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Diffusely invasive supratentorial rosette-forming glioneuronal tumor: illustrative case

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BACKGROUND Rosette-forming glioneuronal tumors (RGNTs) are rare tumors composed of mixed glial and neurocytic components. Most lesions are confined to the posterior fossa, especially in the region of the fourth ventricle, in young adults. In few instances, diffuse involvement of the supratentorial region is identified, thereby creating significant challenges in diagnosis, surgical intervention, and prognostication.

OBSERVATIONS The authors present a 23-year-old female with chronic headaches, papilledema, and hydrocephalus who underwent radiographic evaluation revealing obstructive hydrocephalus and diffuse supratentorial enhancing and nonenhancing cystic and nodular lesions. The patient underwent a right frontal craniotomy and septostomy. An exophytic nonenhancing right frontal horn lesion was resected, and an enhancing third-ventricular lesion was biopsied. Final pathology of both of the lesions sampled was consistent with RGNT. Next-generation sequencing demonstrated tumor alterations in the *FGFR-1* and *PIK3CA* genes. Targeted therapy with the FGFR inhibitor erdafitinib demonstrated a partial remission.

LESSONS Diffuse supratentorial spread of RGNT is an extremely rare presentation of an already uncommon pathology. In some cases, gross-total resection may not be feasible. Goals of surgery include acquiring tissue for diagnosis, maximizing safe resection, and treating any associated hydrocephalus. FGFR inhibitors may be of benefit in cases of disease progression.

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KEYWORDS brain tumor; diffuse; invasive; supratentorial; RGNT; rosette-forming glioneuronal tumor

Rosette-forming glioneuronal tumor (RGNT) represents a rare tumor composed of mixed glial and neurocytic components. Initially described in 2002 by Komori et al.,[1] it is now classified by the World Health Organization (WHO) as a grade I neoplasm. With a slightly higher predominance in females, the majority of tumors are confined to the posterior fossa, particularly in the region of the fourth ventricle. Radiological identification of this rare lesion can be challenging, with its features similar to other more well-known neurological diseases, such as neurocysticercosis or dysembryoplastic neuroepithelial tumor (DNET).[2,3] Patients often present with vague symptoms of headaches secondary to hydrocephalus, visual changes, or seizures. In extremely rare instances, diffuse involvement of the supratentorial region is encountered, thereby creating significant challenges in diagnosis, surgical intervention, adjuvant therapy, and determination of prognosis.

Herein, we present the case of a supratentorial, diffusely invasive RGNT with associated hydrocephalus in a young woman. The case represents an atypical presentation of a rare lesion in which gross-total resection (GTR) was not feasible and off-label use of an Fibroblast Growth Factor Receptor (FGFR) inhibitor was used to control disease progression. We also examine the current literature and review the clinical, histological, and surgical/medical management of the disease.

Illustrative Case

A 23-year-old female from Brazil who was positive for coronavirus disease 2019 and had chronic mild headaches presented to the emergency department following a brief loss of consciousness preceded by a prolonged coughing episode. No focal neurological

ABBREVIATIONS CSF = cerebrospinal fluid; CT = computed tomography; DNET = dysembryoplastic neuroepithelial tumor; FDA = Food and Drug Administration; FGFR = Fibroblast Growth Factor Receptor; GTR = gross-total resection; MRI = magnetic resonance imaging; NGS = next-generation sequencing; OLC = oligodendroglioma-like cell; RGNT = rosette-forming glioneuronal tumor; WHO = World Health Organization.

