemic causes of his symptoms. Magnetic resonance imaging (MRI) of the brain was performed, revealing a well-defined, poorly enhancing mass in the right lateral ventricle attached to and displacing septum pellucidum. The lesion measured 4.9 cm in maximum diameter and was hypointense on T1-weighted images and hyperintense on T2-weighted images. On non-contrast computerized tomography (CT), the lesion was isodense with evidence of obstructive hydrocephalus, as indicated by dilation of the lateral and third ventricles (Fig. 1).

Although the MRI findings were indicative for chordoid glioma as pointed out by previous publications[13-15], due to its scarcity this was not the primary tumor for the pre-diagnosis. Differential diagnoses considered at this stage included central neurocytoma, subependymoma or ganglioglioma. Additional imaging with contrast-enhanced MRI helped further delineate the tumor's margins and its relationship to adjacent brain structures. A biopsy was not performed preoperatively due to the tumor's deep-seated location and the associated risks.

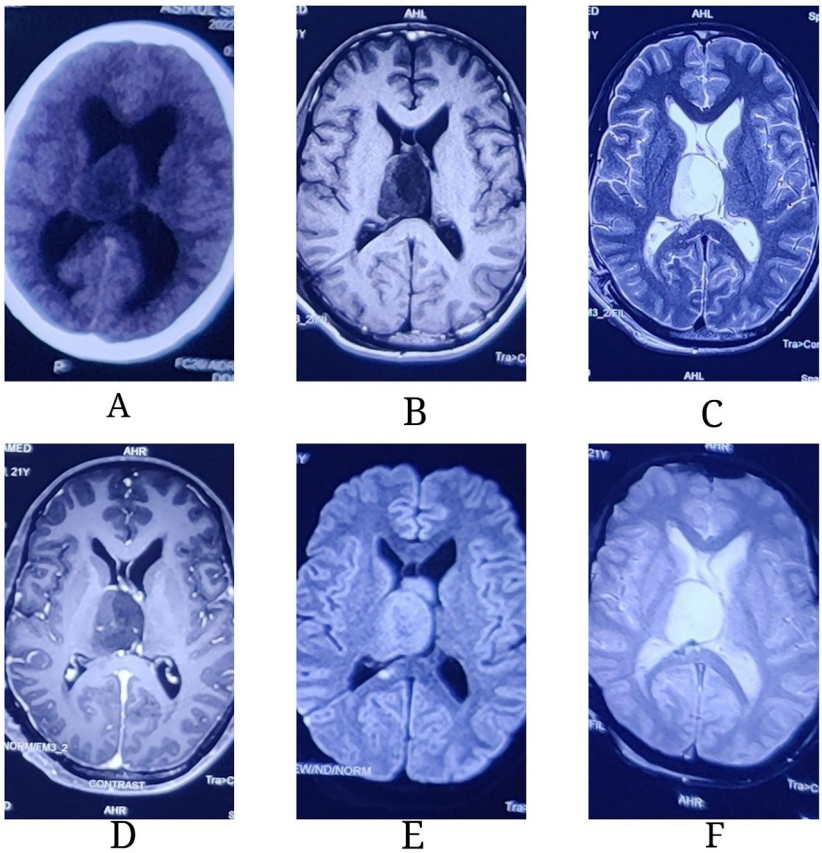

Figure 1. (A) CT scan showing isodense lesion with obstructive hydrocephalus, (B) T1 weighted MRI shows isointense lesion, (C) T2 weighted MRI shows hyperintense lesion, (D) T1 contrast MRI showing no enhancement and (E) no diffusion restriction and (F) no blooming within lesion.

Surgical management

Given the tumor's size, location, and the patient's worsening neurological condition—along with the potential risks of incomplete resection or postoperative hydrocephalus—a decision was made to proceed with cerebrospinal fluid (CSF) diversion via a right ventriculoperitoneal shunt. Following an improvement in neurological status, definitive surgical intervention was planned.

