



Rosette-Forming Glioneuronal Tumor at Septum Pellucidum: Insights Gained from a Common Tumor at Rare Location

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Abstract

The rosette-forming glioneuronal tumor (RGNT) is an uncommon entity and carries a special character because of its mixed glial and neuronal composition in the histomorphological appearance. These lesions have a benign character and carry a good outcome if undergoes gross total resection. Over the past 15 years, there have been a significant change in their nomenclature depending upon the location to histological composition. Herein, we report an interesting case of a 26-year-old lady who was diagnosed to have the lesion at the septum pellucidum with significant symptoms in the form of headache and seizure episodes. A gross total resection was achieved and she made an uneventful recovery. We discuss the literature on the incidence, location, and histological characteristics of the RGNT in various age groups.

Keywords

- ▶ glioneuronal tumor
- ▶ septum pellucidum
- ▶ rosette-forming

Introduction

The rosette-forming glioneuronal tumor (RGNT) is a rare low-grade tumor consisting of glial and neuronal cells at varying stages of differentiation.¹ In the 2007 World Health Organization (WHO) classification, they were called “rosette-forming glioneuronal tumors of the fourth ventricle.”² In the 2016 edition of the WHO classification of central nervous system (CNS) tumors, these tumors were renamed as “rosette-forming glioneuronal tumors” histologically classified as WHO grade I under the category of “neuronal and mixed neuronal-glial tumors” from the earlier entity “rosette-forming glioneuronal tumors of the fourth ventricle” because of their occurrence in optic chiasm, pineal region, septum pellucidum, as well as spinal cord in addition to fourth ventricular cavity.^{3,4}

Herein, the authors present an illustrative case with brief literature review to highlight the caveats associated with very uncommon location of this tumor.

Case Report

A 26-year-old lady presented with complaints of gradually progressive headache, multiple seizure episodes, and weakness over right side of the body for the last 1 month. Her neurological examination was within normal limit, except for bilateral papilledema on fundoscopy. Magnetic resonance imaging (MRI) with gadolinium contrast showed iso- to hypointense on T1 and hyperintense on T2 weighted image with a nonenhancing 1.5 × 1.5 × 0.5 cm predominantly cystic mass attached to the septum pellucidum (▶ Fig. 1). The tumor

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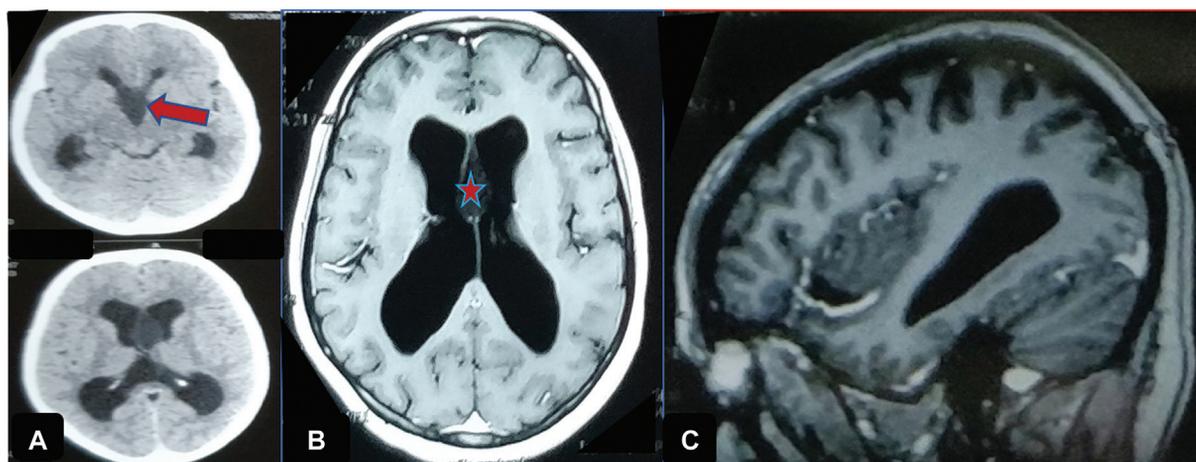


