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Case report

Magnetic resonance imaging of a third ventricular chordoid glioma ☆,☆☆

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ABSTRACT

Chordoid gliomas are uncommon neoplasms located within the anterior portion of the third ventricle. In this article, we aimed to describe the clinical presentation, magnetic resonance imaging characteristics, histological findings, and surgical treatment applied to a case of chordoid glioma. Chordoid gliomas are typically observed as solid masses within the anterior segment of the third ventricle, characterized by homogenous and vivid enhancement. Despite being classified as a low-grade neoplasm, the outcome of chordoid glioma is often uncertain.

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Introduction

Chordoid gliomas are rare and benign central nervous system tumors first described by Brat et al. in 1998 [1]. Fewer than 100 cases of chordoid glioma have been reported in the literature thus far [2,3]. Chordoid gliomas are classified as Grade II tumors according to the World Health Organization (WHO) classification guidelines, updated in 2016 [4].

Clinical symptoms are often caused by obstructive hydrocephalus and the compression of the optic chiasm, leading to nausea, headache, and visual disturbances. The typical radiological features of this type of tumor include a pure solid mass located in the anterior portion of the third ventricle, which often extends into the hypothalamus, showing strong and homogenous enhancement [2,5]. Although chordoid glioma is considered to be a low-grade and slow-growing tumor, the prognosis is often difficult to predict due to its deep

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location and close relationship with the hypothalamus, optic chiasm, and adjacent arteries [1,6]. Moreover, some perioperative and postoperative complications, such as bleeding, pulmonary embolism, and hypothalamic dysfunction, have been reported [7]. In this paper, we aimed to report the clinical presentation, magnetic resonance imaging (MRI) features, histological findings, and treatment of a case of chordoid glioma.

Case presentation

A 45-year-old woman was admitted to our hospital due to 6-month history of headache, memory deficits, visual disturbances, and gaining 20 kg in 6 months. Right temporal hemianopia and left diffuse visual defect were identified on clinical examination. Endocrine test results revealed normal levels of luteinizing, follicle-stimulating, and thyroid-stimulating hormones, prolactin, estradiol, and progesterone. No history of diabetes mellitus or menstrual disturbances were noted.

The brain MRI with contrast injection revealed a well-defined, multilobulated, solid mass in the anterior aspect of the third ventricle. The mass was isointense on T1-weighted images, with heterogeneous signal intensity on T2-weighted images and fluid-attenuated inversion recovery (FLAIR), compared with that of gray matter. Restricted diffusion was observed, which was compatible with a hypercellularity tumor, and the mass showed vivid and homogenous enhancement. The lesion expanded to the supraoptic recess, compressing the optic chiasm downward and forward, bowing the floor of the third ventricle and infundibulum backward and downward, and compressing the bilateral hypothalamus. The mass also contacted the terminal segment of the right internal carotid artery (ICA), the A1 segment of the right anterior carotid artery (ACA), and the tip of the basilar artery (BA). The adjacent brain parenchyma and the optic tracts were also hyperintense on FLAIR. The pituitary gland and suprasellar cistern appeared intact (Fig. 1).

The patient underwent microsurgery via frontal craniotomy using a transcallosal approach guided by neuronavigation. Intraoperatively, a gray, firm, mildly bleeding mass was identified. Partial tumor resection was performed due to the proximity of the mass to the ACA, BA, and optic chiasm. Histological and immunohistochemical findings revealed a typical chordoid glioma (Fig. 2).

The second operation was performed after 2.5 months. An interhemispheric transcallosal approach was applied, and the tumor was removed to the maximum extent possible. No neurological deficits were noticed postoperatively. The visual disturbance was improved.

Discussion

Chordoid glioma is an unusual, slow-growing brain tumor. In 2019, a systematic review was conducted by Johannes et al. [5], including 73 cases of chordoid glioma, in which they described the radiological features, surgical techniques, and im-

munohistochemistry findings associated with these tumors. The tumor is often found in adults, with an average age of onset of 47 years [8]. However, 5 cases have been reported in pediatric patients to date [9]. Chordoid glioma is more frequently detected in women than in men, with a ratio of 3:1 [7]. According to Leeds et al. [10], chordoid glioma derives from the lamina terminalis and the specialized ependyma covering the lamina terminalis and the structures of the ventricular system.

The most common clinical symptoms associated with chordoid glioma include headache (51.4%), visual deterioration (41.4%), and memory deficits (21%) [5]. Polyuria and other symptoms typically associated with endocrine disorders, such as diabetes mellitus, menstrual disturbance, and weight gain, are less common [5]. Roth et al. [11] stated that lesions in the hypothalamic region can affect the medial and posterior nuclei, leading to hyperinsulinemia, leptin resistance, and reduced sympathetic activity, resulting in uncontrolled weight gain. The author also mentioned that this symptom is commonly detected in pediatric patients with craniopharyngioma [11]. Our patient presented with signs that were suggestive of a potential hypothalamic lesion. The visual field defects were caused by the tumor-induced compression of the optic chiasm and bilateral optic tracts. The significant weight gain could be explained by the extension of the tumor to the hypothalamus.