A right parietal craniotomy was performed, utilizing an interhemispheric transcallosal approach to access the lateral ventricle under neuronavigation guidance. Intraoperatively, the tumor appeared firm, yellowish, moderately vascular, and was strongly adherent to the septum pellucidum, fornix, and thalamostriate veins, with extension into the third ventricle (Fig. 2F). Given its adherence to critical neurovascular structures, complete resection was not feasible, and a small residual portion was left behind to minimize the risk of neurological deficits. The procedure was completed without intraoperative complications.

Postoperatively, the patient was closely monitored in the intensive care unit and was gradually weaned off intensive care support before being transferred to the neurosurgical ward. His recovery remained uneventful, and he was discharged on the 10th postoperative day with an intact neurological status and no deficits.

Histopathological findings

Histopathological analysis of the resected tumor revealed clusters of oval-shaped cells embedded within a variably mixed stroma, exhibiting a distinct choroidal morphology. In certain areas, the tumor cells appeared more polygonal and fusiform, with focal regions displaying a fibrosing pattern. Immunohistochemical evaluation demonstrated strong positivity for glial fibrillary acidic protein (GFAP) and CD34, while synaptophysin staining was negative. There was focal positivity for epithelial membrane antigen (EMA) and vimentin, though these findings were not diagnostically significant. The Ki-67 proliferation index was measured at less than 5%, consistent with a low-grade neoplasm (Fig. 2A–E).

The differential diagnosis included chordoid meningioma, which was excluded due to the absence of psammoma bodies and cellular whorls. Likewise, chordoma was ruled out as there was no evidence of physaliphorous cells. The immunohistochemical profile, particularly the strong expression of GFAP and CD34, confirmed the diagnosis of chordoid glioma. The patient provided informed consent for the publication of his clinical details and radiological findings.

This case report has been prepared in accordance with the scare 2023 criteria[16].