Fig. 1 (A–C) CT and MRI findings.

decompression was performed by interhemispheric transcortical approach. Intraoperatively, tumor was seen to be arising from septum pellucidum, extending into lateral ventricle (Right > Left) and was grayish white, soft, suckable, and moderately vascular. Endoscope was used as an assisting tool to achieve gross total excision. Histopathology was suggestive of a biphasic tumor and immunohistochemistry was positive for Synaptophysin and glial fibrillary acidic protein (► **Fig. 2**). MIB labeling index was low (<3%). Except for single episode of generalized tonic clonic seizure on second day of surgery, her postoperative course was uneventful. Subsequent radiology was suggestive of reduction in ventricular size and no residual lesion. After 2 years of follow-up period, she is asymptomatic and doing well.

Discussion

The RGNT was considered as a benign, slow-growing tumor of the fourth ventricular region about two decades back.⁵ In 2002, Komori et al characterized the clinical, radiological, and histopathological features of RGNTs in 11 cases, and they were the first to propose that these lesions cater a distinct clinicopathological entity of mixed glioneuronal tumors.⁶ Recent case reports have indicated that RGNTs could also originate from the spinal cord, third ventricle, and supratentorial brain parenchyma. In a recent study by Yang et al, tumor preponderance was noticed mostly in cerebellum (34.2%) and fourth ventricle (26.3%), followed by supratentorial ventricular system (13.2%), spinal cord and temporal lobe (10.5% each), thalamus and brain stem (7.9% each), frontal lobe and pineal region (5.3% each), and suprasellar region and basal ganglia (2.6% each).⁷

► **Table 1** summarizes the cases of RGNT at uncommon locations (other than fourth ventricular cavity) reported in English literature. The MRI appearance can be divided into cystic, cystic-solid, and solid type, representing 35%, 18%, and 47%, respectively. The cystic components may suggest a relatively benign nature. In most of the RGNT cases, the solid portion showed homogeneous hypointensity on T1WI and homogeneous hyperintensity on T2WI, while contrast

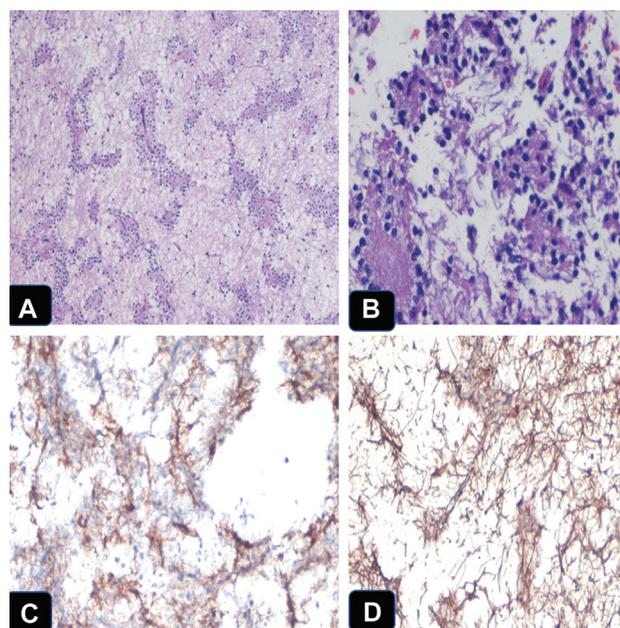


Fig. 2 (A–D) H&E and IHC images.

enhancement was variable with regard to the patterns and degrees of enhancement.

Safe surgical resection of tumor is considered as the gold standard of treatment with limited role of adjuvant chemoradiotherapy only in recurrent cases.