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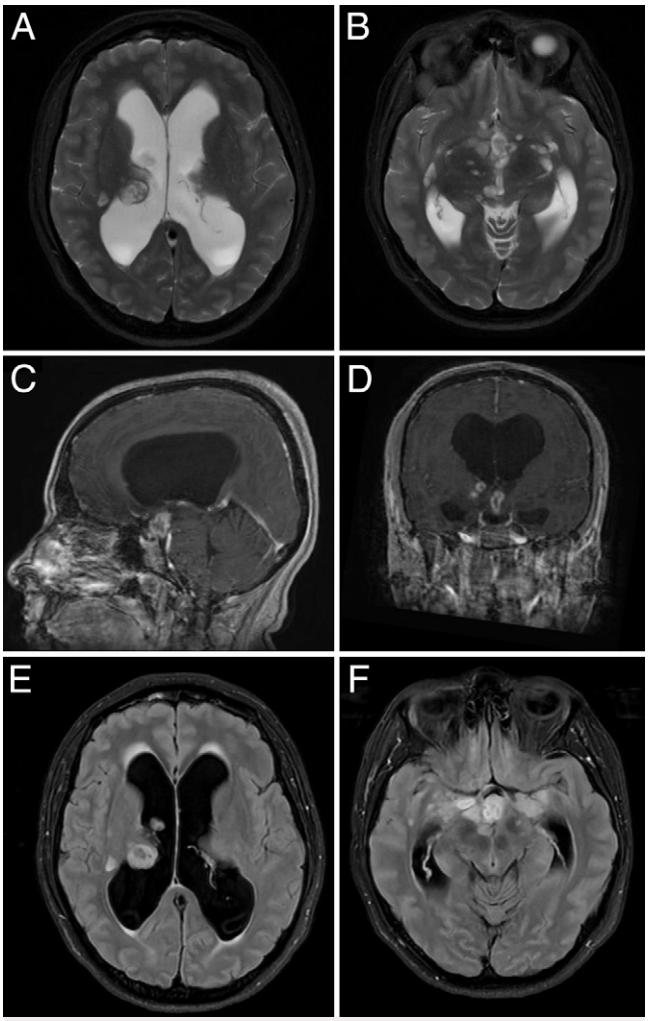

FIG. 1. Axial T2-weighted magnetic resonance imaging (MRI) sequences demonstrate multiple hyperintense subependymal lesions in the wall of the right lateral ventricle (**A**) and in the third ventricle (**B**). Post-contrast T1-weighted MRI shows enhancing lesions involving the hypothalamus and third ventricles on sagittal (**C**) and coronal (**D**) views. Axial T2 fluid-attenuated inversion recovery (FLAIR) sequences (**E–F**) reveal minimal transependymal flow, with FLAIR signal intensity involving the cerebral peduncles and midbrain.

deficits were present; however, ophthalmological exam demonstrated papilledema, confirming the diagnosis of hydrocephalus and elevated intracranial pressure. Computed tomography (CT) of the head demonstrated significant enlargement of the lateral ventricles consistent with obstruction at the level of the third ventricle. Magnetic resonance imaging (MRI) of the brain demonstrated multiple enhancing and nonenhancing, primarily subependymal, cystic and nodular lesions (Fig. 1). Extensive cystic changes were noted in the basal ganglia, thalamus, and midbrain bilaterally. T2-weighted sequences showed minimal transependymal flow surrounding the lateral ventricles and absence of perilesional edema. T1-weighted sequences with gadolinium identified a large, heterogeneously enhancing, third-ventricular lesion likely emanating from the hypothalamus, peripherally enhancing lesions in the right amygdala and temporal periventricular region, as well as multiple homogeneously

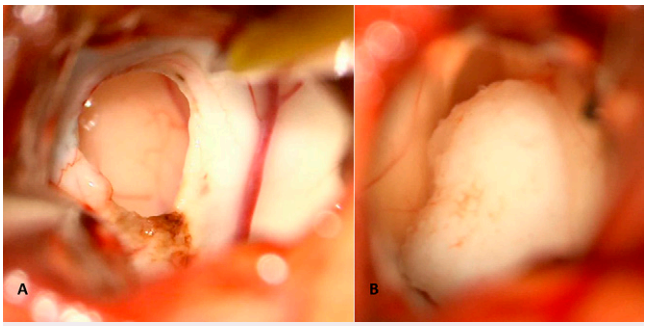

FIG. 2. The septum pellucidum is visualized after resection of the nonenhancing frontal lesion. Septostomy allowing communication between the right and left lateral ventricles (**A**). Right frontal nonenhancing exophytic lesion prior to resection (**B**).

enhancing, subependymal well-circumscribed masses. MRI of the spine was unremarkable. A broad differential diagnosis was developed, including racemic neurocysticercosis, cryptococcal meningitis, toxoplasmosis, and diffuse glioma.

