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Endoscope-assisted Trans-lamina Terminalis Resection of Chordoid Glioma at the Third Ventricle: A Case Report

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Abstract

As per the 2021 World Health Organization (WHO) Classification of Tumors of the Central Nervous System, chordoid glioma (CG) is defined as a slow-growing glial neoplasm categorized as grade II tumor. This tumor is primarily located in the anterior part of the third ventricle, often adheres to important surrounding structures, and is hemorrhagic in nature. Therefore, dissecting this tumor is extremely difficult. In this study, we present the case of a 44-year-old man who initially complained of mild headache and was diagnosed with a homogeneous gadolinium-enhanced lesion in the third ventricle via magnetic resonance imaging. The pathological diagnosis based on his biopsy at the previous hospital was CG. The patient demonstrated no neurological deficit at that time, but the tumor had gradually grown, hydrocephalus appeared 2 years after the tumor was detected, and the patient developed short memory disorder and daytime sleepiness. We resected the tumor via the anterior interhemispheric trans-lamina terminalis approach using a microscope and an endoscope. The residual tumor at the blind spot of the microscopic view was resected under an angled rigid endoscopic view using dedicated tools for transsphenoidal surgery. The tumor was grossly resected, and the histopathological diagnosis was CG. Postoperative neurological findings included slight memory disorder and hypothalamic adrenal dysfunction. No tumor recurrence was reported 3 years post resection. The endoscope-assisted anterior interhemispheric trans-lamina terminalis approach was determined useful for CG resection with minimal surgical complications and without tumor recurrence.

Keywords: chordoid glioma, endoscope, lamina terminalis, microscope, third ventricle

Introduction

Chordoid glioma (CG) has been identified as a rare, slow-growing tumor characterized by ovoid or polygonal epithelioid cells with abundant cytoplasm, organized in clusters and chords. The first description of CG of the third ventricle was provided by Brat et al. in 1998.[1)] CG is mainly located in the third ventricle, often strongly adheres to the surrounding vital structures, and is hypervascular; thus, its total resection is deemed to be extremely difficult without any postoperative complications.[2)] Here, we present a case of CG that achieved a gross total resection via an anterior interhemispheric trans-lamina terminalis approach with endoscopic additional resection, with the patient demonstrating only mild consciousness impairment after the surgery and no tumor recurrence for >3 years.

Case Report

In this study, we report on a case of a 44-year-old man who initially presented with an occipital headache. Magnetic resonance imaging (MRI) revealed a tumor located in the third ventricle that was homogeneously enhanced with gadolinium (Gd) (Fig. 1a and b). He underwent biopsy via the right frontotemporal approach at his previous hospital, with the pathological diagnosis being CG. He was thereafter referred to our department. The patient had no pitui-

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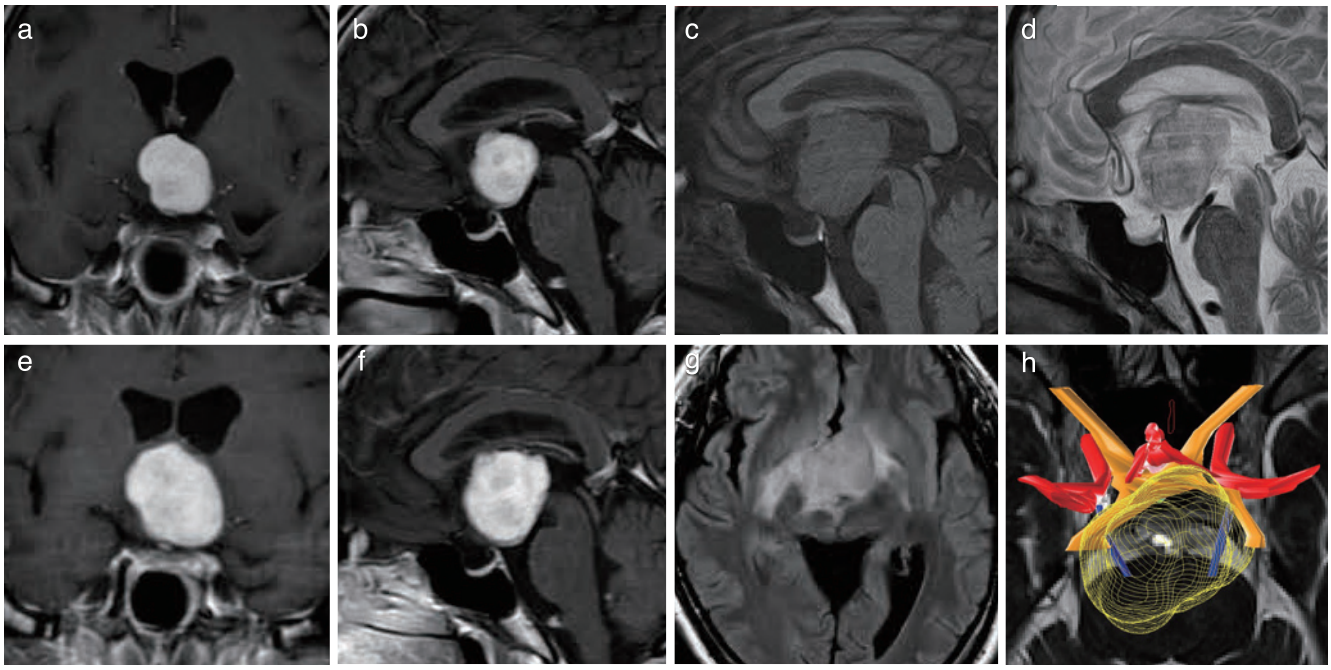

