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Case Report

A Case of Chordoid Glioma Complicated by an Anterior Communicating Artery Aneurysm Treated with Single-Stage Surgery Combining an Interhemispheric Approach and Endoscopic Transsphenoidal Surgery

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Chordoid glioma (CG) is an extremely rare low-grade tumor arising from the suprasellar region. Here, we report a case of CG complicated by an anterior communicating artery aneurysm in a 51-year-old man. The patient underwent a single-stage surgery combining endoscopic transsphenoidal surgery (ETSS) and open craniotomy to clip the aneurysm. CG is typically confined to the third ventricle, but in this case the tumor reached the prepontine cistern and clivus. To decrease the risk of aneurysmal rupture during tumor resection, simultaneous clipping of the aneurysm via craniotomy was performed. Tumor resection was primarily conducted using ETSS, with support provided from the craniotomy side. The extent of tumor resection was favorable, and the patient's vision improved after surgery. However, he later developed complications, including diabetes insipidus and cognitive dysfunction. To our knowledge, this is the first report of combined surgery for CG and highlights the potential of ETSS, thereby providing valuable insights for future treatment strategies.
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Introduction

Lesions in the sellar and parasellar regions include pituitary neuroendocrine tumors (PitNETs), Rathke's cleft cysts, craniopharyngiomas, and parasellar meningiomas. These tumors are relatively common and often present with symptoms related to endocrine dysfunction or visual disturbances, which are caused by lesion proximity to important structures, such as the pituitary gland and optic nerves. These conditions are associated with cerebral aneurysm in 2.3%–7.4% of cases[1–4], and concomitant cerebral aneurysm in patients with sellar or parasellar lesions is a significant challenge because the risk of aneurysmal rupture increases during tumor resection, particularly when endoscopic transsphenoidal surgery (ETSS) is used. In such cases, caution is required in managing both the tumor and aneurysm, to prevent catastrophic complications during surgery.

Previous reports have described the use of combined surgeries. In particular, craniotomy and aneurysm clipping were performed simultaneously with ETSS tumor resection for PitNETs[5,6]. The evidence suggests that this approach is feasible for simultaneously managing the conditions. However, despite the increasing number of studies on sellar and parasellar tumors, there is no report describing the use of a similar combined surgical approach for chordoid glioma (CG), a rare low-grade tumor. Here, we report a unique case of CG complicated by an anterior communicating artery aneurysm. The patient underwent a surgical procedure combining craniotomy for aneurysm clipping and ETSS. Further, the litera-

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ture on managing such cases was reviewed, and the lessons learned from our experience were discussed. To our knowledge, this is the first case to describe combined surgery for CG and emphasize the potential of ETSS. The findings offer valuable insights for future treatment of similar cases.

Case Presentation

A 51-year-old man presented for evaluation of a suprasellar tumor. He had type 2 diabetes mellitus, without diabetic retinopathy, and had been experiencing visual impairment for longer than 1 month, for which he was being monitored. During hospitalization at another hospital for diabetic ketoacidosis–induced unconsciousness, a suprasellar tumor was incidentally discovered on magnetic resonance imaging (MRI), and he was then referred to our institution.

Initial Presentation

The patient was alert and oriented. Visual acuity was 0.7 in the right eye and 0.15 in the left eye was (corrected visual acuity, 1.5 in both eyes). Although there were no visual field defects and no other neurological deficits, the patient reported blurred vision.

Blood Testing

Assessment of tumor marker levels

The laboratory results were as follows:

◯Carcinoembryonic antigen level: 2.6 (normal: <5) ng/mL

◯Alpha-fetoprotein level: <2.0 (normal: <8.8) ng/mL

◯Cancer antigen 19-9 level: 11.0 (normal: <37) U/mL

◯Human chorionic gonadotropin level: <2.3 (normal: <5) mIU/mL

Dynamic hormonal testing showed no evidence of impaired secretion of adrenocorticotropic hormone, cortisol, luteinizing hormone, follicle-stimulating hormone, thyroid-stimulating hormone, or prolactin.