The absence of nuclear atypia, mitotic activities, and necrosis with a low proliferation index in the vast majority of RGNTs indicated a benign biological behavior.⁷ The differential diagnosis of the lesion could be glioma (low, intermediate, or even high grade), germ cell tumors, dermoids, colloid cyst, and neurocytoma. The recent updates of WHO classification of brain tumors have labeled RGNT as “myxoid glioneuronal tumor” as the revised nomenclature for this entity with dual character. Septal nuclei, septum pellucidum, corpus callosum, and periventricular white matter are the preferred locations of occurrence. The available literature suggests a good outcome after tumor decompression and significant resolution of preoperative

Table 1 Summarizing the cases of rosette-forming glioneuronal tumor at uncommon locations reported in English literature

| Author | Age/Sex | Location | Radiological features | Treatment | Outcome (months) |
|--|---------|--|---|-----------|------------------|
| Komori et al ⁶ (2002) | 12/F | Pineal region, aqueduct, tectum | T1 hypo, T2 hyper focal enhancing predominantly cystic lesion | STR | Stable (2) |
| Jacques et al ¹⁰ (2006) | 33/F | Pineal region, left cerebellar peduncles | Multiple cystic lesions with patchy enhancement | GTR | Recurrence (120) |
| Scheithauer et al ¹¹ (2009) | 23/M | Optic chiasm | T1 iso, T2 hyperintense heterogeneously enhancing lesion | STR | Unknown |
| Anan et al ¹² (2009) | 44/F | Cervical-upper thoracic spinal cords | T1 iso/hypo, T2 hyperintense ring enhancing lesion | GTR | Stable (14) |
| Frydenberg et al ¹³ (2010) | 29/M | Pineal region, aqueduct | T1 hypo, T2 hyper focal enhancing cystic lesion | GTR | Unknown |
| Solis et al ¹⁴ (2010) | 16/F | Pineal gland, third ventricle | T1 iso/hypo, T2 hyperintense heterogenous nonenhancing SOL | STR | Stable (2) |
| Xiong et al ⁴ (2019) | 38/M | Septum pellucidum | T1 hypo, T2 hyperintense with heterogenous enhancement | STR | Stable (6) |
| Al Krinawe et al ⁹ (2019) | 7/M | Septum pellucidum | Nonenhancing T1 hypo/T2 hyperintense mass | STR | Stable (24) |
| Sekar et al ¹⁵ (2019) | 18/M | Optic chiasma | T1 hypo, T2 hyper, multiple conglomerate ring enhancing lesion | STR | Stable (9) |
| Present Study (2021) | 32/F | Septum pellucidum | T1 iso/hypo, T2 hyperintense nonenhancing predominantly cystic mass | GTR | Stable (24) |

Abbreviations: GTR, gross total resection; SOL, space occupying lesion; STR, subtotal resection.

symptoms. Follow-up MRI is recommended at 3 months after surgery, semiannually for 2 years, and annually or once in 2 years thereafter.⁷

Recurrence of RGNT is also a well-documented event, which ranges as early as 1 month after surgery to as late as 9 years following decompression. Two cases of malignant transformation several years after surgery into glioblastoma (WHO-IV) have also been reported.⁸

Anatomically, septum pellucidum is one of the rare locations for RGNT and it came into clinical picture because of its tendency to cause ventriculomegaly due to compression over the bilateral foramen of Monro. Approximately 200 cases of RGNT have been reported till now, where incidence of two cases in septum pellucidum has been published by Xiong et al and Al Krinawe et al.^{4,9} The advancement of radiological, histological, and molecular details in establishment of neuropathological diagnosis should reveal the real enigma underlying the natural course of RGNT.

Conclusion

RGNTs are a rare CNS tumor entity and have recently been an interesting topic due to its occurrence at varied locations. Maximal safe resection and close follow-up results in better outcome in this tumor with mixed morphology.

Conflict of Interest

None declared.

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