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Chordoid glioma with uncommon papillary and fusiform structures

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Abstract

Chordoid glioma (CG) is a rare, low-grade glioma predominantly located in the third ventricle, often presenting diagnostic challenges due to its similarity to other tumors. In this report, we presented the clinicopathological features of five cases of CG. The median age was 44 years, with a notable female predominance. All tumors were situated in the third ventricle and frequently misdiagnosed as craniopharyngioma via magnetic resonance imaging (MRI). Microscopically, all cases exhibited typical histopathological features of CG, which accounted for very small proportion in individual case. However, some also presented additional structures, including solid, fusiform, and papillary formations. Papillary and/or solid structures were notably present in cases of incomplete resection, whereas they were absent in cases of complete resection. Immunohistochemical analysis revealed that CK7 showed greater sensitivity in CG compared with pan-CK. Additionally, besides several common immunophenotypic features and *PRKCA* D463H mutation, diffuse and strong positivity for SOX2 was observed in all cases. Similarly, it was present in other gliomas, but not detected in craniopharyngiomas, chordomas and meningiomas. Therefore, these findings not only broadened the histological characteristics, immunological phenotypes, and prognostic factors associated with CG, while also reaffirmed its classification as a distinct tumor type.

Introduction

Chordoid glioma (CG) is a rare, low-grade neuroepithelial tumor primarily located in the anterior portion of third ventricle and predominantly affects adult females. To date, more than 100 cases of CG have been reported. The 2021 WHO Classification of Tumors of the Central Nervous System （CNS） has streamlined the terminology “chordoid glioma of the third ventricle” as noted in the 2016 WHO Classification to “chordoid glioma” [1]. Patients with CG often present with symptoms such as headaches, nausea, vomiting, visual deficits, and memory impairment. In addition to the third ventricle, CGs have been identified in other locations, including frontoparietal occipital lobes, cerebellar hemisphere [2], [3], and pineal region [4].

Despite being classified as low-grade tumors, the prognosis of CG is not universally favorable due to the tumor’s location and associated postoperative complications. Imaging studies typically reveal well-defined masses. Histological analysis characteristically shows epithelioid cells arranged in clusters or cords within abundant mucinous stroma infiltrated by lymphocytes and plasma cells [5]. However, rare structures such as papillary, solid, and fusiform formations are also described [6], [7]. Additionally, mucinous stroma may be absent in focal areas [8]. Therefore, it is crucial to identify less common histological features to prevent misdiagnosis. Immunohistochemistry typically reveals strong and diffuse positivity for glial fibrillary acidic protein (GFAP), vimentin, and thyroid-transcription factor-1 (TTF-1) in tumor cells [8]. CD34, cytokeratin, and epithelial membrane antigen (EMA) exhibit focal positivity. Recently, the PRKCA D463H mutation has been exclusively identified in CG and suggested to be an oncogenic, gain-of-function mutation driving tumorigenesis [9], [10]. To date, 51 cases of CG with the *PRKCA* D463H mutation have been documented. However, no commercial diagnostic kits are available, and next-generation sequencing (NGS) is not widely implemented in most basic hospitals. SOX2, a member of the SOX transcription factor family, is involved in several cellular processes, including the maintenance of embryonic stem cells pluripotency, differentiation of neural stem cells, and cancer development [11], [12]. The expression of SOX2 is noted in several primary brain neuroepithelial tumors, particularly gliomas [13]. However, its expression in CG remains inadequately documented.

In this study, we presented five cases of CG, detailing their clinical, radiological, histological, immunohistochemical, and genetic characteristics to enhance the current understanding of CG pathology. Moreover, we assessed the sensitivity and specificity of SOX2 as a potential diagnostic marker in CG.

Section snippets

Cases

Five cases of CG were collected from the archives of Department of Pathology of the Second Affiliated Hospital of the Fourth Military Medical University between January 2017 and December 2023. The criteria for selecting CG was based on the 5th edition of the WHO Classification of Tumor of CNS. Essential criteria included the presence of a glial neoplasm with

chordoid features located in the third ventricle. In addition, in order to compare the differentiation of SOX2 in tumors that may be …

Characteristics of clinic and imaging

The clinical characteristics of five cases were summarized in Table 1, which indicated a female predominance of 4:1, with a median age of 44 years (ranging from 31 to 49 years). Three out of five patients were hospitalized due to headaches; notably, case 3 presented with visual deficits. The other two patients were admitted following imaging studies prompted by physical examinations. Magnetic resonance imaging (MRI) revealed well-demarcated solid masses in the suprasellar region and the third …

Discussion

CG is a rare, low-grade glial neoplasm that predominantly occurs in the third ventricle. In our study, we found a female predominance among the patients, with a median age of 44 years, which was slightly younger than reported in previous studies [8], [14], [15]. MRI characteristics of CGs often resemble those of craniopharyngiomas, the most commonly occurring tumors in the suprasellar region. Consequently, distinguishing two tumor types based solely on imaging findings is challenging. Due to …

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We declare that this manuscript is original, has not been published before and is not currently being considered for publication elsewhere.

We confirm that the manuscript has been read and approved by all named authors and that there are no other persons who satisfied the criteria for authorship but are not listed. We further confirm that the order of authors listed in the manuscript has been approved by all of us. …

CRediT authorship contribution statement

Gong Li: Writing – review & editing, Project administration, Methodology. **Liu Xiaoyan:** Methodology, Formal analysis. **Zhang Wei:** Writing – review & editing. **Han Yu:** Validation, Data curation. **Wang Zhuo:** Investigation. **Wang Shumei:** Writing – review & editing, Data curation. **Zhu Shaojun:** Formal analysis. **Feng Lanlan:** Writing – original draft, Investigation, Data curation, Conceptualization. **Mu Xiaorong:** Investigation, Data curation. **Li Junting:** Resources, Data curation. **Wang Yuanyuan:** Validation, …

Declaration of Competing Interest

All authors disclosed no relevant relationships. …

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