Imaging

A non–contrast-enhanced CT scan of the head revealed a 31-mm, slightly hyperdense mass in the suprasellar region with surrounding edema. On MRI, the tumor had a low signal intensity on T1-weighted images, a mixed high-to-low signal intensity on T2-weighted images, and significant heterogeneous enhancement on contrast-enhanced T1-weighted images. Magnetic resonance angiography (MRA) revealed an anterior communicating artery aneurysm anterior to the tumor (**Figure 1a**). Thin-slice coronal and sagittal T2-weighted MRI revealed that the tumor extended from the third ventricle to the prepontine cistern and clivus (**Figure 1b and 1c**). The clival portion of the tumor had minimal contrast enhancement (**Figure 1d–f**). Angiography identified a 3.5-mm anterior communicating artery aneurysm, projecting inferiorly (**Figure 2a and 2b**). Ophthalmologic examination showed no signs of diabetic retinopathy.

Treatment Plan

To relieve pressure on the optic nerves and perform diagnostic biopsy, tumor resection was planned. A combined surgical approach was used because of the tumor location, which made it challenging to access via craniotomy alone, and the risk of aneurysm rupture during resection. The strategy included transnasal endoscopic tumor resection and aneurysm clipping via craniotomy (**Figure 3d**).

Surgery

Craniotomy and intranasal manipulation in ETSS were initiated simultaneously. While dissecting the interhemispheric fissure under a microscope, intranasal manipulation in ETSS was paused. However, tumor resection was performed using both approaches concurrently, with the interhemispheric approach (IHA) playing a supplementary role. The tumor, which was gray and firm, was gradually decompressed and cautiously dissected from the surrounding structures (**Figure 3a and 3b**). Near-total resection was achieved, leaving only a small superficial portion intact. Next, the aneurysm was clipped (**Figure 3 c and 3e**). Intraoperative Doppler ultrasonography confirmed that aneurysmal blood flow had completely stopped. Blood flow in the adjacent vessels was preserved.

Pathological Examination

Hematoxylin and eosin staining revealed clusters and cords of tumor cells with infiltrating lymphocytes and plasma cells. Immunohistochemistry showed diffuse expression of glial fibrillary acidic protein and vimentin, with strong nuclear staining for thyroid transcription factor 1. S-100 staining was partially positive, and the Ki-67 labeling index was <1%. These findings confirmed the diagnosis of CG (**Figure 4**).

Postoperative Course

Brain tumor resection was subtotal, with small ischemic lesions (**Figure 5**), and the patient's vision improved. However, he developed diabetes insipidus, which was