The patient initially underwent a right frontal craniotomy, septostomy, biopsy, and placement of an external ventricular drain (Fig. 2). Prior to disturbing the tumor, cerebrospinal fluid (CSF) was collected for cytology. The exophytic nonenhancing right frontal horn lesion was resected in its entirety (Fig. 2B). The enhancing third-ventricular lesion was identified filling the right foramen of Monro and merging with the right fornix. The appearance was consistent with that of the floor of the third ventricle/hypothalamus. Multiple biopsy specimens were sent for histological examination. The patient remained neurologically intact and consented to the placement of a left frontal ventriculoperitoneal shunt 3 days later. She reported immediate improvement in her headaches and was discharged on postoperative day 1 from shunt placement.

Final pathology demonstrated RGNT. The nonenhancing right frontal lesion demonstrated the biphasic neurocytic and glial cytoarchitecture characteristic of RGNT (Fig. 3), whereas the third-ventricular specimen showed primarily the glial component. CSF cytology was negative. Immunohistochemistry confirmed the glial and neuronal components of the tumor. The Ki-67 labeling index was approximately 0.4%. Microvascular proliferation and necrosis were absent. Next-generation sequencing (NGS) of the tumor showed alterations in the *FGFR-1* and *PIK3CA* genes. Methylation profiling was consistent with RGNT.

Two-month follow-up MRI showed stable disease. However, 4-month MRI demonstrated an increased size of the ring-enhancing lesions in the left thalamus and third ventricle. The decision was made to proceed with targeted therapy with erdafitinib, an FGFR inhibitor that the Food and Drug Administration (FDA) approved for urothelial carcinoma, and to consider radiation in the future with additional disease progression. MRI 3 months after starting the erdafitinib showed a partial response to treatment, with a decrease in the enhancement of multiple lesions (Fig. 4).

Patient Informed Consent

The necessary patient informed consent was obtained in this study.

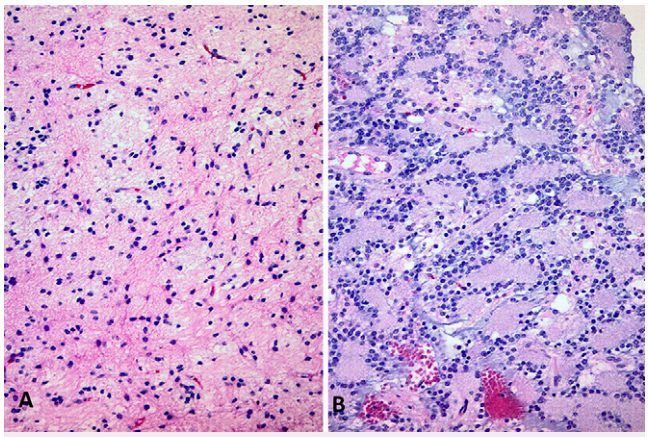

FIG. 3. Hematoxylin-and-eosin stain demonstrating biphasic glial (**A**) and neurocytic (**B**) cytoarchitecture. Ki-67 labeling index of 0.4%. Original magnification ×20.

Discussion

Observations

Clinical Presentation

Rosette-forming glioneuronal tumor is classically encountered in young adults, with a slight predominance in females at a ratio of 1:1.75 (male to female).[1] The mean age at diagnosis is 23.6 years (27.6 for those presenting with supratentorial RGNT), with rare reports of patients in their 80s and as young as 4 years old. Obstructive hydrocephalus is frequent and is a common clinical manifestation.[4,5] Zhang et al.[6] found the rate of hydrocephalus to be 43.6% in a cohort of 41 patients of fourth-ventricular RGNT.

