The rosette-forming glioneuronal tumor mimicked cerebral cysticercosis: a case report

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Abstract
Introduction Rosette-forming glioneuronal tumor (RGNT) is a rare variety of slow growing mixed glioneuronal tumor involving primarily fourth ventricular region. This is a comprehensive analysis of a 22-year-old woman with RGNT composed of mainly cystic components. In addition, the case showed multiple lesions located in brain parenchyma which mimicked cerebral cysticercosis. Here, we analyzed this case and listed some characteristics of RGNTs in reported literature which occurring in atypical locations for further understanding it.

Case report A 22-year-old woman presented with a history of transient dizziness, nausea, and vomiting. Magnetic resonance imaging (MRI) showed multiple cystic lesions in brain parenchyma and then the patient was diagnosed with cerebral cysticercosis possibility. Empirical anti-infective therapy in addition to a follow-up post 2 weeks of MRI examination showed the lesions unchanged. Finally, a biopsy of the right cerebellar hemisphere lesions verified RGNT.

Conclusion RGNT is an uncommon tumor classified as grade I glioma by World Health Organization (WHO). The imaging findings of RGNT are not specific especially in atypical areas. RGNT is rare, but we should also consider the possibility in diagnosis and differential diagnosis.

Keywords Rosette-forming glioneuronal tumor · Cerebral cysticercosis · Magnetic resonance imaging · Hemorrhage

Introduction

The rosette-forming glioneuronal tumor (RGNT) was first described by Komori et al. in 2002. As it was initially thought as dysembryoplastic neuroepithelial tumor (DNT) of the cerebellum [1]. In 2007, it was classified as grade I glioma by World Health Organization (WHO). RGNT occurs most often in young women with mean age of onset at 23.57 years [2]. There are few literatures regarding the imaging features and prognosis of RGNT. For most of the literatures on RGNT are case reports. RGNT is most commonly located in the fourth ventricle; however, recent reports demonstrated that RGNT can also occur at sites outside its usual locations. The lesions are mostly comprised of cystic-solid or solid, and the solid components present heterogenous enhancement. Here, we describe a rare case of a 22-year-old woman with RGNT in bilateral cerebellar hemisphere, brain stem, and left thalamus who was misdiagnosed as cerebral cysticercosis before biopsy.

Case report

A 22-year-old woman presented with a history of transient dizziness, nausea, and vomiting. No neurological deficits were apparent; however, on further evaluation, initially with computed tomography (CT) scan, revealed multiple cystic hypo-dense mass lesions in bilateral cerebellar hemisphere, brain stem, and left thalamus with unclear boundary (Fig. 1a–c). Magnetic
Fig. 1  a–c CT findings show the tumors in left thalamus, brain stem, and bilateral cerebellar hemisphere (red arrow). d–l MRI findings. The lesions in bilateral cerebellar hemisphere. d Hyper-intense in T2-weighted image and small nodule-like higher signal (red arrow) along with circled solid component. e Hypo-intense in T1-weighted image. f T2 flair image showed iso-hyper intense. g Hypo-intense in DWI. h Hyper-intense in ADC. i No obvious enhancement majority and minority presented as mild annular enhancement. j–k Perfusion-weighted imaging color map, decreased regional cerebral blood flow, and regional cerebral blood volume. l Minor hemorrhage in the lesions, l SWI showed minor hemorrhage. m–o MRI findings for post 2 weeks. m–o Corresponding T1WI, T2 flair, and T1WI enhancement, no obvious changes compared with before.
Discussion

RGNT is an unusual disease, and it is considered an independent entity of glioma, which is categorized as grade I by World Health Organization (WHO) due to its characteristics of containing both neural and glial components [3]. It is generally considered to be benign, but there have been reports that some can be invasive [4]. The disease was initially thought to only occur in the fourth ventricle, and the typical imaging characteristics are mid-line lesions which appeared in the fourth ventricle and extended to adjacent structures [5]. On MRI, RGNT typical imaging findings are relatively well circumscribed, with both solid and cystic components with T1-hypo-intense and T2-hyper-intense located in or around the fourth ventricle. Gadolinium-based contrast enhancement could show variable or no enhancement, but with increasing reports of the disease, other positions have also been reported, including the pineal region, pons, thalamus, spinal cord, optic chiasm, cerebellar hemisphere, optic pathway, lateral ventricle, septum pellucidum, cerebellar vermis, and even temporal lobe [6–9].