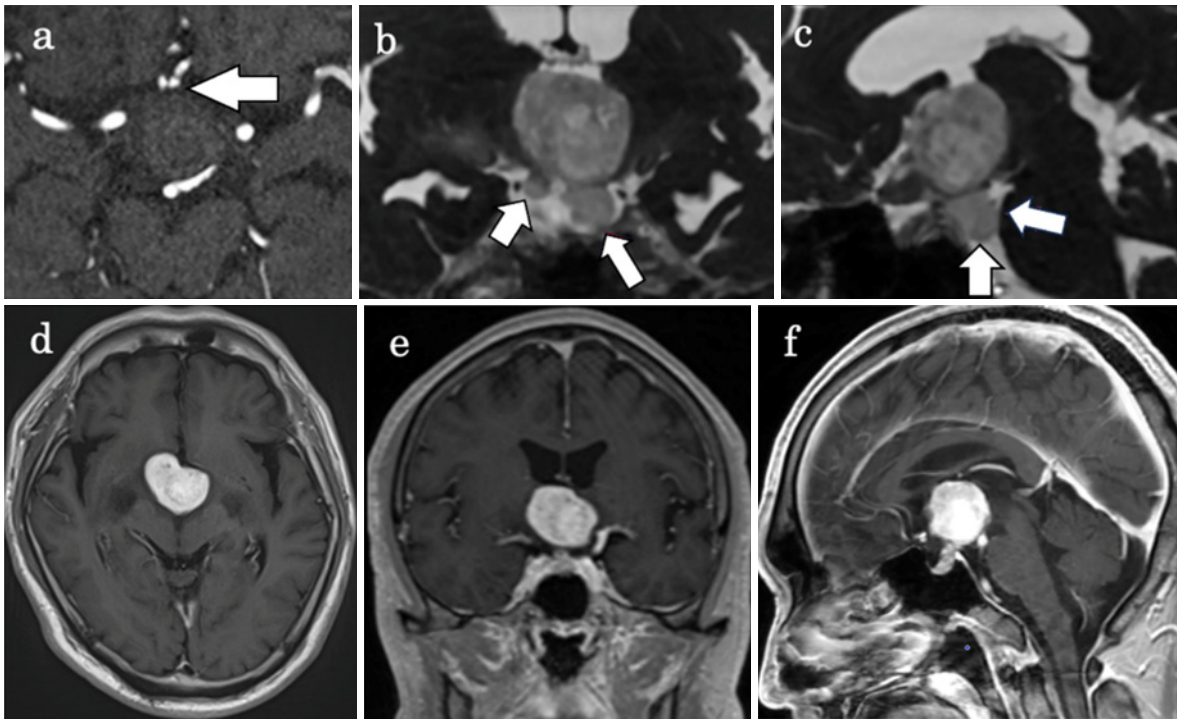

Figure 1 Preoperative findings
A magnetic resonance angiogram (MRA) showing the tumor coexisting with an anterior communicating artery aneurysm, indicated by the arrow (a). T2-weighted magnetic resonance imaging (MRI) revealed that the tumor was located in front of the brainstem, and the tumors, indicated by the arrows, reached the clivus (b, c). Gadolinium-enhanced MRI showed strong homogeneous enhancement of the tumor (d) postcontrast axial MRI, (e) coronal MRI, (f) sagittal MRI.

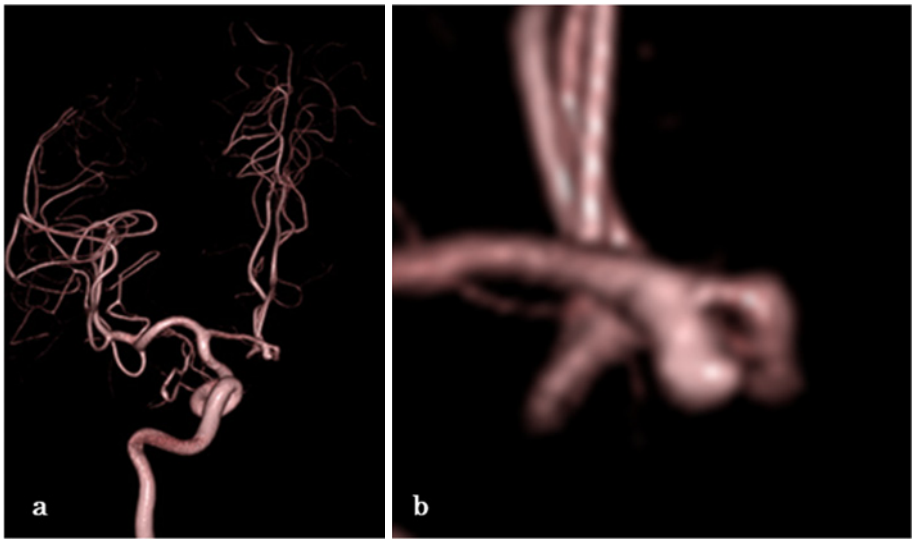

Figure 2 Cerebral angiogram showing an anterior communicating artery aneurysm with a width of 3.5 mm

managed with desmopressin (420 μg/day). Although the patient continued his activities of daily living, he exhibited cognitive deficits, including impaired reasoning, mood changes, memory loss, and attention deficits and was thus transferred to a rehabilitation hospital. Short-term memory impairment persisted after discharge.