We performed a literature review utilizing the search terms "rosette-forming glioneuronal tumor" in PubMed in August 2023, resulting in 167 articles. Forty-nine articles were excluded after initial title and abstract screening. Articles presenting supratentorial cases were included, and the excluded cases were infratentorial (n = 71) or spinal (n = 9). A total of 49 cases of supratentorial RGNT were included among 38 studies (Table 1).[3,4,7–42] Our analysis suggests that hydrocephalus is a predominant finding in patients with a supratentorial presentation (63% of 49 cases), with a higher rate than reported in infratentorial cases. Patients typically present with headaches and visual changes, nausea or vomiting, gait disturbances, seizures, vertigo, weakness, paresthesias, and dysmetria. Symptoms vary depending on the location of the lesions.[4–6,8] Zhang et al.[6] reported headaches in 68.3% of cases, ataxia in 39%, and seizures in less than 5% of cases. We found that 67.3% (33/49 cases) of supratentorial cases presented with headaches, 16.7% (7/49 cases) presented with ataxia, and 44% (15/49 cases) presented with seizures. Unsurprisingly, seizures were found to be more prevalent in supratentorial than in infratentorial RGNT, whereas ataxia seems to be more prevalent in infratentorial cases. Three of the cases that presented with seizures were patients with congenital epilepsy or a prior history of seizures in adolescence that worsened in the presence of RGNT.

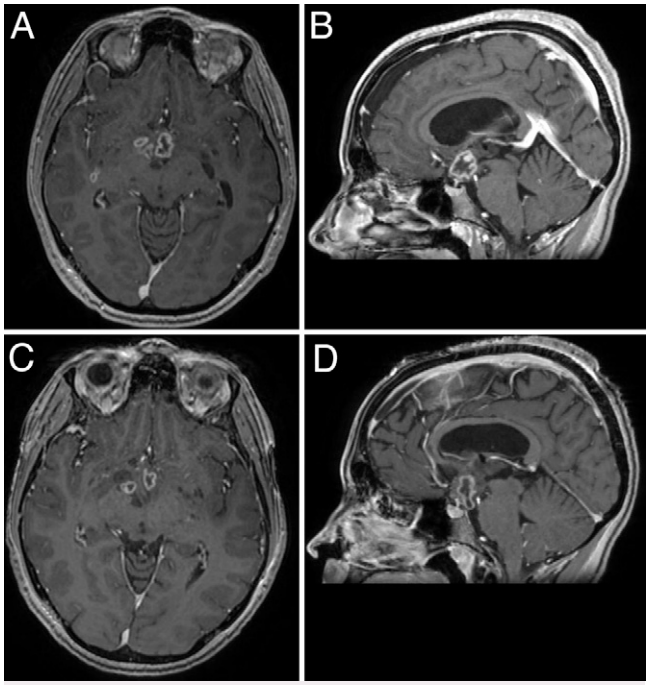

FIG. 4. Postcontrast axial (**A**) and sagittal (**B**) T1-weighted MRI 1 month postoperatively continues to demonstrate multiple enhancing lesions in the third ventricle extending into the optic chiasm and suprasellar cistern. Postcontrast axial (**C**) and sagittal (**D**) images 3 months post–erdafitinib therapy showing decreasing enhancement of the third-ventricular lesion.

Imaging Characteristics

Rosette-forming glioneuronal tumor typically arises in the fourth ventricle and can invade surrounding posterior fossa structures and rarely the spinal cord. Rarely, lesions can involve supratentorial structures including the pineal region, optic chiasm, and septum pellucidum (Table 1).[6] In our analysis, we found that 71% of cases (35/49 cases) were unifocal supratentorial presentations, whereas 29% of cases (14/49 cases) presented with a multifocal supratentorial pattern (invading >1 supratentorial location). Our patient demonstrated diffuse supratentorial disease of the bilateral thalami, crus cerebri, medial temporal lobes, and hypothalamus. Additionally, extensive subependymal spread involving the right lateral and third ventricles was found at the time of initial presentation. The current literature lacks homogeneity on how to properly classify the progression or spread of RGNT and is an area requiring further study.