With regards to the English literature through a comprehensive search of Web of Science and PubMed using the search term “the rosette-forming glioneuronal tumor” nearly a decade, more than 100 articles have been published to date. After full text screening, excluding articles that were less relevant to the characteristics of RGNTs, nearly 70 articles were included by December 2020 finally. In general, 101 cases of RGNTs were reported located in the fourth ventricle, while 51 cases were located in atypical site. However, the imaging manifestations of RGNTs occurring outside the fourth ventricle are not specific; so, they are often misdiagnosed. Here, the characteristics of RGNTs located outside the fourth ventricle in 51 published cases were listed (Table 1). The lesions can be solid-cystic, cystic, or simple solid, and generally, the former is the most common. The average age of these published cases

Fig. 2  a–d Pathological pictures of the tumor. a Glioneuronal tumor with glial and neurocytic components. b Neurocytic rosette: small round nuclear tumor cells distributed in a network and a ring-like array of neurocytic tumor cell nuclei around an eosinophilic neuropil core (blue arrow) (hematoxylin-eosin: a×200; b×400). c, d Synaptophysin immunopositivity in the pericapillary area of a perivascular pseudorosette (red arrow) (c×200; d×400). Diffuse positivity for glial fibrillary acidic protein (GFAP) in the glial component of the tumor(e×200)
| Author and year | Lesion | Case number | Age, sex | Location | Contrast enhancement | T1WI | T2WI | Hemorrhage | Management | Recurrence | Number of lesions | Follow-up |
|----------------|--------|-------------|----------|----------|----------------------|------|------|------------|------------|------------|-----------------|-----------|
| Pierre-Aurelien Beuris et al., 2015 | Cystic | 1 | 13/F | Left cerebellar hemisphere | No enhancement | Hypo | Hyper | NA | STR | No | 1 | NA |
| Aaron Halfpenny et al., 2019 | Cystic | 2 | 5/F | Left temporal lobe | Nodular enhancement | Iso/hypo | Hyper | NA | GTR | Yes, 10Y | 1 | 10Y |
| Lian Duan et al., 2017 | Cystic-solid | 3 | 26/F | T9–11 | Heterogeneous enhancement | Hypo | Hyper | NA | GTR | No | 1 | 15M' |
| Lian Duan et al., 2017 | Cystic-solid | 4 | 35/F | C3–7 | Patchy and inhomogeneous | Hypo | Hyper | NA | GTR | No | 1 | 17M' |
| Shuji Hamachi et al., 2019 | Cystic-solid | 5 | 37/F | C2–5 | Slight enhancement | Hypo | Hyper | NA | GTR | No | 1 | 2Y |
| Marc Eastin et al., 2016 | Cystic | 6 | 33/F | Right thalamic, the ventricle | No enhancement | Hypo | Hyper | NA | GTR | No | 2 | 12M' |
| Adrien Collin et al., 2018 | Cystic-solid | 7 | 40/F | C7–8 | Heterogeneous enhancing | Hypo | Hyper | NA | GTR | No | 1 | 6M' |
| Yazeed Al Krinawe et al., 2020 | Solid | 8 | 7/F | Septum pellucidum | No enhancement | Hypo | Hyper | NA | STR | No | 1 | 2Y |
| Bharadwaj, Rishab et al., 2020 | Cystic-solid | 9 | 12/M | The optic pathway | No enhancement | Hypo | Hyper | NA | Biopsy | No | 1 | 6M' |
| Fumine Tanaka et al., 2019 | Cystic-solid | 10 | 18/M | Pons | Partial rim enhancement | Hypo | Hyper | NA | Biopsy | No | 1 | 17Y |
| Emily P Sieg et al., 2016 | Cystic-solid | 11 | 8/F | Right hypothalamus | Ring-like enhancement | Hypo | Hyper | NA | STR | No | 1 | 3Y |
| Arunkumar Sekar et al., 2019 | Cystic-solid | 12 | 16/M | Optic chiasm | Ring-like enhancement | Hypo | Hyper | NA | STR | No | 1 | NA |
| Gautam Bera et al., 2017 | Cystic-solid | 14 | 16/M | Left side of the vermis | Patchy enhancement | Hypo | Hyper | NA | GTR | No | 1 | 1Y |
| Ji Xiong et al., 2012 | Cystic-solid | 15 | 38/M | Septum pellucidum, the bilateral ventricles | Heterogeneous enhancement | Hypo-iso | Mainly hyper | NA | STR | No | 2 | 6M' |
| Kiren S.J. Allinson, 2015 | Mainly cystic | 16 | 33/M | The fourth ventricle, the third and lateral ventricles | Patchy enhancement | Hypo | Hyper | NA | Biopsy | Proximately doubled in size | multiple | 1Y |
| Noriko Sumitomo et al., 2017 | Cystic | 17 | 9/M | The right parietal lobe | No enhancement | Hypo | Hyper | NA | STR | No | 1 | NA |
| Caleb P. Wilson et al., 2020 | Cystic-solid | 18 | 19/M | Left temporal, left gangliocapsular region, bilateral thalamus, tectum, cerebellum. | Slight enhancement | Hypo | Mildly hype | NA | STR | Dramatic expansion, number increasing | multiple | 6Y |
| L. Gao et al., 2018 | Cystic | 19 | 16/M | Cerebellar hemisphere | Slight enhancement | Iso-hypo | Hypo-hyper | NA | GTR | NA | 1 | NA |
| L. Gao et al., 2018 | Cystic | 20 | 29/M | Lateral ventricle | Slight enhancement | Iso-hypo | Hypo-hyper | NA | GTR | NA | 1 | NA |
| Author and year | Lesion            | Case number | Age, sex | Location                                      | Contrast enhancement | T1WI   | T2WI   | Hemorrhage | Management | Recurrence | Number of lesions | Follow-up |