Informed Consent

Written informed consent for publication of this case report and accompanying images was obtained from the

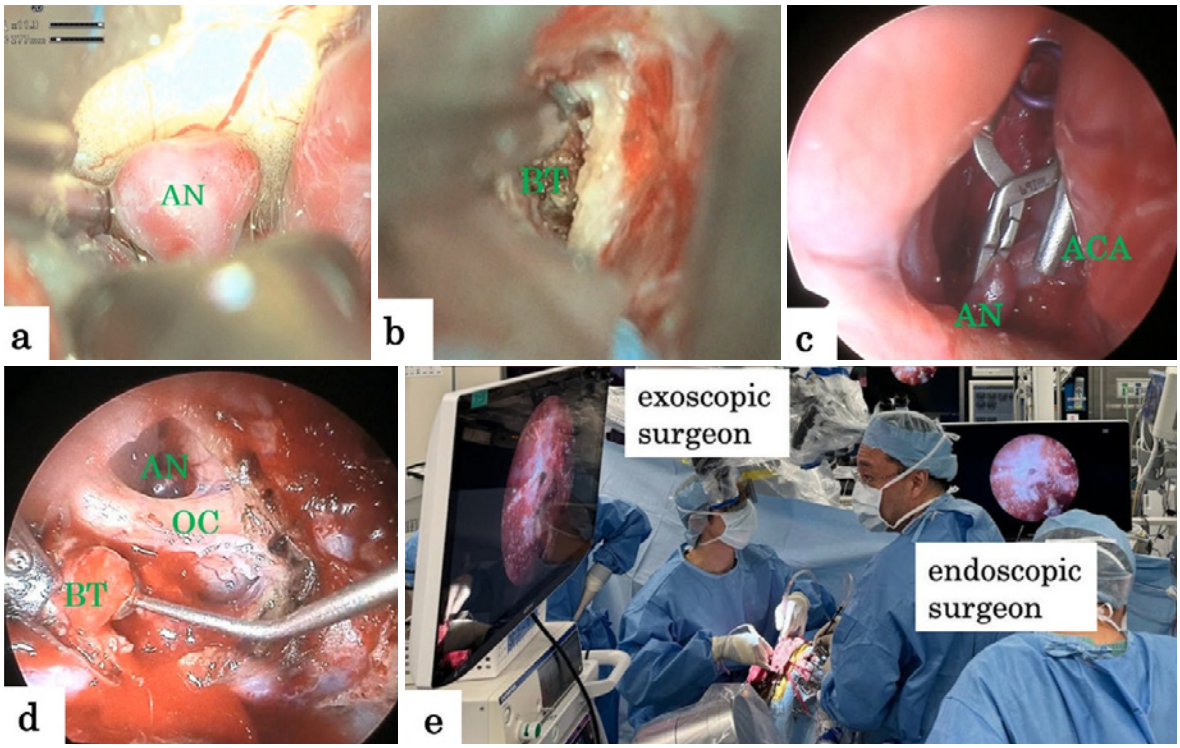

Figure 3 Surgical images
Exoscopic view of the clipping of the aneurysm (a) and tumor resection (b); endoscopic view of the clipping of the aneurysm (c) and tumor resection (d). One operator was observing the exoscopic monitor, and the other was observing the endoscopic monitor (e).

patient.

Discussion

CG is a well-demarcated glioma that primarily arises from the anterior wall of the third ventricle. Accounting for <0.1% of all primary brain tumors, it is an extremely rare tumor[7–9]. In almost all cases of CG, a specific p. D463 H missense mutation of the protein kinase C alpha gene has been identified. Most patients are adults (median age, 45 years), with a female predominance at a ratio of 1:2. CG is generally slow-growing and rarely extends beyond the third ventricle. In the 2021 World Health Organization Classification of Tumors of the Central Nervous System (5th edition), CG is classified as a grade 2 tumor, indicating low malignant potential. However, because of the tumor's proximity to important structures, surgical resection can be challenging, and severe complications may occur postoperatively. A primary aim of treatment is preventing complications such as diabetes insipidus and endocrine dysfunction.

