Thalamic rosette-forming glioneuronal tumor stable for more than 11 years after subtotal removal: Case report and review of literature

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Abstract

BACKGROUND: Rosette-forming glioneuronal tumors (RGNTs) are rare and slow-growing (WHO grade 1); they mainly involve the posterior fossa. We here report a rare RGNT originating from the thalamus; after subtotal removal it remained stable for more than 11 years.
CASE PRESENTATION: A woman in her early 20s consulted us due to a 6-month history of memory difficulties, headaches, and blurred vision. Magnetic resonance imaging (MRI) showed obstructive hydrocephalus and an 18-mm non-cystic third ventricular tumor that arose at the right thalamus. It was isointense on T1-wighted images, high-intense on T2-weighted images, and non-enhances T1-weighted images. Through a right trans-ventricular subchoroidal approach we made subtotal resection of the soft tumor, leaving a small remnant attached to the posterior thalamic wall. Histologically, the tumor was composed of an alveolar component that included rosettes surrounding cores of eosinophilic neuropils or small vessels and a solid component resembling pilocytic astrocytoma. The cells composing the rosettes were positive for olig-2, MAP, and synaptophysin. The Ki-67 index was around 1%. Postoperatively her symptoms disappeared, and she commenced her engineering career. MRI performed 11.3 years after the surgery found the absence of recurrence.
CONCLUSION: This RGNT is quite unique because it arose from the thalamus, very rare site from which RGNTs originate, and remained stable for more than 11 years after subtotal resection.

Key words: Rosette-forming glioneuronal tumor, RGNT, thalamus, hydrocephalus, long-time-survival

Introduction

Rosette-forming glioneuronal tumors (RGNTs) are rare WHO grade 1 brain tumors composed of an alveolar component harboring neurocytic rosettes and a solid component resembling pilocytic astrocytoma[1]. They mainly involve the fourth ventricle and cerebellum and were first designated "rosette-forming glioneuronal tumors of the fourth ventricle" by Komori *et al*[2]. RGNTs at the third ventricle[3-5], spinal cord[6, 7], and cerebrum[8] have since been reported. Although RGNTs are thought to be indolent, some were aggressive and showed malignant changes[3, 9- 13]. To understand the nature of these tumors, more cases with prolonged follow-up must be accumulated.

We encountered a 24-year-old woman with a third ventricular RGNT originating from the right thalamus. It had been subtotal removed without adjuvant treatment and the residual tumor remained stable for 11.3 years. We also reviewed thalamic RGNTs reported earlier.

Case report

A 24-year-old female without any family and medical history reported a 6-month history of memory difficulties, headaches, and blurred vision. Fundoscopy revealed bilateral papilledema, but there were no physical findings other than the aforementioned symptoms. Magnetic resonance imaging (MRI) showed a right thalamic, well-circumscribed tumor that protruded into the third ventricle and hydrocephalus (Fig. 1). It was homogeneously isointense on T1- and high-intense on T2-weighted images. No cystic changes, enhancement effect, peritumoral edema, restricted diffusion, or synchronous lesions in the posterior fossa were observed. Magnetic resonance spectroscopy showed that the N-acetyl-aspartate/choline ratio was 0.52. After the injection of fludeoxyglucose or methionine, positron emission tomography (PET) revealed no tracer accumulation in the tumor.

Our preoperative diagnosis was low grade astrocytoma and maximally safe tumor resection was attempted. A right trans-ventricular subchoroidal approach exposed the middle- to the posterior part of the third ventricle between the right thalamus and the right internal cerebral vein. The tumor was totally covered with the third ventricular ependyma and was not involving Sylvian aqua duct (Fig. 2). After endoscopic third ventriculostomy, more than 90% of the grayish-pink and softish tumor was removed under a microscope. There was no major bleeding. A small tumor remnant

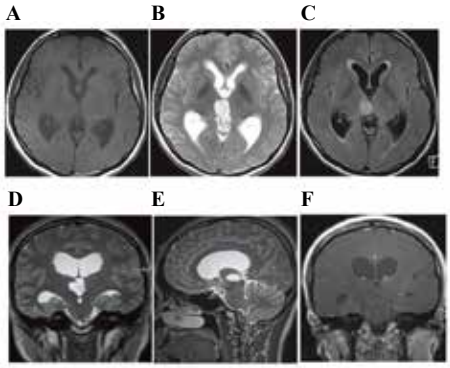

Figure 1. Preoperative magnetic resonance imaging (MRI)