|-----------------|-------------------|-------------|----------|-----------------------------------------------|----------------------|--------|--------|------------|------------|------------|-------------------|-----------|
| L. Gao et al., 2018 | Cystic-solid      | 21          | 23/M     | Cerebellar vermis                             | Heterogeneous        | Hypo-hyper | Hypo-hyper | NA         | STR        | NA         | 1                 | NA        |
| L. Gao et al., 2019 | Cystic            | 22          | 24/M     | Left temporal lobe                            | No enhancement       | Hypo    | Hyper   | NA         | GTR        | No         | 1                 | NA        |
| L. Gao et al., 2019 | Cystic            | 23          | 30/M     | Cerebellar vermis                             | No enhancement       | Hypo    | Hyper   | NA         | GTR        | No         | 1                 | NA        |
| Sajjad Muhammad et al., 2019 | Cystic-solid     | 24          | 22/NA    | Pineal region                                 | Partially and        | Hypo    | Hyper   | NA         | STR        | No         | 1                 | 8W        |
| Ibrahim Alnami et al., 2013 | Solid          | 25          | 57/M     | The posterior third ventricle.                | Heterogeneous        | Iso     | Hyper   | NA         | Biopsy     | No         | 1                 | 6M        |
| Ibrahim Alnami et al., 2013 | Cystic-solid    | 26          | 28/M     | Posterior third ventricle extending into the aqueduct | Nodular enhancement | Hyper   | NA      | Biopsy     | No         | 1          | NA                | 1 Y       |
| Özlem Yapişer et al., 2018 | Cystic           | 27          | 55/F     | Mesial temporal lobe                          | No enhancement       | Iso-hypo| Hyper   | NA         | GTR        | No         | 1                 | NA        |
| H. Cebula et al., 2016 | Cystic-solid     | 28          | 75/F     | Left posterior thalic                          | Heterogeneously      | Hypo-hyper| Hyper   | Na         | STR        | Dead       | Multiple 3               | 3Y        |
| Sonia Garcia Cabezas et al., 2014 | Cystic-solid    | 29          | 24/M     | Both cerebellar hemispheres, the left cerebellopontine angle, spinal cord | Intense and heterogeneous enhancement | Hypo    | Hyper   | NA         | STR        | No         | 1                 | 2Y        |
| Shi-Yun Chen et al., 2016 | Cystic-solid     | 30          | 17/M     | Right basal ganglia                           | Heterogeneous        | Hypo    | Hyper   | NA         | STR        | No         | 1                 | 3Y        |
| Shi-Yun Chen et al., 2017 | Cystic           | 31          | 33/M     | Left parietal lobe                            | No enhancement       | Hypo    | Hyper   | NA         | STR        | No         | 1                 | 3Y        |
| Shi-Yun Chen et al., 2018 | Cystic-solid     | 32          | 21/F     | The third and fourth ventricles and the suprasellar region | Heterogeneous        | Hypo    | Hyper   | NA         | STR        | Dead       | Multiple 3               | 3Y        |
| Yasutaka Fushimi et al., 2011 | Cystic-solid    | 33          | 28/F     | The cerebellar vermis                         | No enhancement       | Hypo    | Hyper   | NA         | STR        | No         | 1                 | 2Y        |
| David Cachia et al., 2014 | Cystic           | 34          | 36/F     | Right frontal lobe, right midbrain tectum, and cerebellar vermis | No enhancement       | Hypo    | Hyper   | NA         | STR        | No         | Multiple 7               | 7M        |
| Philip George Eye et al., 2017 | Cystic-solid     | 35          | 35/M     | The third ventricle                           | No enhancement       | Iso     | Hyper   | NA         | STR        | NA         | 1                 | NA        |
| Orestes E. Solis et al., 2011 | Cystic-solid     | 36          | 16/F     | The pineal gland region                       | No enhancement       | Iso-hypo| Hyper   | NA         | STR        | No         | 1                 | 2M        |
| Ji Xiong et al., 2013 | Cystic           | 37          | 23/M     | Left frontal lobe                             | No enhancement       | Hypo    | Hyper   | NA         | GTR        | No         | 1                 | 8M        |
| Gorky Medhi et al., 2015 | Cystic-solid     | 38          | 32/M     | Midline posterior fossa, vermis and cerebellar hemispheres | Heterogeneous        | Hypo-hyper| Hyper   | Yes        | STR        | No         | Multiple 11               | 11M       |