Fig. 1 Magnetic resonance imaging (MRI) at the first visit demonstrated a tumor occupying the third ventricle. Homogenous enhancement with gadolinium (a, b). The tumor gradually grew, and the bilateral ventricles grew for 14 months. Iso-intensity on sagittal T1-weighted imaging (T1WI) (c), low- to iso-intensity on sagittal T2-weighted imaging (d), and homogenously enlarged tumor in the third ventricle on coronal T1WI (e) and sagittal T1WI (f) with gadolinium. Perifocal edema in the bilateral basal ganglia was observed on fluid-attenuated inversion recovery (g). Constructive interference in a steady state with gadolinium (h) showed that the tumor was located dorsal to the chiasm.

tary endocrine dysfunction and neurological deficit at that time. However, the tumor had gradually grown; he was developing hydrocephalus for 14 months. Moreover, the patient developed short memory disorder and daytime sleepiness. His endocrine function demonstrated severe growth hormone deficiency (growth hormone peak value, 8.86 ng/mL). MRI demonstrated hydrocephalus, a homogenously Gd-enhanced tumor occupying the third ventricle (Fig. 1c, d, e, and f), and a high-intensity signal in fluid-attenuated inversion recovery (FLAIR) spread to the bilateral (right > left) basal ganglia (Fig. 1g). Gd-enhanced MRI construct interference in steady state revealed that the tumor was located dorsal to the chiasm (Fig. 1h). We assumed that the origin of the tumor was the laminar terminalis; therefore, we opted for the anterior interhemispheric translamina terminalis approach.

First, we resected the tumor piecemeal and achieved internal decompression, to avoid the hypothalamic arteries (Fig. 2a). The tumor was partially gelatinous but was mostly fibrous, elastic, hard, and bled easily. After internal decompression, the tough, thick, fibrous external wall of the tumor was dissected from the surrounding structures, coagulating the feeding arteries on the tumor surface. The tumor was easily dissected from the left and posterior walls of the third ventricle; however, the tumor severely adhered to the right wall of the third ventricle and the caudal side of the chiasm. It was coincident to the preoperative FLAIR MRI findings. Particularly, the tumor on the right caudal side of the chiasm was fibrous and bled such that this region was suspected to be of tumor origin. After subtotal tumor resection under the microscope, we inserted an angled (30° and 45°) rigid endoscope (Storz®: high-definition type with irrigation sheath) into the tumor cavity via the surgical tract (Fig. 2b). We observed the tumor bed and then resected the residual tumor located at the blind spots (the caudal side of the anterior communicating artery and chiasm) in the microscopic view (Fig. 2 c). At the end of the surgery, we confirmed that no residual tumor or hemorrhage in the tumor bed was evident under endoscopic view.

Post operation, MRI revealed total tumor resection (Fig. 2d and e) without any infarction and hemorrhage. The patient demonstrated mild cognitive dysfunction (impaired short-term memory for 3 months), which gradually improved over time. However, he demonstrated panhypopituitarism, developing postoperative extra-axial cerebrospinal fluid collection (PECC) in the right frontal subdural space, and cerebrospinal fluid (CSF) was collected in the left frontal subcutis (Fig. 2f). He underwent ventriculoperitoneal shunt (VPS) placement via the left posterior horn 22 days post-tumor resection. Further, pulmonary embolism from lower extremity venous thrombosis occurred 31 days postsurgery. Heparinization was initiated immediately, and the patient's circulatory and pulmonary conditions