Axial T1-weighted- (A), T2-weighted- (B), and fluid attenuated inversion recovery (FLAIR) scans (C) show hydrocephalus and a tumor in the third ventricle (arrows). The tumor is iso-intense with the contralateral thalamus in (A) and hyperintense in (B) and (C). Coronal and sagittal T2-weighted images (D, E) show the intra-thalamic part of the tumor (arrowheads). It is not gadolinium-enhanced (F).

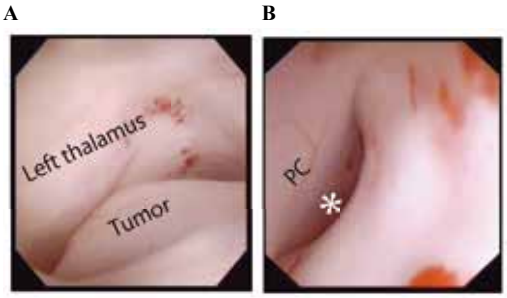

Figure 2. Intraoperative endoscopic exposure of the tumor through the right transventricular subchoroidal approach

A: The tumor is covered with ependyma and protrudes into the third ventricle from the right thalamus. B:The Sylvian aqueduct (*) is not involved by the tumor. PC: posterior commissure

that was attached to the most posterior part of the right thalamic wall was left in situ because manipulation under direct microscopic observation was difficult. Her postoperative recovery was smooth and her symptoms improved gradually. Postoperative MRI scans revealed a small tumor remnant and no hydrocephalus (Fig. 3).

Histologically, the tumor consisted of two components (Fig. 4A). One was alveolar; compact cells with round nuclei formed clusters or rosettes surrounding cores of fine fibrils or tiny vessels (Fig. 4B). An accumulation of mucinous matrix was seen around the rosettes (Fig. 4C). The compact-cell nuclei were positive for olig-2 (Fig. 5A). MAP2 and synaptophysin were frequently positive in the cytoplasm of these compact cells and in the perivascular area and central core of the rosettes (Figs. 5B, 5C). Trapped fibers and scattered larger neurons were positive for neurofilament protein (Fig. 5D). The compact cells and fibrils around rosettes were negative for grail fibrillary acidic protein (GFAP). The Ki-67

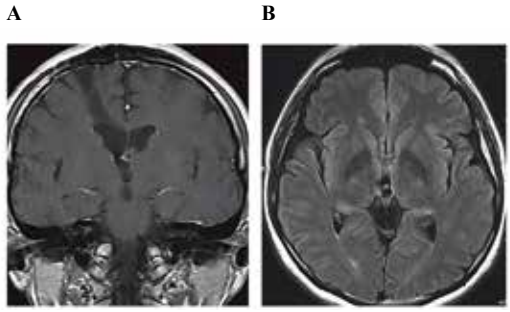

Figure 3. MRI performed 3 months postoperatively

A: Coronal post-gadolinium T1-weighted image shows the surgical corridor (arrowheads) and confirms disappearance of hydrocephalus. B: Axial FLAIR image showing a small residual tumor at the most posterior part of the right thalamic wall (arrow).

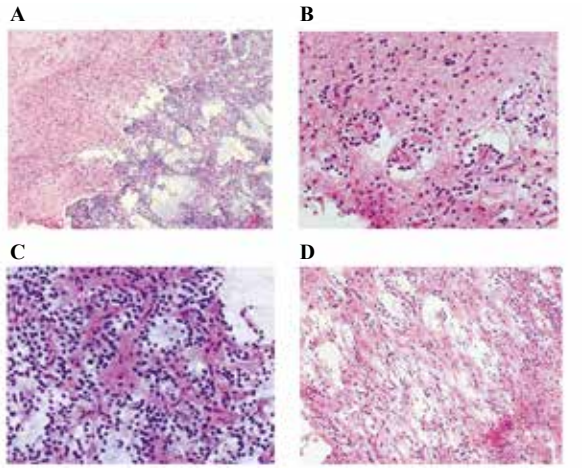

Figure 4. Tumor histology (H&E staining)