| Gorky Medhi et al., 2015 | Cystic-solid     | 39          | 38/F     | Pineal region                                 | Heterogeneous        | Hypo-hyper| Hyper   | Yes        | STR        | Residual lesions, stable | 3Y        |
| Cystic-solid | 40 | 24/M | No enhancement | Iso-hypo | YES       | STR       | NA | Multiple | NA |
| Author and year                  | Lesion    | Case number | Age, sex | Location                                                                 | Contrast enhancement                                                                 | T1WI | T2WI | Hemorrhage | Management | Recurrence | Number of lesions | Follow-up |
|---------------------------------|-----------|-------------|----------|---------------------------------------------------------------------------|---------------------------------------------------------------------------------------|------|------|------------|------------|------------|------------------|-----------|
| Gorky Medhi et al., 2015        | Cystic    | 41          | 12/M     | Cerebellar hemispheres, vermis, midbrain, pons, medulla                   | Heterogeneously hyper                                                                  | Hypo | Hyper | NO          | GTR         | Residual lesions | 1                | 3M'       |
| Gorky Medhi et al., 2015        | Cystic    | 42          | 40/F     | Pineal region                                                             | No enhancement                                                                       | Hypo | Hyper | NO          | GTR         | Residual lesions | 1                | NA        |
| S. Kemp et al., 2012            | Cystic    | 43          | 33/M     | Left lateral ventricle                                                    | No enhancement                                                                       | Hypo | Hyper | NA          | GTR         | NA         | 1                | NA        |
| Ewa Matyja et al., 2014         | Cystic    | 44          | 22/M     | The left temporal lobe                                                     | No enhancement                                                                       | Hypo | Hyper | NA          | GTR         | No         | 1                | 3.6Y      |
| Junqing Xu et al., 2012         | Cystic    | 45          | 39/M     | Pineal gland, the third ventricle                                         | Iso-hyper                                                                            | Hypo | Hyper | NA          | GTR         | No         | Multiple          | 42M'      |
| Benjamin Thurston et al., 2012  | Cystic    | 46          | 8/F      | Left superior cerebellar peduncle                                        | Heterogeneous enhancement                                                              | Hypo | Hyper | NA          | GTR         | Yes, 9M     | 1                | 9M'       |
| Anil K. Mahavadi et al., 2020   | Cystic    | 47          | 41/M     | The third ventricle                                                       | Heterogeneous enhancement                                                              | Mainly hypo | Mainly hypo | NA          | STR         | Residual lesions, stable | 1          | 6M'       |
| Pankaj Sharma et al., 2011      | Cystic    | 48          | 16/F     | The tectal region of the midbrain                                        | No enhancement                                                                       | Hypo | Hyper | NA          | Biopsy     | Stable     | 1                | 6M'       |
| Pankaj Sharma et al., 2012      | Cystic    | 49          | 17/M     | Suprasellar and interpudendial cistern, the third ventricle              | Peripheral enhancement                                                                 | Mainly hypo | Mainly hypo | YES         | Biopsy     | Stable     | Multiple          | NA        |
| Seiji Yamada et al., 2019       | Cystic    | 50          | 16/F     | Right temporal lobe                                                       | Multinodular enhancement                                                               | Hypo | Hyper | NA          | GTR         | NA         | 1                | NA        |
| Tannoy Kumar Maiti et al., 2014 | Solid     | 51          | 2/M      | The posterior third ventricle                                             | No enhancement                                                                       | Hypo | Hyper | NA          | STR         | Dead       | 1                | 13M'      |
| Tannoy Kumar Maiti et al., 2015 | Cystic    | 52          | 12/M     | The posterior third ventricle                                             | Mild contrast enhancement                                                              | Hypo | Hyper | NA          | GTR         | No         | 1                | 9M'       |