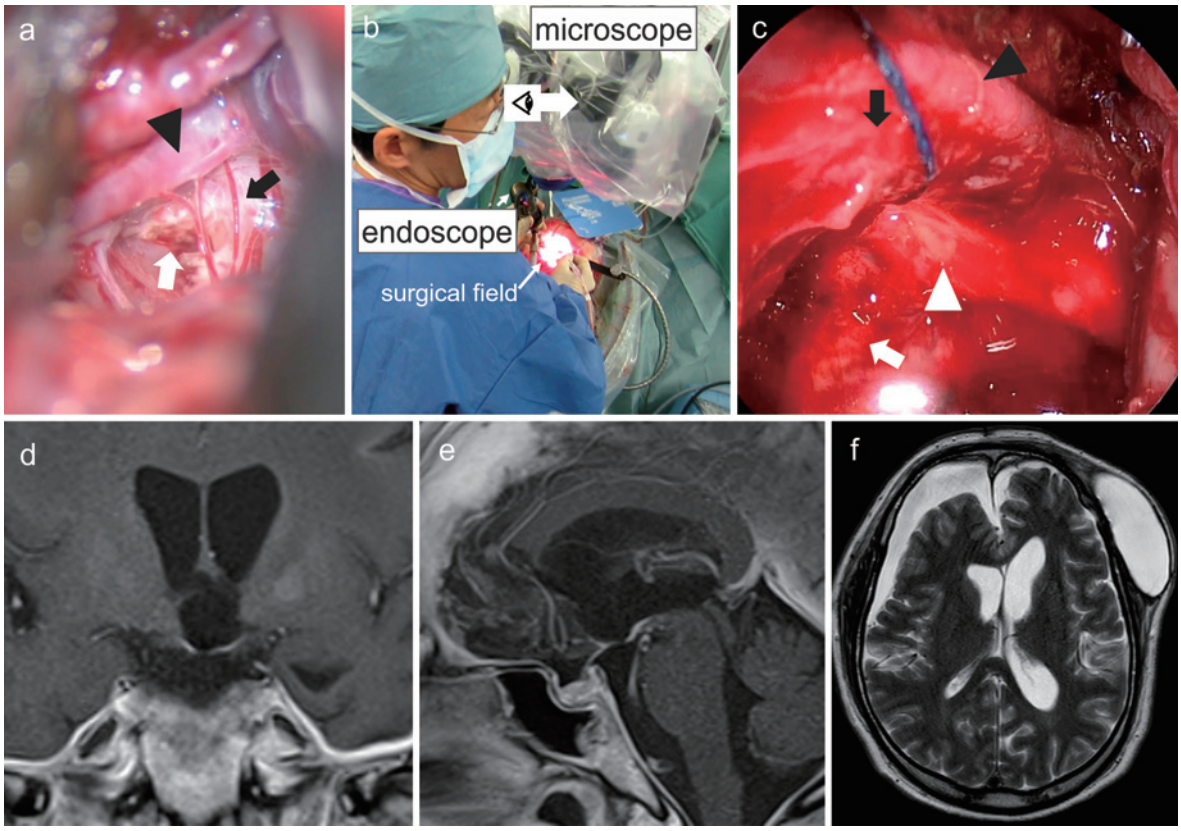

Fig. 2 Intraoperative view from a microscope (a) via the anterior interhemispheric trans-lamina terminalis approach shows the tumor (white arrow) is located in the third ventricle; anterior communicating artery (black arrowhead); and hypothalamic artery (black arrow). The figure shows the positional relations of the microscope, endoscope, and surgical field. The operator watching the endoscopic monitor beside the microscope (b). Intraoperative view from a 45° rigid endoscope (c). The residual tumors (black and white arrows) were detected at the caudal side of the anterior communicating artery (black arrowhead) and chiasm (white arrowhead). Postoperative magnetic resonance imaging revealed gross total resection of the tumor (d, e). Axial enhanced T1-weighted imaging demonstrated postoperative extra-axial cerebrospinal fluid collection (f).

were stabilized. No tumor recurrence was identified 3 years post-tumor resection.

Histopathological examination showed proliferation of the eosinophilic heterocyst in the context of myxoma with chronic inflammatory cell invasion. Immunohistochemically, the tumor was found positive for glial fibrillary acidic protein (GFAP), epithelial membrane antigen (EMA), vimentin, cluster of differentiation (CD) 34, and thyroid transcription factor-1 (TTF-1), with little reactivity for S-100 and negative for Brachyury (Fig. 3). According to these results, the permanent pathological diagnosis was CG, as per the 2021 WHO Classification of Tumors of the Central Nervous System.