Rosette-forming glioneuronal tumor can be visualized on both CT and MRI; however, a wide range of radiological features renders image-based diagnosis challenging. Tumors can demonstrate solid and/or cystic components. Focal contrast enhancement can be seen as nodular, linear, ring, or spot-like patterns. On CT, varying amounts of calcification can be seen in up to 25% of tumors. On T1-weighted MRI, the lesions can present as isointense or hypointense. T2-weighted MRI demonstrates predominantly hyperintense lesions.[5]

Interestingly, RGNT was once considered an infratentorial form of DNET, a classically low-grade supratentorial lesion associated with seizures in young adults.[2] Subsequent studies have found a superficial relation but with multiple histological and clinical distinguishing characteristics. An origin from the subependymal plate or the cerebellar internal granule cell layer has been suggested for fourth-ventricular RGNT.[1] Dysembryoplastic neuroepithelial tumor arises from the cortical gray matter, specifically the secondary

TABLE 1. Supratentorial rosette-forming glioneuronal tumor cases reported in the literature

Case No.	Authors & Year	Age (yrs)	Location	Hydrocephalus	Headaches	Ataxia	Seizures	Surgical Management	Recurrence/ Progression
1	Michel et al., 2022[7]	23	Pineal region	–	+	–	–	GTR	–
2		48	Bilat thalamus	+	–	–	+	STR & ETV	–
3		20	Pineal region & 3rd ventricle	+	+	–	–	GTR & VPS	–
4		51	3rd & lat ventricles	+	–	–	+	Biopsy & septostomy & VPS	–
5	Lin et al., 2021[8]	30	Pineal region	+	+	–	–	STR & VPS	–
6		40	Pineal region	+	+	–	–	GTR	NA
7		23	Pineal region	+	+	–	+	GTR & VPS	–
8		42	Pineal region	+	–	+	–	STR & ETV	–
9		17	Pineal region	+	+	–	–	GTR & VPS	–
10		18	Pineal region	+	+	–	–	GTR & ETV	–
11	Uchiyama et al., 2021[9]	9	Temporal lobe	–	+	–	+	STR	+
12	Zhu et al., 2021[3]	22	Thalamus, brainstem, & cerebellum	–	–	–	–	Biopsy	NA
13	Bharadwaj et al., 2020[10]	12	Optic pathway	–	–	–	–	Radiotherapy	–
14	Al Krinawe et al., 2020[11]	7	Septum pellucidum	+	+	–	–	STR	–
15	Mahavadi et al., 2020[12]	41	Lat ventricle	+	+	+	–	STR	–
16	Muhammad et al., 2020[13]	22	Pineal region	–	+	–	–	STR	–
17	Wilson et al., 2020[4]	19	Temporal lobe & 3rd ventricle	+	–	+	+	STR & VPS	NA
18	Yapicier et al., 2020[14]	55	Hippocampus	–	+	–	+	GTR	NA
19	Halfpenny et al., 2019[15]	5	Temporal lobe	–	–	–	+	GTR	+
20	Morassi et al., 2019[16]	24	Lat & 3rd ventricles	–	+	–	–	STR	+
21	Sekar et al., 2019[17]	18	Optic nerve	–	–	–	–	STR	–
22	Singh et al., 2019[18]	20	Corpus callosum	NA	+	–	+	STR	NA
23		29	Suprasellar	NA	+	–	+	GTR	–
24	Yamada et al., 2019[19]	16	Temporal lobe	–	–	–	+	GTR	NA
25	Eye et al., 2017[20]	35	3rd ventricle	+	+	–	–	STR & ETV	–
26	Sumitomo et al., 2017[21]	9	Parietal lobe	–	–	–	+	GTR	NA
27	Tamura et al., 2017[22]	29	Cerebral aqueduct	+	+	–	–	Biopsy & ETV	+
28	Cebula et al., 2016[23]	75	Thalamus	+	+	+	–	STR & ETV	–