The current surgical approaches for CG include the interhemispheric transcallosal approach, the transcortical transventricular approach, the transsylvian trans–lamina terminalis approach, and ETSS[10–14]. However, there is no preferred approach, because factors such as tumor size, location, and the direction of CG extension must be considered. The interhemispheric transcallosal approach affords easy access to the inferior part of the third ventricle, but access to the undersurface of the optic chiasm and optic nerves is difficult because they cannot be visualized. In a report by Huo et al.[12], more than half of the patients underwent surgery using this approach; however, the transsylvian trans–lamina terminalis approach resulted in significantly better patient outcomes than did other surgical approaches. Nevertheless, accessing the undersurface of the optic chiasm and optic nerves is also challenging with this approach.

ETSS offers a direct view of the undersurface of the optic chiasm and optic nerves. Zeinalizadeh et al.[15] described the first case successfully treated with extended ETSS. Although outcomes have been favorable for some patients with CGs confined to the third ventricle who underwent ETSS, this approach may increase the risk of damaging hypothalamic nuclei in the infundibular area. Moreover, its success may be limited to smaller lesions and those without significant extension away from the midline[12].

There have been frequent reports of patients with con-

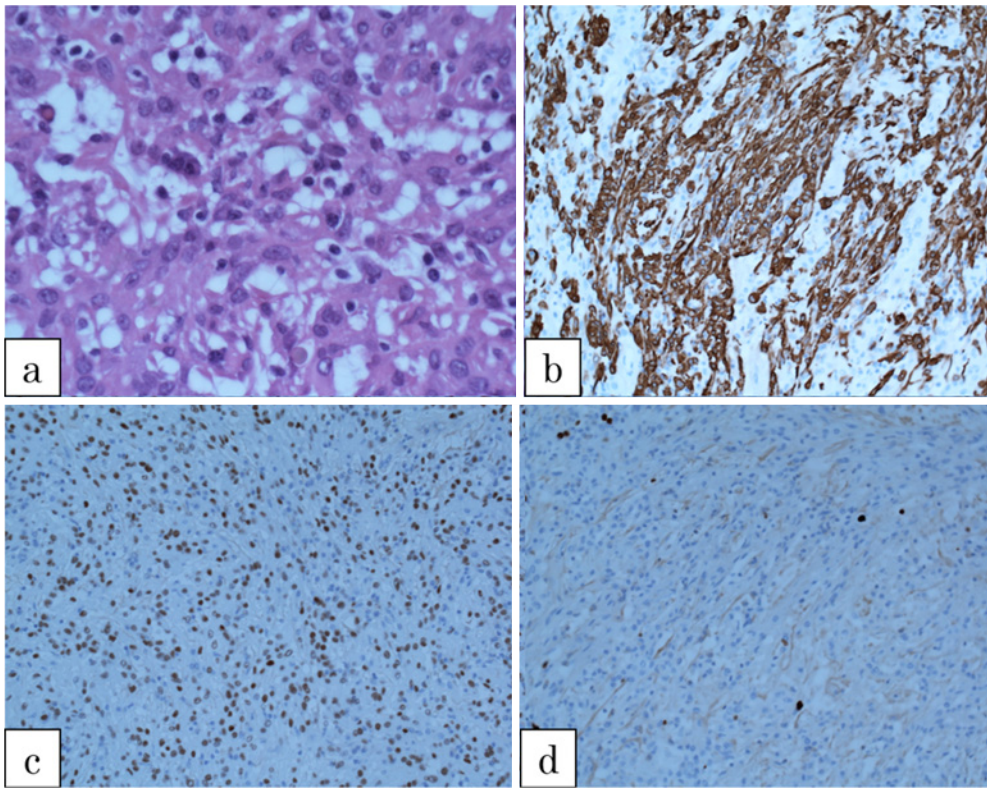

Figure 4 Pathological examination of the chordoid glioma
There were clusters and cords of epithelial cells within a mucinous stain (a) hematoxylin & eosin (×40). Staining of the tumor specimen was positive for GFAP (b), TTF-1 (c), and Ki-67 (d) (b–d, ×40).

