Embryonal Tumors with Multilayered Rosettes: A Comprehensive Case Report and Systematic Review

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Introduction

- Embryonal tumor with multilayered rosettes (ETMR), reclassified in 2016, is a rare and aggressive tumor occurring mostly in children (Fig 1, 2).[1-5]
- Characterized by amplification of the microRNA cluster locus 19q13.42 (C19mc).[1-5]
- Due to rarity, ETMRs pose significant clinical challenges due to limited data, and treatment options still have a poor prognosis.

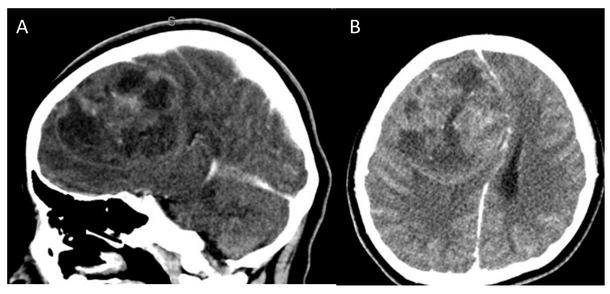

Figure 1. Sagittal (A) and axial (B) CT of frontal mass

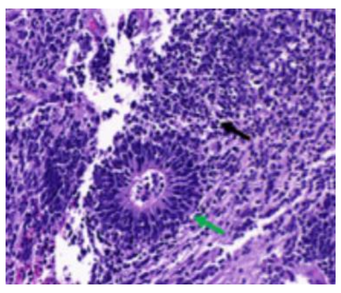

Figure 2. Multilayered rosette

Objectives

This systematic review and case report aims to synthesize current knowledge and treatment outcomes of ETMR to enhance prognostication and clinical management.

Methods and Materials

- Conducted a systematic review following PRISMA guidelines (Fig 3).
- Utilized PubMed, Embase, and Web of Science up to August 2023.
- Included case reports, case series, and retrospective/prospective studies.
- The patient case focused on clinical presentation, management, and outcome.

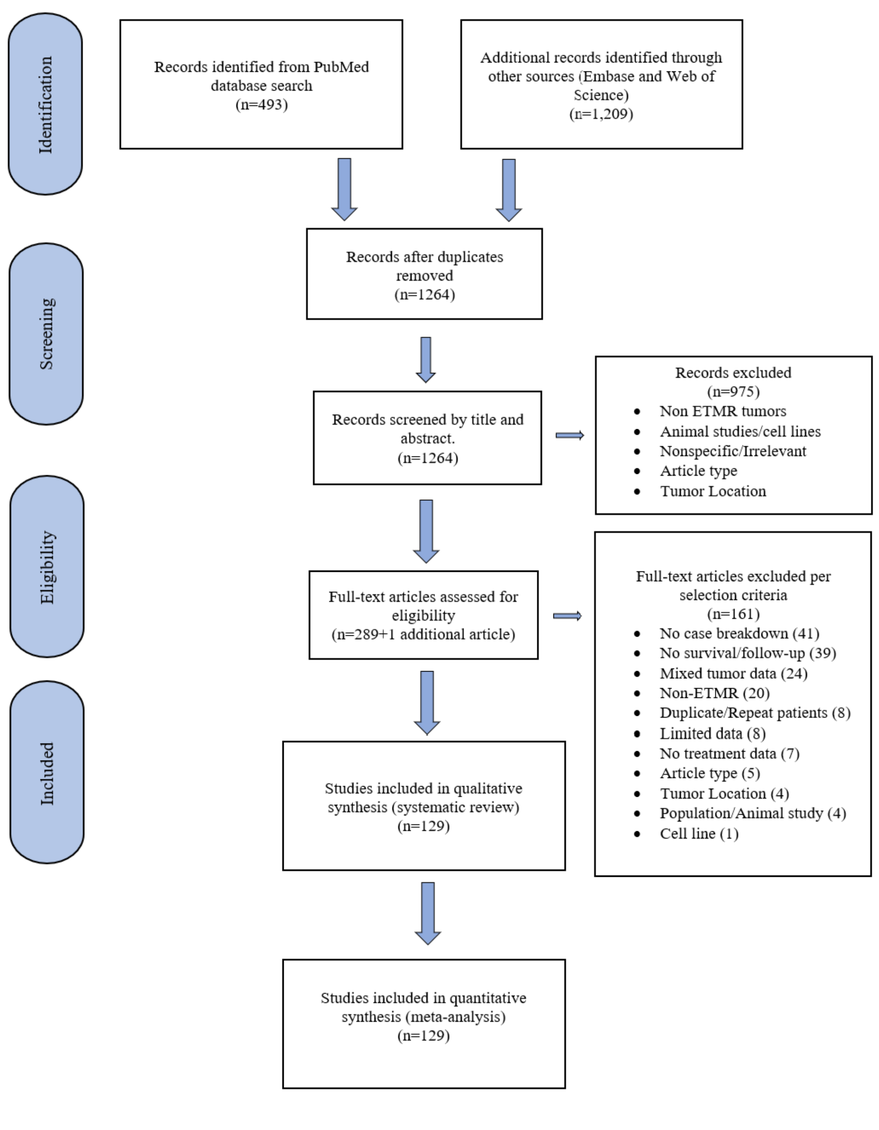

Figure 3. Publication retrieval displayed in a PRISMA flow chart

Results

9-year-old female presented with one week of headaches, N/V, and seizures. Molecular testing confirmed frontal lobe ETMR with C19mc amplification (Fig 1). Treatment consisted of gross total resection (GTR), chemotherapy, and radiation. The patient is in remission 24 months following diagnosis.