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TABLE 1. Supratentorial rosette-forming glioneuronal tumor cases reported in the literature

Case No.	Authors & Year	Age (yrs)	Location	Hydrocephalus	Headaches	Ataxia	Seizures	Surgical Management	Recurrence/ Progression
29	Eastin et al., 2016[24]	33	Thalamus & 3rd ventricle	+	+	–	–	Biopsy & ETV	–
30	Medhi et al., 2016[25]	38	Pineal region	+	+	–	+	STR	–
31		12	Pineal region	+	+	–	–	GTR	–
32	Allinson et al., 2015[26]	13	Lat, 3rd, & 4th ventricles	+	+	–	–	Biopsy & ETV	NA
33	Chen et al., 2015[27]	21	Suprasellar, 3rd & 4th ventricles	+	+	–	–	STR	NA
34	Maiti et al., 2015[28]	12	3rd ventricle	+	+	–	–	GTR	–
35	Yamamoto et al., 2015[29]	8	Hypothalamus	+	+	–	–	STR & radiation	+
36	Matyja et al., 2014[30]	22	Temporal lobe	–	–	–	+	GTR	–
37	Alnaami et al., 2013[31]	57	3rd ventricle	+	+	+	–	ETV & biopsy	–
38		28	3rd ventricle	+	+	–	–	ETV & biopsy	NA
39	Xiong et al., 2013[32]	23	Ant cingulate cortex & frontal lobe	–	–	–	+	GTR	–
40	Kemp et al., 2012[33]	33	Lat ventricle	–	–	–	–	GTR	NA
41	Xiong et al., 2012[34]	38	Lat & 3rd ventricle	+	–	–	–	STR	NA
42	Xu et al., 2012[35]	39	Pineal gland & 3rd ventricle	+	+	–	–	GTR	–
43	Sharma et al., 2011[36]	17	Hypothalamus & 3rd ventricle	+	+	–	–	STR & VPS	–
44	Frydenberg et al., 2010[37]	29	Pineal region	+	+	–	–	GTR	NA
45	Ghosal et al., 2010[38]	22	Pineal region & thalamus	–	+	–	–	STR	NA
46	Solis et al., 2010[39]	16	Pineal region	+	+	–	–	STR	–
47	Scheithauer et al., 2009[40]	23	Optic chiasm	–	+	+	–	STR	NA
48	Wang et al., 2009[41]	16	Lat, 3rd, & 4th ventricles	+	–	–	+	Biopsy & radiation	–
49	Lu et al., 2009[42]	79	3rd ventricle	+	–	+	–	ETV	NA

Ant = anterior; ETV = endoscopic third ventriculostomy; NA = not available; STR = subtotal resection; VPS = ventriculoperitoneal shunt; – = no; + = yes.

germinal layer.[2] Komori et al.[1] showed RGNT to have an expansive growth pattern leading to progressive clinical symptoms due to increased intracranial pressure and hydrocephalus. This clinical scenario is typically not encountered in DNET. Moreover, DNET is classically limited to the cortex, while RGNT can exhibit invasion of the white matter.[1]

The radiological features of RGNT can be variable and often lead to misdiagnosis. Initial misdiagnosis of RGNT as neurocysticercosis, metastasis, DNET, pilocytic astrocytoma, oligodendroglioma, ependymoma, primitive neuroectodermal tumor, and other primary central nervous system tumors has been reported. Rosette-forming glioneuronal tumor confined to the posterior fossa can resemble cystic pilocytic astrocytoma, while supratentorial multinodular lesions can be interpreted as DNETs. Local brain invasion combined with the classic infratentorial location can be helpful in differentiating RGNT radiologically.[5]