Chordoid glioma is frequently found at the lamina terminalis of the third ventricle or the suprasellar region and often extends into the hypothalamus [12]. Other locations have also been reported, such as the fourth ventricle, lateral ventricle, thalamus, and parieto-temporal lobe [3,9,13]. These tumors present as solid, round or ovoid, well-defined, and multilobulated masses on cross-sectional imaging. On computed tomography (CT), they appear as hyperdense masses compared to the adjacent brain parenchyma [12]. On MRI, these tumors appear hypo- or isointense on T1-weighted images (96.8%), hyper- or isointense on T2-weighted images (96.9%), with vivid and homogenous enhancement following contrast administration [5]. Intratumoral necrosis, calcification, or cystic components are occasionally detected [5]. Signal intensity of these tumors on diffusion-weighted image is variable according to previous reports. Six cases reported by Yang et al. [3] showed no restricted diffusion. One pediatric case in lateral ventricular reported by Chen et al. [9] was isointense on diffusion-weighted image. Rim and mural nodule enhancement have been reported for tumors found within the fourth ventricle [8,13]. According to Grand et al. [13], chordoid glioma does not show an increase in perfusion compared with the adjacent white matter, with a relative cerebral blood volume (rCBV) of 1. The author explained that the vivid enhancement feature of chordoid glioma is likely the result of a breakdown in the blood-brain barrier rather than intratumoral hypervascularity. However, a case associated with multiple lesions in the ventricular system was reported with an rCBV of 6.95 [14]. In our case, the tumor was located in the anterior aspect of the third ventricle, with the typical signal intensity and characteristic enhancement on MRI.

Tumors of the sellar and suprasellar regions and those found within the third ventricle are the primary differen-