Results

- Included 340 patients with a median age of 2.5 years, 54.5% were female.
- C19mc amplification was reported for 111 (32.4%) patients.
- Patients with supratentorial tumors had longer median survival times (13 mo) compared to those with infratentorial tumors (7 mo), p-value < .05.
- The top 3 tumor locations were frontal lobe (14.6%), brainstem (± 4th ventricle, ± cerebellum) (14.6%), and parietal lobe (9.9%).
- Approximately 50% of patients survive to 1 year (Fig 4).
- GTR was significantly associated with improved survival outcome (Fig 5).
- The median overall survival was 11 months, with those who received GTR chemotherapy, and radiation, having the longest median survival of 24 months (Fig 6).

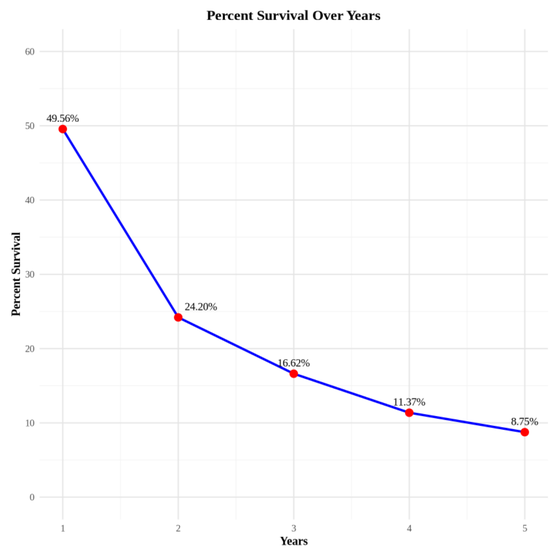

Figure 4. Percent survival from 1-5 years

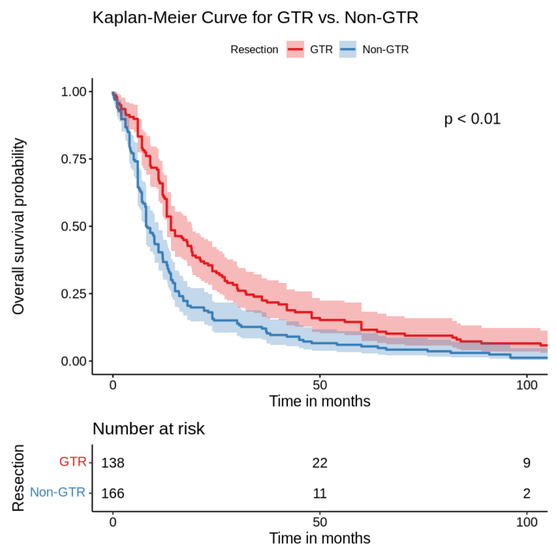

Figure 5. Kaplan-Meier curve of GTR vs. non-GTR

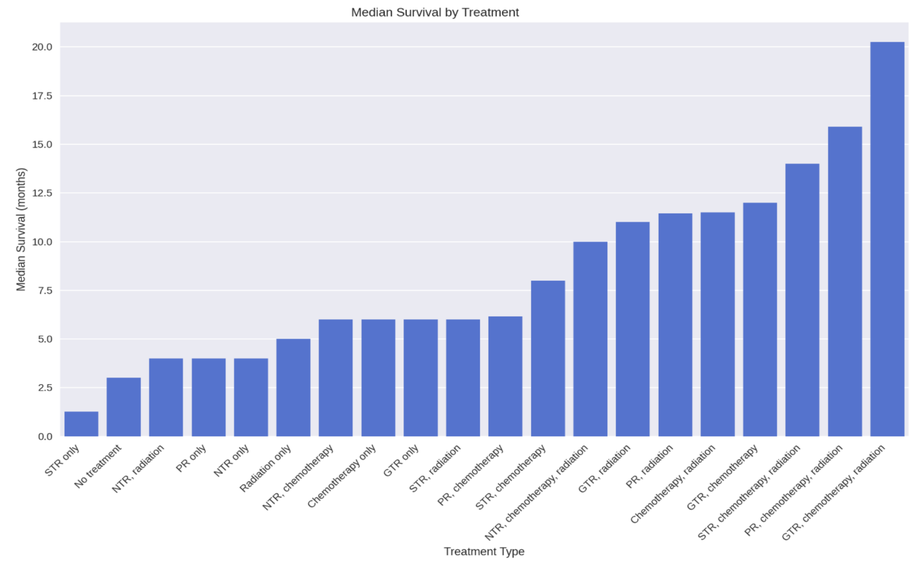

Figure 6. Median survival time by treatment received

Discussion

- This review highlights the critical need for a multidisciplinary approach in managing and treating ETMR.
- This diagnosis still carries a poor prognosis regardless of the association between comprehensive treatment, including GTR, and improved survival outcomes.
- Thus, this stresses the need for collaborative networks and multicenter studies to enhance surgical and adjuvant therapy options.

Conclusions

- This is the largest systematic review of ETMR based on the WHO 2021 classification.
- Tumor location, degree of surgical resection, and adjuvant therapy were associated with improved survival time.
- This study may help with prognostication in patients diagnosed with ETMR and hence aid in patient and parent counseling.

Contact

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