The patient and his family have provided consent for the publication of this case report.

Discussion

Commonly, MRI findings of CG demonstrate an enhanced homogenous lesion with Gd in the anterior part of the third ventricle, and perifocal edematous lesions are detectable in the bilateral basal ganglia.[3] The imaging findings of our case have closely resembled these features (homogenously Gd-enhanced lesion in the anterior part of the third ventricle and a high-intensity signal in FLAIR) in the bilateral basal ganglia. The high FLAIR lesion was larger on the right side, considering that the right side of the tumor has severely adhered to the third ventricle.

The histopathological appearance of CG resembles that of chordoma or chordoid meningioma. However, CG commonly lacks bone infiltration and physaliphorous cells. GFAP, CD34, TTF-1, EMA, cytokeratin, vimentin, and S-100 are typically immunopositive in CG, with a low MIB-1 index, which is <5% in almost all CGs.[1,4] The immunostainings in our case were in accordance with the features of the reported cases.

In this case, external CSF collection was observed postsurgery (Fig. 2f). In another study, CSF collection postsurgery for CG and external ventricular shunting were considered a remedy.[5] Open surgery of the ventricle using the anterior interhemispheric approach can cause PECC, and VPS can be an effective treatment.[6] PostVPS placement,

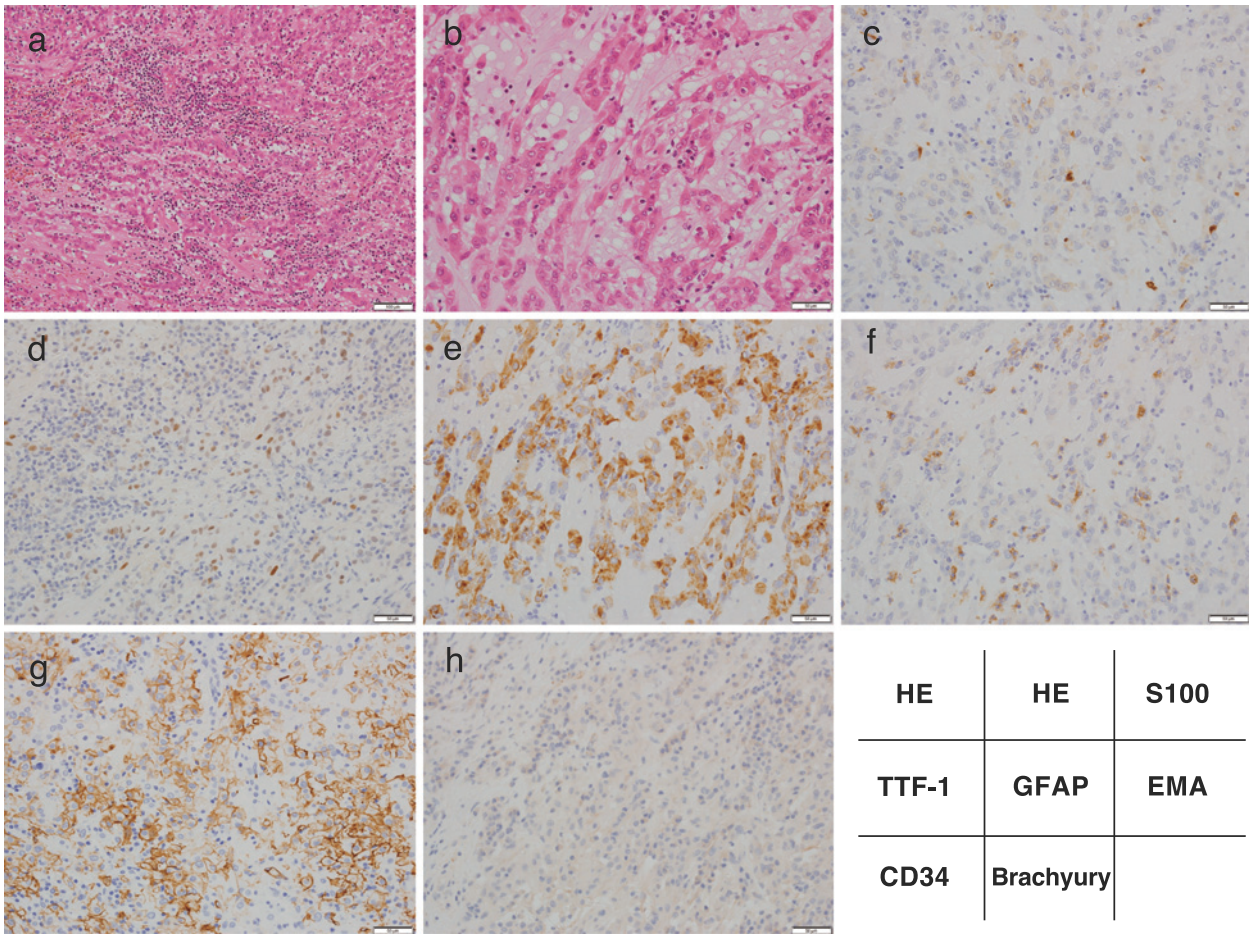

Fig. 3 Photomicrographs of the surgical specimen at the second surgery, hematoxylin and eosin (HE) staining (a, b), S100 (c), thyroid transcription factor-1 (TTF-1) (d), glial fibrillary acidic protein (GFAP) (e), epithelial membrane antigen (EMA) (f), cluster of differentiation 34 (CD34) (g), and Brachyury (h). HE stains revealed regular epithelioid cells with eosinophilic cytoplasm embedded in a mucinous matrix. Chordoid glioma strongly expressed GFAP (e) and CD34 (g), whereas TTF-1 and EMA were partially positive, and Brachyury was negative (h).

hydrocephalus, CSF collection in the left subcutaneous region, and impaired consciousness have remarkably improved. Ampie et al. reported that the 1- and 5-year overall survival rates of patients with CG were 78.2% and 63.7%, respectively.[2] Severe complications of CG include pulmonary thrombosis, which can occur 31 days after surgery in this case. This complication can be caused by hypothalamic dysfunction, long operation duration (in our case, 535 min) due to its difficulty, and postoperative long-term bed rest.[7,8]

Because CG is a slow-growing benign tumor, its appropriate surgical indication is difficult and conflicting. Most patients who present with CG have initial manifestations of headache and hydrocephalus.[9] According to the tumor size, other symptoms may appear. Our patient had already been diagnosed with CG by biopsy in the previous hospital; thus, we observed it until the symptoms of consciousness deterioration and hydrocephalus emerged, considering the characteristics of the tumor and the difficulty of the surgery.