*M* man, *F* female, NA not available, GTR gross total resection, STR subtotal resection, iso iso-intensity, hypo hypo-intensity, hyper hyper-intensity, *Y* year, *M’* month
is 38 years old. Hemorrhage is rare in RGNTs, and only six cases presented positive for bleeding. Management of RGNTs has been accordant with the literatures. Surgery remains the primary treatment option, with gross total resection (GTR) recommended and subtotal resection (STR) as alternatives. The prognosis of RGNT is generally good, and recurrence is uncommon with a total of 4 cases recrudesced of the 51 cases. However, two patients died of these presented cases. Most of the tumors were single lesions with only 10 cases showed multiple lesions.

In summary, the misdiagnosis in the above case reflects how RGNT is under-emphasized and poorly researched. Thus, based on the analysis of the present case and limited data available from review of literature, we propose that the following two aspects may have contributed significantly in misdiagnosing RGNT. Firstly, the case we highlighted occurred in the brain parenchyma, while more than 69.7% of previously reported RGNTs involve the fourth ventricle [3]. And multiple lesions involving the bilateral cerebellar hemisphere, brain stem, and left thalamus at the same time are rarely reported. Secondly, the imaging findings of this case overlapped with cerebral cysticercosis. As we all know, cerebral cysticercosis is the most common parasitic disease of the central nervous system (CNS). The imaging manifestations of the parenchymal active phase are multiple cystic lesions. The enhancement is not obvious and the hypo-perfusion on perfusion imaging. For the above reasons, the tumors were misdiagnosed as cerebral cysticercosis deservedly.

Admittedly, there are distinct radiological signs which highlight uncertainties regarding the previous diagnosis: most notably include tiny spot-like hypo-intense in bilateral cerebellar hemisphere on the susceptibility weighted imaging (SWI) (Fig. 1I). After excluding tiny calcifications on CT (Fig. 1c), it can be assumed that there is minor hemorrhage within the lesions. Hemorrhage is almost invisible in cerebral cysticercosis, although there were also few reports of hemorrhages in RGNT. Two possible reasons may account for the latter. One being that the SWI sequence is rarely a routine sequence, and minor hemorrhages in many reported cases may go undetected because they are difficult to show on other sequences in MRI. On the other hand, it is generally assumed that RGNT is a benign tumor, microvascular proliferation is rare; therefore, the hemorrhages are infrequent as well. The evidence between microvascular endothelial proliferation and hemorrhages has been documented in RGNT [10, 11]. L. Gao et al. reported several cases of RGNTs with intratumoral hemorrhage in 2017. They summarized that intratumoral hemorrhage was one of the additional indications to the diagnosis of RGNT. Other indications included “green bell pepper sign,” CSF dissemination, and multiple satellite lesions. Medhi et al. also summed up that hemorrhage and CSF dissemination may be the characteristics of RGNT through the summary of 7 cases [12]. In our case, intratumoral hemorrhage and multiple satellite lesions are consistent with their conclusions. Therefore, the above signs may aid in diagnosing RGNTs. However, whether this microvascular proliferation and intratumoral hemorrhage are related to prognosis needs further research.

**Conclusion**

RGNT is an uncommon low-grade neuroglial tumor which is generally considered benign with slightly longer course [2]. Headache is the most recorded common symptom. Histopathologically, RGNT consists of two components: a neurocytic component that forms rosettes, and an astrocytic component that resembles a pilocytic astrocytoma [13]. Through the case we reported, we have discovered that RGNT can be multiple cystic lesions, and the brain parenchyma can be the major affected areas. The intratumoral hemorrhage shown by SWI sequence may have some significance for our diagnosis of it. In conclusion, RGNT often presents significant diagnostic dilemma, and hence, further knowledge of this tumor is essential as they are relatively slow growing and exhibit benign histological characteristics.

**Declarations**

**Conflict of interest** The authors declare no competing interests.

**Ethical approval** This article does not contain any studies with humans.

**Informed consent** We would like to state that the informed consent has been obtained from the patient.

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