The intraparenchymal multicystic appearance and variable enhancement pattern of diffuse supratentorial RGNT also places neurocysticercosis high on the differential. Indeed, our patient demonstrated these findings, and given her previously living in South America, neurocysticercosis remained high on the differential. Although rarely seen in RGNT, intratumoral hemorrhage has been reported and may be distinguishing from other pathologies with similar radiological features, particularly cysticercosis.[3]

Histopathology

Rosette-forming glioneuronal tumor is a WHO grade I neoplasm. Histologically, it has a biphasic appearance consisting of both glial and neurocytic components. The neurocytic elements form uniform neurocytic rosettes or perivascular pseudo-rosettes and may lie in microcystic mucinous areas. The glial elements can appear similar to a pilocytic astrocytoma with Rosenthal fibers and eosinophilic granular bodies. It may also have oligodendroglioma-like cells (OLCs) that form microcysts. Rosette-forming glioneuronal tumor typically shows minimal cellular atypia, without mitotic activity, necrosis, or microvascular proliferation.[5] Its histopathology differs from that of DNET, which classically has a multinodular intracortical growth pattern with bundles of axons lined by OLCs, forming perpendicular columns to the cortical surface.[5,27,43] Rosette-forming glioneuronal tumor can stain positive for glial fibrillary acidic protein (GFAP) from its glial components, as well as positive for synaptophysin from its neurocytic rosette component.[5,43]

Molecular testing or NGS can be helpful in distinguishing tumors with RGNT-like histological features. Lucas et al.[44] performed a comprehensive analysis of low-grade neuroepithelial tumors with *FGFR-1* alterations and concluded that RGNT is unique in its distinct epigenetic mutations in *FGFR-1*, *PIK3CA*, or *PIK3R1* and sometimes *NF-1* or *PTPN11*. This implies that Ras-Raf-MEK-ERK and PI3-kinase-Akt-mTOR signaling pathways play key roles in the pathogenesis of RGNT.

Surgical Management of Invasive Supratentorial RGNT

Rosette-forming glioneuronal tumors confined to the posterior fossa are typically benign and slow-growing and follow an indolent course. Gross-total resection, when possible, especially in tumors confined to the cerebellum, is associated with the best overall long-term outcomes. The low-grade histology lends to long-term control, and low recurrence rates are common with aggressive resection. In cases of tumors predominantly confined to the cerebellum, GTR is often feasible. Unfortunately, many tumors involve the brainstem in the region of the fourth ventricle and Sylvian aqueduct, limiting potential intervention to biopsy or debulking. Given this location, obstructive hydrocephalus is common and may represent a life-threatening emergency. As a result, surgical management must be tailored to individual tumors and the patient presentation.

Diffuse supratentorial spread of the tumor, as was encountered in our patient, although previously reported, is an extremely rare presentation. Gross-total resection, and even aggressive debulking, in these cases is impossible, and goals of surgical management include treating the hydrocephalus and establishing a diagnosis. A systematic approach and prioritization of goals are essential to the successful management of this challenging clinical scenario.

Of greatest importance in this case was the presentation of symptomatic obstructive hydrocephalus. The life-threatening nature of this finding was highlighted by the patient's brief loss of consciousness, which signified a diminishing tolerance of long-standing hydrocephalus. Identification of the third-ventricular lesion as the underlying cause of obstruction was important to developing the surgical plan. In RGNT confined to the brainstem or posterior fossa, endoscopic third ventriculostomy is a potential option for the management of obstruction at the level of the aqueduct or fourth ventricle. Endoscopic third ventriculostomy, in this case, was precluded, as the tumor emanates from the hypothalamus/floor of the third ventricle, fills the third ventricle, and results in obstruction at the level of the bilateral foramina of Monro.

