Rosette-forming Glioneuronal Tumour:
An Educational Overview with Illustrative Cases

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Introduction

- ✓ Rosette-forming glioneuronal tumour (RGNT) is a rare CNS WHO grade 1 neoplasm.
- ✓ First described as a subtype of DNET in the cerebellum by Kuchelmeister et al. (1995).
- ✓ Recognised as a distinct clinicopathological entity by Komori et al. (2002).
- ✓ Incorporated into the WHO CNS classification in 2007, with updates in 2016 and 2021.
- ✓ It can pose a diagnostic challenge due to its rarity and overlapping features with other pathologies.

Objective

- ✓ A concise educational review of RGNT with emphasis on its key radiological characteristics.
- ✓ Illustrative cases from our neurocentre highlight typical radiological features and long-term behaviour.

Clinical presentation

- ✓ RGNT typically presents in young adults with symptoms related to its mass effect in the posterior fossa, such as headache, ataxia and visual symptoms.
- ✓ Typically indolent, but can disseminate or recur late.

Histopathology

- ✓ **Biphasic architecture:**
  - ✓ Neurocytic: rosettes and perivascular pseudorosettes.
  - ✓ Glial: pilocytic astrocytoma–like.
- ✓ **Immunohistochemistry:** synaptophysin+ in rosette cores; GFAP and S100+ in glial areas.
- ✓ **Molecular profile:** Hallmark is FGFR1 mutations.

Imaging features

- ✓ Location: Most frequently in the fourth ventricle and cerebellum, but has also been reported in lateral and third ventricles, pineal, hypothalamus, optic chiasm and spinal cord.
- ✓ General appearance: Well-demarcated, cystic and/or solid.
- ✓ CT: hypoattenuating – occasional calcifications or hydrocephalus - haemorrhage is rare.
- ✓ T1: hypointense.
- ✓ T2 & T2-FLAIR : hyperintense.
- ✓ DWI: no diffusion restriction due to low cellularity.
- ✓ Postcontrast: Highly variable - heterogeneous, rim, focal or none. Green bell pepper sign may be characteristic.
- ✓ Multiple satellite lesions – CSF dissemination.

Differential diagnoses

- ✓ Pilocytic astrocytoma: cyst with an enhancing mural nodule.
- ✓ Dysembryoplastic Neuroepithelial Tumour (DNET): supratentorial, cortically-based 'bubbly' multicystic lesions.
- ✓ Medulloblastoma: diffusion restriction.
- ✓ Ependymoma: plastic growth along ventricles – calcifications are more common.
- ✓ Gangliocytoma/Lhermitte Duclos: 'tigroid' appearance.
- ✓ MVNT: multiple small 'bubbly' lesions; no significant mass effect, more common supratentorially.
- ✓ Cysticercosis: A rare mimic.

Treatment

- ✓ Surgical resection is the primary treatment with excellent outcomes in most cases.
  - ✓ Gross total resection → lowest recurrence rates.
  - ✓ Subtotal resection: risk of recurrence; mandates long-term follow-up.

Illustrative cases

Case 1 – A 22-year-old female patient presented with an incidental posterior fossa tumour on MRI spine for leg pain.