The tumor was composed of an alveolar- (A, lower right) and a solid component (A, upper left) (H&E, original magnification × 40). In the alveolar component, an accumulation of compact cells formed rosettes around the eosinophilic core of fine fibrils and microvessels (B, × 200). The accumulation of mucinous matrix around the rosettes and enlargement of the intercellular space produced an alveolar appearance (C, × 200). The solid component (D, × 100) consisted of fascicular arrangements of piloid cells on a microcystic and fibrillary background, resembling pilocytic astrocytoma.

index was around 1%.

The other component was neuronal; long bipolar cells proliferated in fascicular fashion against a fibrillary and microcystic background and resembled pilocytic astrocytoma (Fig. 4D). All of these cells were GFAP-positive (not shown). Many thin-walled vessels and some areas of fine calcification were identified.

Mitosis was absent in both components. These histologic features were compatible with reported findings on RGNTs in the fourth ventricle[1, 2].

The patient's symptom disappeared after surgery and she was able to commence her working career. Follow-up annual- and then bi-annual MRI scans confirmed

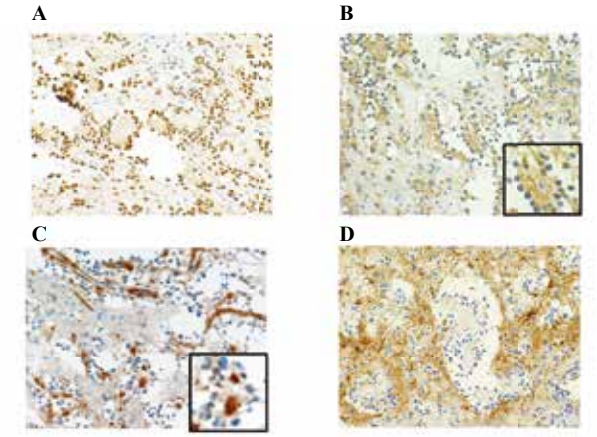

Figure 5. Immunohistochemical stains of the alveolar component

The nuclei of compact cells were clearly stained by olig-2 (A, × 200). MAP2 (B, × 200) and synaptophysin (C, × 200) were expressed in the perivascular area and the central core of the rosettes. They were also positive in scant cytoplasm of the compact cells (insets in B and C). Neurofilament protein immunoreactivity was limited to trapped fibers and scattered larger neurons (D, × 200).

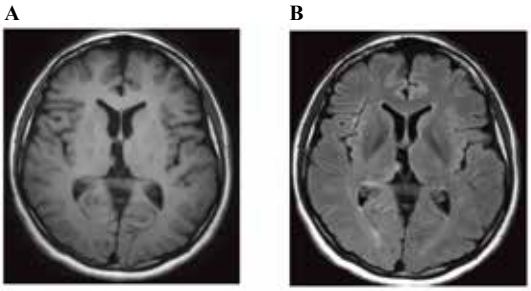

Figure 6. MRI scans obtained 11.3 years after subtotal RGNT removal

Axial T1- (A) and FLAIR (B) images show no regrowth of the residual tumor (arrows) or disseminated lesions.

the absence of tumor recurrence. At 11.3-year follow-up after the operation she was without neurological symptoms and MRI confirmed the stability of the residual tumor (Fig. 6).

Discussion

RGNTs are slow-growing brain tumors that preferentially affect young adults. They tend to arise in the fourth ventricle and cerebellum. Histologically they are composed of two distinct histological components, one is alveolar and the other is solid[1, 2]. It has been hypothesized that the RGNTs originate from pluripotent stem cells in the subependymal plate[14, 15]. In our patient, the RGNTs was also expected to arise from the pluripotent stem cells in the subependymal plate.