CG, which is hemorrhagic in nature, originates from the laminar terminalis and severely adheres to the surrounding structures.[10] For these reasons, reports on CG argue that several surgical approaches exist for CG of the third ventricle. The anterior interhemispheric (AIH) approach has been identified as the most appropriate for maximum tumor resection of CG while avoiding surgical impairments.[2] However, some studies have indicated that residual CG reccurs postsurgery, which may then require secondary surgery to achieve gross total resection.[1,11,12] Adjuvant radiotherapy is effective for the stabilization of tumor growth,[5,13,14] and a safe and useful strategy, such as planned subtotal resection followed by stereotactic radiosurgery, has been reported.[15] However, how radiotherapy affects CG is yet to be elucidated. To achieve gross total resection, the use of a rigid neuroendoscope, in addition to craniotomy with a microscope via the same corridor, has been found to be useful to detect and resect the residual tumor, even

in the third ventricle that is located deep in the anterior skull base.[16,17] Especially, the high-definition type of endoscope can discriminate the tumor easily from surrounding structures due to its high resolution. Ichikawa et al. reported that hybrid microscopic-endoscopic surgery was useful for eliminating the microscopic blind spot.[17] We routinely used this method for craniopharyngioma resection in the AIH approach. However, it remains difficult to insert the endoscope into the third ventricle via a narrow interhemispheric fissure and small gap in the lamina terminalis without injuring surrounding structures. To avoid this problem, we inserted the endoscope under microscopic view and used the endoscopic monitor beside the microscope to observe and resect the tumor (Fig. 2b). Furthermore, we placed the angled endoscope at the posterior part of the third ventricle, made a working space between the tumor and the angled endoscope, and used dedicated tools for transsphenoidal surgery to resect the tumor during endoscopic procedure.[18,19] These steps are effective for delicate surgical procedures in the deep surgical field under endoscopic view. As a result, the endoscope-assisted trans-lamina terminalis approach was able to safely achieve gross total CG resection at the third ventricle in our case.

Conclusion

In this study, we demonstrated how a CG of the third ventricle has undergone gross total resection via the anterior interhemispheric trans-lamina terminalis approach with a microscope and an endoscope using the same surgical corridor. The endoscope was useful for residual tumor resection at the blind spot of the microscope. In conclusion, the anterior interhemispheric trans-lamina terminalis approach is a promising procedure for patients with CG, with minimal surgical complications and no tumor recurrence.

Conflicts of Interest Disclosure

There are no conflicts of interest to declare.

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