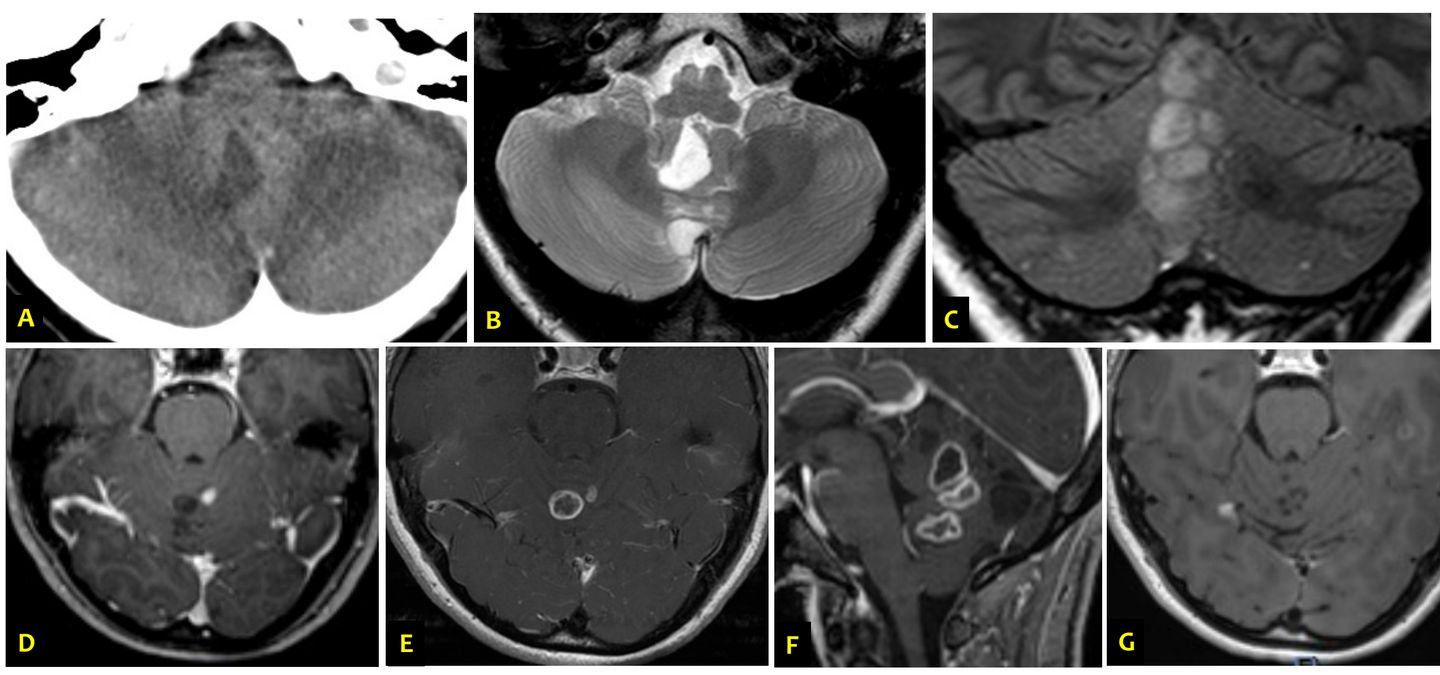

Figure 1 – A) Hypodense cystic lesions on CT. B-C) T2 and FLAIR hyperintense cystic lesions. D) Nodular enhancement in one of the cystic lesions with lack of enhancement in others. E-F) One year later, there is change in the size and enhancement pattern of the lesions. G) 13 years later, further change in size of the lesions with resolution of enhancement. Histopathology confirmed RGNT.

Case 2 – A 21-year-old male patient presented with an incidental posterior fossa lesions on a CT head after minor head trauma.

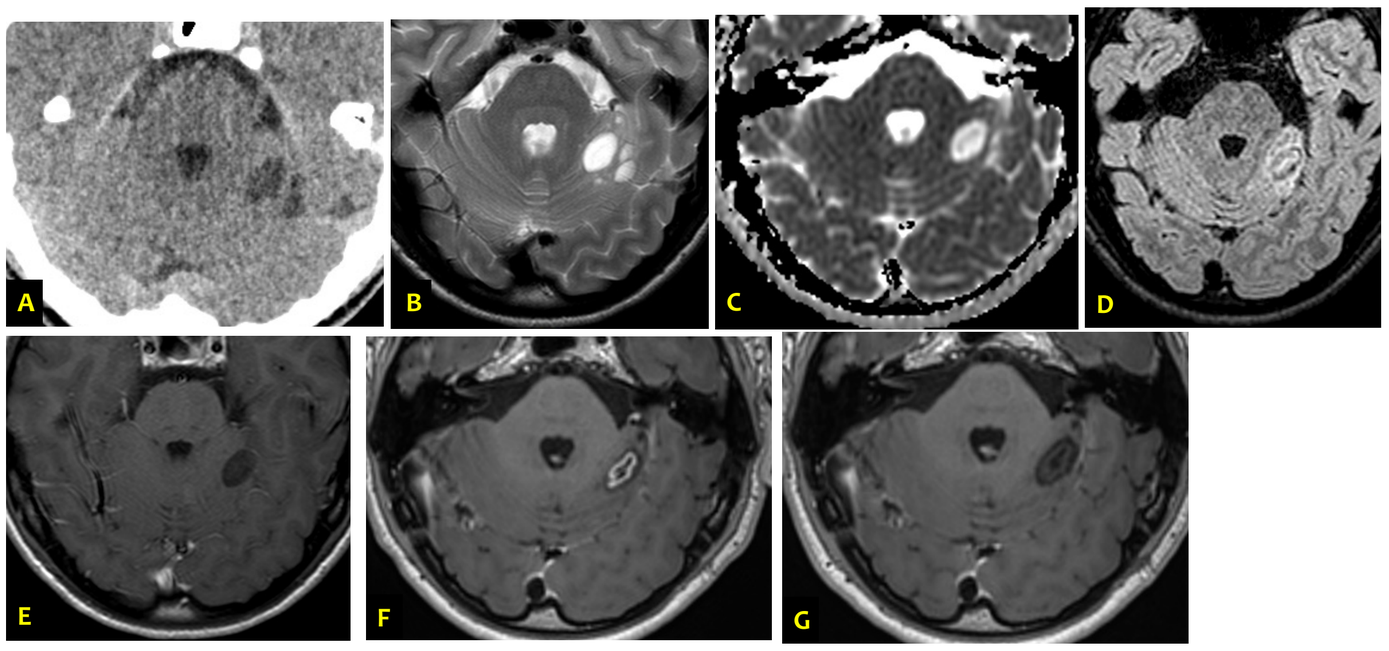

Figure 2 – A) Hypodense cystic lesions on CT. B) T2 hyperintense cystic lesions. C) Hyperintense lesion on ADC map in keeping with facilitated diffusion. D) Hyperintense signal on T2-FLAIR. E) No postcontrast enhancement. F) 6 years later, ring enhancement has developed in one of the cystic lesions with change in size of the cystic lesions. G) 1 year later, resolution of enhancement. Histopathology confirmed RGNT.

Conclusion

- ✓ Despite its rarity and variable imaging features, RGNT typically presents with recognisable radiological and histopathological features.
- ✓ The two cases presented show **a waxing-waning or migratory pattern of enhancement** which has been described in the literature and does not necessarily indicate tumour progression.
- ✓ Long-term imaging follow-up essential to capture behaviour variability.

Selected References

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Disclosures: Nothing to declare.