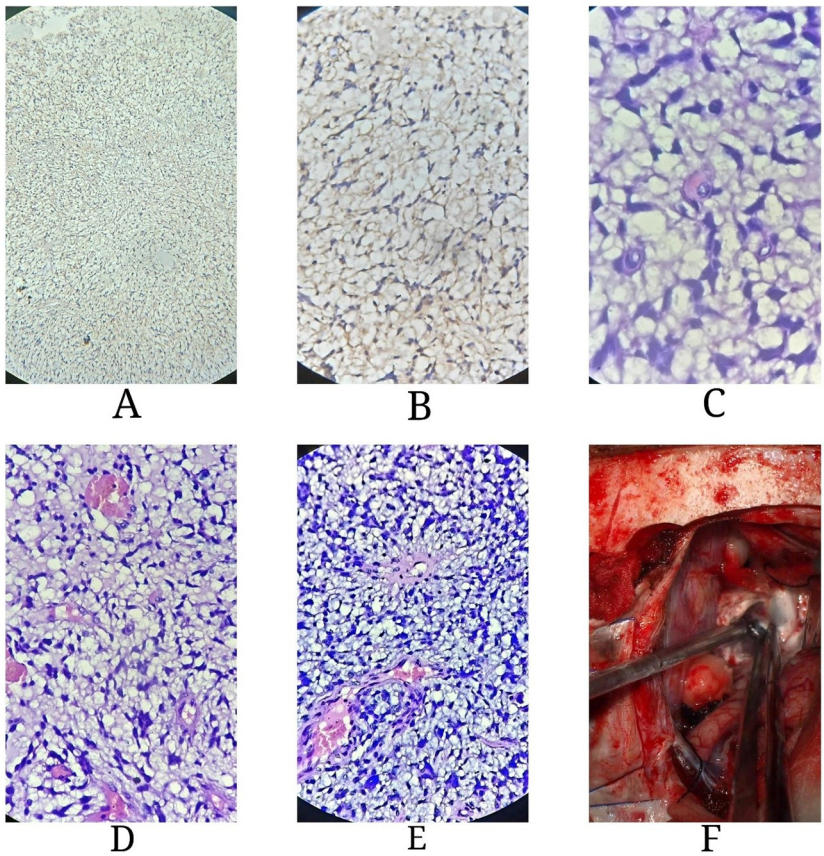

Figure 2. (A&B) Strong positive for glial fibrillary acidic protein stain; (C, D, E) Haematoxylin & eosin staining showing clusters of oval cells in myxoid stroma and choroidal appearance; (F) firm yellowish tumor attached to septum pellucidum and fornix.

Discussion

Chordoid glioma is a distinct clinicopathologic entity, recognized for its unique histological and immunohistochemical features. It is crucial to differentiate chordoid gliomas from other neoplasms due to differences in treatment approaches and prognosis. The expression of GFAP, vimentin, and CD34 is characteristic of chordoid gliomas, while other markers, such as S-100 protein, EMA, and cytokeratin, provide additional diagnostic clues[17].

Surgical resection remains the cornerstone of treatment, aiming for gross total resection due to the tumor's benign nature and low proliferative index[15,18]. However, the tumor's location near critical brain structures often makes complete resection challenging, and the risk of postoperative complications such as diabetes insipidus, hypothalamic dysfunction, and memory impairment is significant[2]. A review of the literature indicates that adjuvant radiotherapy has been utilized in nine reported cases. However, clinical follow-up data are limited and relatively short, with a mean duration of 22.5 months. Surgical intervention is associated with considerable morbidity and mortality, largely influenced by the tumor's anatomical location rather than its histopathological characteristics[19]. Despite these challenges, gross total resection offers the best chance for a favorable outcome, with lower rates of recurrence compared to subtotal resection[3].

Diagnostic considerations

The localization-related symptoms experienced by chordoid gliomas patients are nonspecific and chiefly due to pathology's preference for the anterior third ventricle and often suprasellar area, as well as lateral ventricle involvement. Manifestations observed clinically are mainly due to the presence of obstructive hydrocephalus, endocrine dysfunction, hypothalamic feeding disorder with electrolyte imbalance, and optic nerve compression causing visual disturbance[5].

Imaging characteristics

A chordoid glioma is an oval, well-defined, non-invasive lesion, Appearing on the MRI as a strikingly, well-outlined, oval, non-invading tumor. If they do not have any distinctive signaling features, their typical location in the anterior third ventricle could point toward the diagnosis. In some lesions, cystic component radiologically may be seen. The solid portion shows vivid contrast enhancement, on T1-weighted imaging (T1WI) it is hypo- to isointense while on T2-weighted imaging (T2WI), it is iso- to hyperintense. DWI shows no restriction of diffusion and the lesions demonstrate lowered level of N-acetyl aspartate and elevated level of choline which indicate low grade tumor on magnetic resonance spectroscopy[20].

Chordoid gliomas pose significant surgical challenges due to their deep-seated location and close proximity to critical neurovascular structures. Achieving gross total resection (GTR) is often difficult, frequently resulting in subtotal resection (STR), which is associated with higher recurrence rates. A systematic review of 81 cases found that GTR was significantly correlated with improved progression-free survival compared to STR. The choice of surgical technique plays a crucial role in determining postoperative outcomes. The trans-lamina terminalis approach has been associated with a trend toward lower postoperative morbidity compared to other approaches. Consequently, increasing attention is being directed toward preoperative planning strategies to enhance the extent of maximal safe resection while minimizing complications[21].

The integration of advanced intraoperative technologies, including neuronavigation, diffusion tensor imaging (DTI) tractography, intraoperative MRI (IoMRI), and fluorescence-guided surgery, has significantly enhanced the precision of tumor resection. Neuronavigation systems, in particular, facilitate accurate preoperative planning and real-time intraoperative guidance, enabling more extensive tumor removal while minimizing damage to adjacent healthy tissue. A randomized controlled trial demonstrated that neuronavigation significantly increases the extent of maximal safe resection, with an average residual tumor volume of 13.8% compared to 28.9% using conventional techniques. These advancements have been associated with improved progression-free and overall survival in glioma patients, though their specific impact on chordoid gliomas remains to be fully elucidated[22].