RGNTs have been also identified outside the fourth ventricular region, i.e., at the chiasmal-optic nerve[16], the

Table 1. Patients with rosette-forming glioneuronal tumors arising at the thalamus

Author (year)	Age/ Sex	Symptoms	Laterality	Location	Size	Hydro-cephalus	Another lesion	Surgery	Adjuvant treatment	Follow-up	Latest condition
Alnaami I (2013) [21]	57/ M	Headache	Right	Posterior	1 cm*	+	Absent	ETV+Biopsy	No	6 months	Alive without recurrence
Eastin M (2015)[23]	33/ F	Blurred vision, Headache, Tinnitus	Right	Medial	3 cm*	+	Absent	ETV+Biopsy	No	NM	NM
Cebula H (2016)[22]	75/ F	Headache, Unsteady gait, Drowsiness	Left	Posterior (pulvinar)	1.5 cm*	+	Absent	ETV+Biopsy	No	1 year	Alive without recurrence
Present case (2022)	24/ F	Headache, Blurred vision, Memory disturbance	Right	Posterior	1.8 cm	+	Absent	ETV+ Transventricular subtotal removal	No	11.6 years	Alive without recurrence

ETV: endoscopic third ventriculostomy, NM: not mentioned, *: judged from images in the previous case reports

posterior part of the third ventricle[3, 4, 17], the spinal cord[6, 7], the entire ventricular system[18], and the cerebrum[8].

In our patient the tumor occupied the posterior part of the third ventricle and elicited obstructive hydrocephalus. A part of her RGNT was embedded in the right thalamus, presumably the tumor origin. Intraoperatively it was totally covered with ependyma of the third ventricular wall, which confirmed its origin to be the right thalamus.

RGNTs arising in the thalamus are rare. Yang *et al.*[13] who reviewed 188 RGNTs including their own 36 cases found that only 5 (2.7%) manifested thalamic involvement; but the details of these 5 were not described. Our own literature review found 8 RGNTs including ours that involved the thalamus. Of these, 3 were extensions of the fourth ventricle or tectal RGNTs[2, 18, 19]. Another RGNT was detected at autopsy of a patient with multiple neoplastic lesions scattered along the walls of the third ventricles; but there was no clear evidence of thalamic origin[20]. Thus, the remaining four cases were RGNTs truly originated at the thalamus (Table 1) (three females and one male; mean age of 47.3 years) [21-23]. All of these tumors presented with hydrocephalus. Endoscopic third ventriculostomy was performed in all followed by biopsy in three and subtotal removal in our case. None of the patients received adjuvant therapy and three, including ours, were reported to be alive, at 6- and 12 months and 11.3 years after the operation without tumor recurrence.

Preoperative differential diagnoses of tumors arising in the posterior part of third ventricle include high grade gliomas, diffuse astrocytoma, pilocytic astrocytoma, ependymoma, and germ cell tumors. Considering thalamic origin of tumor, tumors from brain parenchyma including glioma should be a primarily preoperative diagnosis in our case.

According to a review by Hsu *et al.* [24], among 47 RGNTs, 40% were solid, 34% were mixed solid and cystic, and 26% were solely cystic on MRI scans. Nearly 90% of the RGNTs reviewed by Hsu et al. [24] and Yang et al.[13] were hypointense on T1- and hyperintense on T2-weighted images. Yang *et al.* reported that among gadolinium-enhanced RGNTs, 3.9% were homogeneous, 44.2% were heterogeneous, 15.5% showed rim- and 11.6% focal enhancement[13]. No enhancement was observed in 24.8% of the tumors. Diffusion-weighted imaging generally sh13owed no evidence of restricted diffusion. Magnetic resonance spectroscopy revealed that the choline level was slightly elevated while NAA was decreased[13].

Because our patient's tumor was T1-isointense, non-enhanced, and solid without cystic formation, our tentative preoperative diagnosis was diffuse astrocytoma (IDH mutated) rather than RGNT, pilocytic astrocytoma, or high grade glioma[17, 25-27]. The absence of tracer accumulation in the tumor on PET scans also denied malignant neoplasms including diffuse midline glioma (H3 K27M mutant) [28].

Genetically, RGNTs were reported to be characterized by highly recurrent genetic alterations affecting both MAPK- and PI3K signaling pathways and by FGFR1 hotspot mutations[29]. Others suggested a relationship between PI3K mutations and RGNT recurrence[4]. At the time we treated this patient, the genetic survey on gliomas currently performed by us using a tailored gene sequencing panel targeting 48 genes including these three genes was not yet available[30]. From now on, genetic information is essential for better understanding of the biological characteristics of RGNTs and for developing effective treatment strategies.

