Clinical Presentation and Optimal Management of Intracranial Germ Cell Tumors Based on 98 Cases

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Disclosure

• There is no conflict of interest to disclose.
Aims and Objectives

• Germ cell tumors (GCTs) are a rare and challenging subset of intracranial neoplasms.

• Timely identification and multidisciplinary management are essential.

• We aim to;
  - document natural history in a large, longitudinal series
  - evaluate secondary clinical and/or cerebrospinal fluid (CSF) features that might enhance diagnostic speed and accuracy.
Methods

• Retrospective analysis of CNS GCT treated at Mayo Clinic, 1988-2017
• All cases involved neurosurgery service
• 80 cases of primary intracranial GCT
  (47 cases were germinoma: 59.8 %)
• 18 cases of metastatic intracranial GCT
• Clinical data, incl. age, sex, symptoms, location, treatment, recurrence, death were extracted
Most cases presented with endocrinological dysfunction (75.8 %), often accompanied by visual disturbance (45%).
Initial Symptom of Pineal GCT

**Hydrocephalus at presentation**
- n=9 (17%)
- n=45 (83%)

**Ophthalmic symptom at presentation**
- Diplopia (subjective): 9
- Upward-gaze palsy on exam: 10
- Pupillary light reflex deficit on exam: 18

*Most cases presented with hydrocephalus and disordered extraocular movements (86.1%).*
Cell Fraction in Pretreatment CSF

In germinomas, pre-treatment CSF testing showed significantly decreased monocyte and increased lymphocyte predominance compared with non-germinomatous GCT.
Local radiation demonstrated significantly worse progression-free and overall survival, as compared to cases treated with whole-brain/whole-ventricle radiotherapy.
Discussion

• Systematic review of GCTs demonstrated at least one hormonal abnormality in >90% intracranial GCT cases (Jorsal, et al.).

• Hankinson et al. reported almost all patients of pineal GCTs (28 out of 29) showed the ophthalmologic symptoms at presentation. Primary resection and post-chemotherapy resection were associated with worsening symptoms.

• Abundant lymphocytes in the CSF in germinoma cases may reflect the pathological characteristic of germinoma; abundant infiltration of small-sized lymphocytes among tumor cells described as “two-cell pattern”.

• Prior evidence support local RT as insufficient for germinoma in particular and CNS GCTs in general.

- Alapetite et al. Neuro Oncol 12(12):1318-1325, 2010
Conclusions

• Both anatomy and histopathology significantly impact GCT presentation and outcome.

• Pineal GCTs present with hydrocephalus and extraocular movement disorders, whereas neurohypophyseal GCTs present with pituitary insufficiency and visual deficit.

• Among germinoma, prognosis is significantly enhanced by ventricular coverage in radiotherapy.

• CSF cell fraction may have diagnostic significance, and should be prospectively studied as a potential diagnostic adjunct.