comitant cerebral aneurysms and tumors in this region, and the elevated incidence of cerebral aneurysm in patients with PitNETs is well documented. Some studies suggest that PitNETs are associated with rupture of cerebral aneurysms[16], which can cause pituitary apoplexy or severe epistaxis[17]. Cerebral aneurysms may also rupture during ETSS[18,19], which can lead to catastrophic outcomes. Hence, noninvasive imaging such as MRA is recommended preoperatively to confirm the presence of aneurysms[20].

A previous study described the use of combination surgery comprising ETSS for PitNETs and craniotomy for aneurysm clipping[21]. However, to our knowledge, the present report is the first to describe the use of a combined IHA and ETSS for a CG with an anterior communicating artery aneurysm. A unique feature of this case was the relatively close proximity of the aneurysm to the tumor (**Figure 1a**). Considering the risk of aneurysm rupture during tumor resection, the aneurysm should be treated either before or simultaneously with the tumor. In this case, the surgical approach was dictated by the difficulty of achieving complete tumor resection using only IHA or endoscopy, the inability to determine preoperatively whether the tumor was a CG, and the presence of brain edema suggesting the possibility of a rapidly growing tumor (such as craniopharyngioma, chordoid glioma, or another type of glioma). Therefore, simultaneous surgery was performed without preceding aneurysmal treatment. IHA was chosen over the transsylvian approach for aneurysm clipping because it reduced interference between the operators during combined surgery. Furthermore, the tumor extended beyond the third ventricle into the prepontine cistern, reaching the clivus—another distinguishing feature of this case. Accessing the undersurface of the optic chiasm and optic nerves through an open craniotomy would have been challenging, so ETSS was performed simultaneously. This approach allowed direct visualization of the undersurface of these structures. We believe that the combined approach, with mutual assistance from both directions, provided the best access.

If aneurysm clipping was attempted solely via the ETSS, the limited surgical fields and instruments would increase the risk of rupture. Coil embolization was considered, but tumor resection should be scheduled at least 3 months after embolization, when the aneurysm is com-