Because RGNTs are generally benign, surgery is thought to be the first-line treatment. We applied a right-sided transcortical, transventricular, subchoroidal approach because its risk of cognitive

function impairment is lower than the transcallosal-, interfornicial-, left-sided transventricular, or transchoroidal approach[31]. We obtained good exposure of the mid- to the posterior half of the third ventricle without splitting major veins or damaging the fornices. Subtotal tumor removal successfully addressed our patient's obstructive hydrocephalus. However, with our approach it was difficult to see the tumor part that was attached to the most posterior portion of the right ventricular wall.

After the first subtotal removal procedure without adjuvant therapy, the residual RGNT in our patient remained stable for 11.3 years. Ours was the 3rd-longest follow-up among approximately 200 RGNTs reported earlier; the average follow-up lasted only 14 - 28 months[6, 13, 32-35]. Therefore, the long-term postoperative course of patients with RGNTs remains unknown. Although most RGNTs are benign, postoperative recurrence[36, 37], malignant transformation[9, 10], and intraventricular dissemination[4, 20, 38, 39] have been reported. In our patient, there were no findings of high-grade gliomas in the glial component or of microvascular proliferation.

Of 124 RGNTs with known prognoses reviewed by Yang et al. [13], 87.9% were stable at the end of a mean follow-up period of 28.0 months; 12.1% exhibited aggressive behavior, 7.3% in-situ progression, and 1.6% dissemination. Three patients (2.4%) died of tumor-related issues. They suggested that in pediatric patients, the presence of a solid tumor and inadequate tumor resection were independent factors predictive of shorter progression-free survival. A review by Wilson et al. [12] revealed that the type of resection affected the interval to RGNT recurrence; in 6 of 62 patients (9.7%) who had undergone gross total removal, the average time to recurrence was 6.17 years; in 3 of 32 patients (9.4%) with subtotal removal it was 2.29 years. As 3 RGNTs recurred 7-, 9-, and 10 years even after gross total resection[4, 10, 40], we should continue to closely monitor the clinical course of our patient.

CONCLUSION

In our patient the RGNT originated from the right thalamus and remained stable for 11.3 years after subtotal resection. More cases with prolonged follow-up must be accumulated and the genetic background of the tumors must be elucidated for the development of effective treatment strategies addressing this rare clinical entity.

COI declaration

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

Ethical approval

This report is a case report and is exempt from the "Ethical Guidelines for Medical Research Involving Human Subjects." Since sufficient consideration has been given to personal information, the ethical review has been waived with the consent of the patient and her family. Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

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亜全摘出後 11 年以上再発なく経過している視床発生のロゼット形成性グリア神経細胞腫瘍：症例報告と文献レビュー

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和文要約

ロゼット形成性グリア神経細胞腫瘍は稀な腫瘍で，緩徐に発育し（WHO grade 1），後頭蓋窩に好発する．我々は，亜全摘出後11年以上再発なく経過する，視床を起源とした稀なロゼット形成性グリア神経細胞腫瘍を経験したため報告する．患者は20歳代前半の女性で，6カ月前からの記憶障害，頭痛，霧視を自覚し当科を受診した．Magnetic resonance imaging (MRI)では閉塞性水頭症，18mmの右視床から生じた非嚢胞性第三脳室腫瘍を認めた．病変は，T1強調像で等信号，T2強調像では高信号を呈し，ガドリニウムによる造影効果は認めなかった．右側からのtrans-ventricular subchoroidal approachにより，視床後壁にごく少量の遺残を残して，柔らかい腫瘍を亜全摘出した．組織学的には，周囲に好酸球性神経細胞性コアや小血管コアを伴うロゼットを含めたalveolar component，および毛様細胞性星細胞腫に類似した充実性成分で構成されていた．ロゼットを構成する細胞は，Olig-2，MAP，synaptophysinが陽性で，Ki-67 indexは1％であった．術後，症状は消失した．術後11年3か月後のMRIでは再発なく経過していた．本症例は，ロゼット形成性グリア神経細胞腫瘍の発生部位としては稀な視床から発生し，亜全摘出後11年経過しても再発なく経過している点において，極めて稀な症例である．