Obstruction of both the foramina of Monro and the inability to debulk/resect the lesion expanding the hypothalamus also necessitates considerations for shunting. The simplest shunt construct entails 1 ventricular catheter, thus necessitating a septostomy. Access to the right frontal horn and third ventricle for biopsy and septostomy can be achieved via a right frontal transcortical approach or interhemispheric approach. The right frontal approach was chosen for multiple reasons. First, the significantly enlarged lateral ventricles provided direct access to the right frontal horn and third-ventricular lesions. Second, a right frontal ventriculostomy was placed to provide drainage in the intensive care unit while awaiting final pathology. Finally, a small linear, sagittally oriented, right frontal incision avoided the left frontal region, which was preserved for placement of a left ventriculoperitoneal shunt.

The second surgical consideration was the identification of a lesion suitable for biopsy and was a critical component of preoperative planning. The heterogeneous nature of the lesions, consisting of enhancing and nonenhancing, cystic and solid, and subependymal and parenchymal lesions, complicated this process, particularly given the broad differential diagnosis. The exophytic right frontal horn lesion was chosen because of its noneloquent location and nonenhancing profile. The third-ventricular lesion was chosen for its enhancement and its accessibility through the foramen of Monro. Importantly, at the time of surgery, it was essential to appreciate that the third-ventricular lesion was expanding the hypothalamus, thereby limiting the amount of tissue that could be safely biopsied. Aggressive resection or even large biopsies of this lesion could have potentially resulted in severe disabling neurological deficits.

Finally, obtaining a CSF sample at the time of craniotomy was a third consideration. Prior to the right frontal corticectomy and disturbing the tumor, a ventriculostomy was placed into the right frontal horn, and CSF was obtained for cytology. CSF was negative for abnormal cells; therefore, it is likely that the diffuse nature of our case was due to intraparenchymal spread rather than CSF dissemination.

Clinical Management and Prognosis

Recurrence following aggressive resection is rare but has been reported and may be associated with CSF dissemination. Observation and serial imaging are appropriate in most cases of postoperative residual disease; however, chemotherapy and radiation have been used as adjuvant treatment in more aggressive RGNT cases. Hockman et al.[43] identified 21 cases of multifocal RGNT of both supratentorial and infratentorial origin, finding that 43% of cases had CSF dissemination and 48% had intraparenchymal spread. Seventeen percent of cases showed progression of disease, and this was only found in cases of CSF dissemination. CSF dissemination of RGNT was found to be associated with more aggressive behavior, requiring early and aggressive management, as opposed to nondisseminated disease. These cases were treated with adjuvant

radiation alone or radiation and chemotherapy, all resulting in death between 6 months and 6 years. They concluded that observation for 12 months or longer is appropriate before consideration of adjuvant therapies.

Erdafitinib is a pan-FGFR inhibitor, approved by the FDA to treat urothelial cancers. Recent reports have investigated the use of erdafitinib in other tumors expressing FGFR mutations, including gliomas.[45] *FGFR-1* alteration is a common mutation associated with RGNT, as was found in our patient. To the best of our knowledge, this is the first report in the literature of erdafitinib used to treat RGNT. The decision to use erdafitinib off-label was made after subsequent radiological follow-up revealed disease progression. Follow-up MRI after erdafitinib therapy showed radiological improvement of the tumor enhancement. Further studies are needed to explore the efficacy of FGFR inhibitors as a possible medical treatment for RGNT in cases in which resection is not feasible.

Lessons

Diffuse spread of RGNT to the supratentorial compartment is an extremely rare pathology and poses significant challenges in diagnosis, surgical management, and planning of adjuvant therapy. The typically young age at which these patients present and the overall paucity of data further complicate the long-term management and establishment of prognosis. Goals of surgery are establishing a diagnosis, maximizing the extent of resection, and treating associated hydrocephalus while preserving neurological function. In cases of progressive and/or diffuse RGNT in which the *FGFR-1* mutation is present, target therapies such as erdafitinib may be of benefit. Additional case reports and management discussions are critical to the literature, as large-scale studies remain unlikely with such a rare tumor.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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