Postoperative follow-up is essential, with serial imaging to monitor for any signs of recurrence. In cases of subtotal resection or recurrence, adjuvant radiation therapy may be considered, although its role remains uncertain due to the rarity of this tumor and the lack of large-scale studies[4]. Recent advances in neuroimaging and surgical techniques have improved the diagnosis and management of chordoid gliomas, though the optimal treatment approach continues to be refined[6].

Long-term prognosis and follow-up strategies for chordoid glioma patients require more attention, particularly due to the risk of recurrence and postoperative complications. Given the tumor's rarity and the complexity of its surgical resection, it is essential to establish standardized protocols for long-term monitoring. Research on glioma patients has demonstrated that factors such as age, tumor grade, and the extent of resection play a critical role in determining patient outcomes. A study of 335 glioma patients found that a complete resection correlates with better prognosis and reduced recurrence rates[23]. Therefore, long-term follow-up strategies should include regular imaging (MRI and CT) to monitor for recurrence, along with clinical assessments to detect early signs of complications such as memory impairment, diabetes insipidus, or hypothalamic dysfunction. These monitoring protocols are essential to maximize survival and quality of life for patients who undergo resection of chordoid gliomas.

The preoperative radiological differentiation of chordoid gliomas from other intraventricular tumors is a significant challenge due to the overlap in imaging characteristics. These tumors often present as well-defined, ovoid masses in the anterior third ventricle, which can be easily confused with other lesions such as neurocytomas or meningiomas. However, chordoid gliomas have certain distinguishing features that can aid in their identification. MRI typically shows a hyperintense lesion on T2-weighted images, with uniform contrast enhancement, and the presence of a cystic component in some cases. Furthermore, CT scans reveal hyperdensity in most cases, providing additional diagnostic clues. A detailed analysis of these imaging characteristics has been outlined in several studies, such as those found in a comprehensive review of intraventricular tumors, which emphasizes the importance of contrast-enhanced MRI for accurate preoperative diagnosis[24]. Incorporating these findings into the manuscript will not only enhance its diagnostic value but

also help clinicians in making more accurate preoperative assessments and tailoring their surgical approach accordingly.

This is a single case report with a relatively very short period of follow-up thus, these findings are not generalizable. However, as pointed throughout the manuscript, chordoid gliomas are rare and lateral ventricle seated lesions are even rarer. Thus, this report may serve as an additional evidence for more comprehensive studies.

Conclusion

Chordoid glioma, although rare, should be considered in the differential diagnosis of lateral ventricular tumors, especially in young adults presenting with visual and neurological symptoms. Early diagnosis and surgical intervention are crucial for improving outcomes, with gross total resection being the preferred treatment. Histopathological and immunohistochemical analysis remains vital for definitive diagnosis, and ongoing research is needed to explore the role of adjuvant therapies in incompletely resected tumors. Long-term follow-up is essential to monitor for recurrence and manage any late-onset complications.

Chordoid glioma, though rare, should be included in the differential diagnosis of lateral ventricular tumors, particularly in young adults presenting with visual and neurological symptoms. Early diagnosis and surgical intervention are crucial for improving outcomes, with gross total resection being the preferred treatment. Histopathological and immunohistochemical analyses are vital for definitive diagnosis, and ongoing research is needed to explore the role of adjuvant therapies in cases of incomplete resection. Long-term follow-up is essential to monitor for recurrence and manage any late-onset complications.

Ethical approval

Ethical approval not required for case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author's contribution

Writing the paper: K.K.N.; study concept or design, data collection, data analysis or interpretation: G.D., B.C.

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