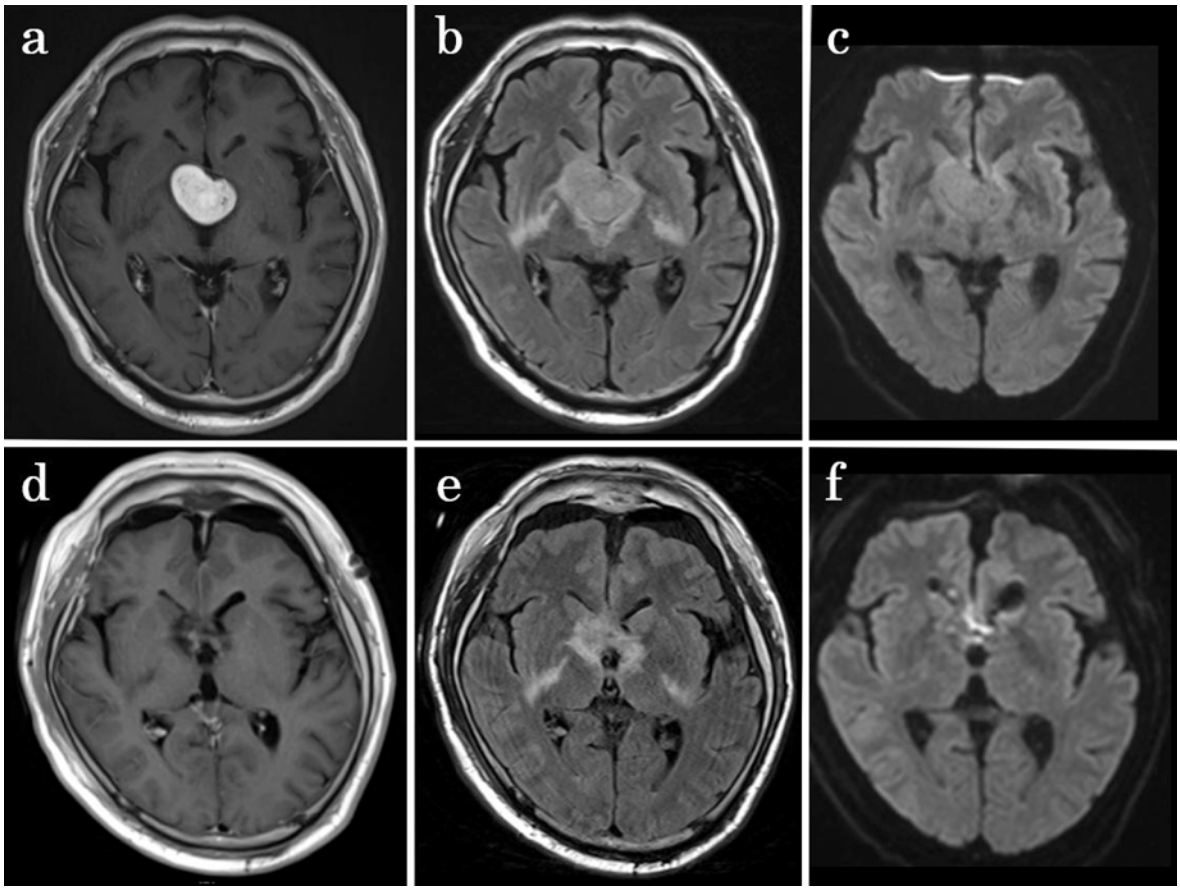

Figure 5 Axial MRI images
(a–c) are pre-operative, and (d–f) were obtained on the day after surgery. (a) and (d) are contrast-enhanced T1-weighted images. (b) and (e) are T2 Fluid Attenuated Inversion Recovery (FLAIR). (c) and (f) are diffusion-weighted images (DWI).

pletely thrombosed. However, progression of the patient's visual symptoms was a concern. Moreover, the use of stents might have required long-term antiplatelet therapy, which increases the risk of perioperative hemorrhage and potential bleeding from residual tumors. Therefore, open craniotomy for aneurysm clipping was selected.

Postoperatively, the patient developed higher brain dysfunction, specifically cognitive impairment, mood changes, memory deficits, and attention disorders, which correspond to the symptoms of hypothalamic syndrome[22]. In particular, dysfunction of the basal forebrain, mamillary body, and fornix contributed to cognitive impairment and memory deficits. These contusions can occur with a transcortical approach or ETSS because of the tumor's proximity to these structures. Jia et al.[23] reported memory loss in 61.54% of patients with intra–third ventricle-type craniopharyngiomas who underwent ETSS. Hypothalamic syndrome can be attributed to damage to the ventromedial and dorsomedial hypothalamus or the fornix, causing endocrine and metabolic disorders, autonomic dysfunction, thermoregulatory issues, eating and sleep disturbances, sexual dysfunction, neuropsychiatric changes, and epilepsy. The transnasal approach was required in order to access the tumor undersurface. Nevertheless, caution must be taken to prevent damage to the infundibulum and hypothalamus. Since this case also involved the development of hypothalamic syndrome, further examination and caution regarding the approach were deemed necessary.

Conclusion

A case of concurrent CG and an anterior communicating artery aneurysm was successfully managed with a combined surgical approach. Although the tumor was nearly totally resected, the patient developed hypothalamic syndrome, indicating that further examination and caution regarding the approach were necessary. Patients with complex tumors and associated aneurysms require an individualized surgical approach.

Author Contributions: Taisei Aoki drafted the manuscript.

Youhei Nounaka assisted in the surgery and revised the manuscript. Fumihiro Matano performed the surgery and revised the manuscript. Koshiro Isayama, Asami Kubota and Tadashi Higuchi assisted in the surgery. Shigeyuki Tahara performed the surgery and supervised the study. All authors read and approved the final manuscript.

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