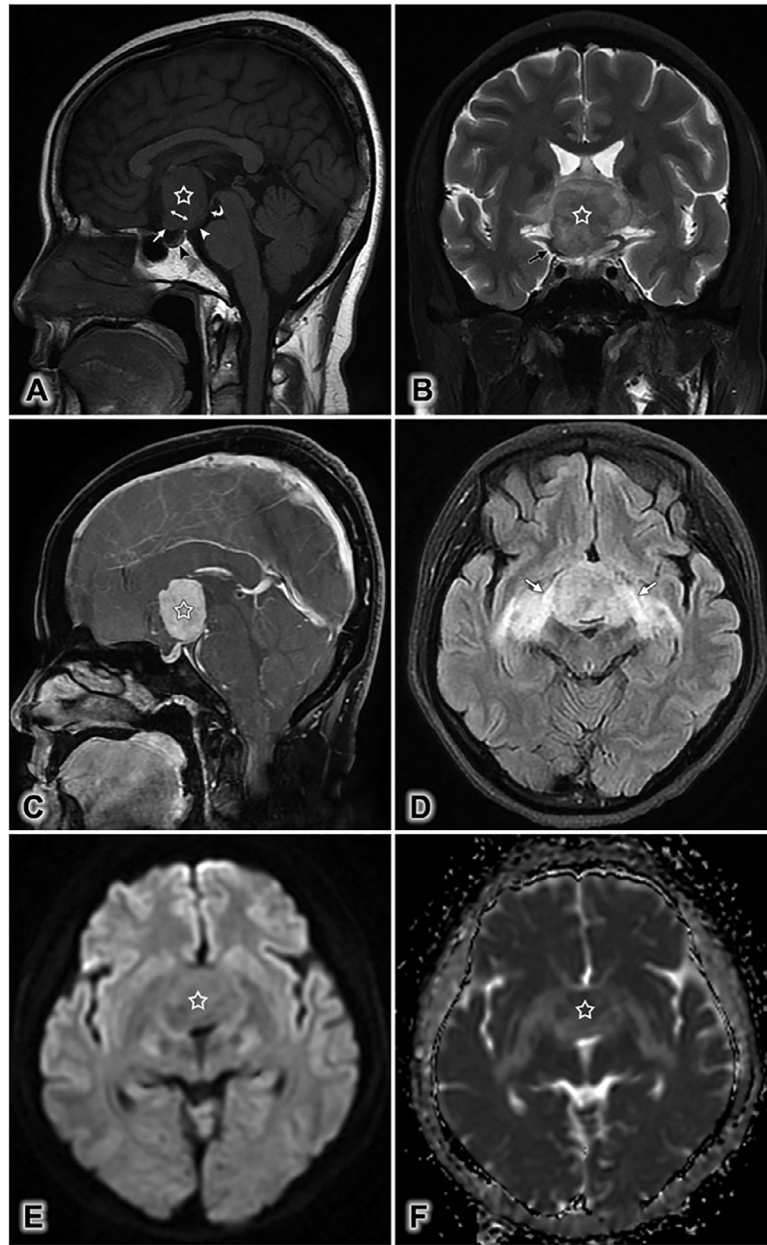


Fig. 1 – Brain MRI showing a well-defined, multilobulated, solid mass in the anterior portion of the third ventricle. (A) Mid-sagittal T1-weighted image showing an isointense mass (star) expanding into the supraoptic recess (double-headed arrow), compressing the optic chiasm downward and forward (arrow), and bowing the floor of the third ventricle (curved arrow) and infundibulum backward and downward (white arrowhead). The pituitary gland and suprasellar cistern appeared intact (black arrowhead). (B) Coronal T2-weighted image showing a heterogenous hyperintense mass (star) contacting the terminal segment of the right ICA (arrow). (C) Sagittal T1-weighted post-contrast image showing the strong and homogenous enhancement of the mass (star). (D) Axial FLAIR showing the edema of the adjacent brain parenchyma and bilateral optic tracts (arrow). (E and F): Diffusion-weighted image (E) and apparent diffusion coefficient image (F) showing the restricted diffusion of the tumor (star). MRI: magnetic resonance imaging; ICA: internal carotid artery; FLAIR: fluid-attenuated inversion recovery. (Color version of the figure is available online.)

tial diagnosis for chordoid gliomas. The mass effects, such as the downward and backward bowing of the floor of the third ventricle and infundibulum and the downward compression of the optic chiasm, associated with an intact pituitary gland and supraoptic recess, represent important findings

that can distinguish chordoid gliomas from tumors of the sellar and suprasellar regions. Third ventricular craniopharyngiomas have been reported in the literature [15] with similar localization as chordoid gliomas. However, craniopharyngiomas often show heterogeneous enhancement, whereas

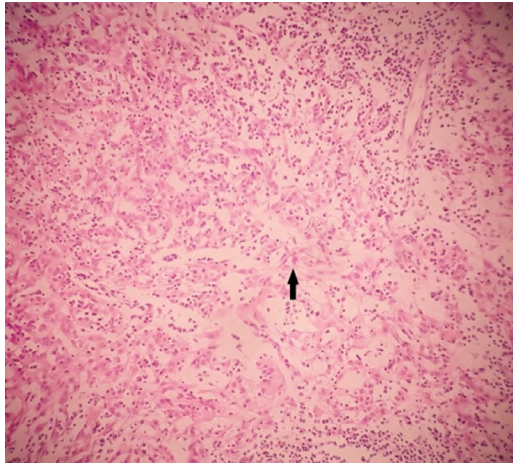


Fig. 2 – Histological imaging demonstrates epithelioid cells arranged in clusters or cords, with stroma containing lymphoplasmacytic infiltrate (black arrow). Hematoxylin and eosin (HE, $\times 100$). (Color version of the figure is available online.)

vid and homogeneous enhancement is a typical finding of chordoid gliomas [5,15].

Surgical management is the treatment of choice for chordoid gliomas. The interhemispheric transcallosal and trans-lamina terminalis approach techniques are commonly used, although temporal transcortical and transsphenoidal approaches have also been reported [2,5]. The gross total resection of the mass is optimal but can often be associated with a high risk of perioperative and postoperative complications [6]. Severe complications can include bleeding, pulmonary embolism, hypothalamic dysfunction, cardiovascular effects, and pneumonia [7,16]. According to Kobayashi et al. [6], partial tumor removal followed by gamma knife radiosurgery represents a promising treatment that is minimally invasive and associated with a lower risk of complications. Besides, gamma knife radiosurgery showed as ineffective therapy in one case reported no change in tumor size after 66 months of treatment with this method [6]. Our patient was approached through an interhemispheric transcallosal approach guided by microscope-based neuronavigation. Partial tumor removal was performed due to the close positioning of the lesion to the optic chiasm, the arteries, and the hypothalamus. No severe peri- or postoperative complications were detected.

Conclusion

A pure solid mass located in the anterior segment of the third ventricle and extending to the hypothalamus, showing vivid and homogenous enhancement, represent typical radiological features of chordoid glioma. The prognosis of this tumor remains uncertain due to its complex relationship with adjacent functional structures.

Ethical statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

Author contributions

Nguyen DH and Nguyen MD contributed to this article as co-first authors. All authors have read the manuscript and agree to the contents.

Patient consent

Informed consent for patient information to be published in this article was obtained.

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