### EPEN-12. MULTIPLE RECURRENCES OF ANAPLASTIC EPENDYMOMA WITH EXTRA AXIAL AND EXTRA NEURAL METASTASIS IN A PEDIATRIC PATIENT

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Background: Ependymomas are the third most common CNS tumor in the pediatric population, accounting for 10% of all CNS tumors. Co-occurring extraneural and extracranial metastasis of ependymomas are extremely rare, with only 1 reported adult case in current literature. Case Description: We describe the case of a patient with multiple reoccurrences of anaplastic ependymoma. Initial imaging showed a 5 x 8 x 8 cm complex cystic mass with nodular enhancing components within the left occipital lobe. The 4th ventricle was intact and imaging was negative for metastasis. Pathology following resection demonstrated perivascular pseudorosettes, areas of calcification, and increased mitotic activity. Biopsy revealed GFAP, EMA, neurofilament, INI-1, and was negative for CAM5.2, confirming anaplastic ependymoma. Methylation studies for PFA or PFB subgroup differentiation were not available. The patient had recurrences at 4-, 5-, and 6-years after his initial diagnosis. Seven years from his initial diagnosis, the patient underwent resection of four nodular lesions from the occipital lobe and surrounding soft tissue. Pathology of these lesions and the lymph nodes/soft tissue confirmed anaplastic ependymoma. A PET scan showed increase uptake in the supraclavicular lymph nodes and had multiple bilateral pulmonary nodules. Scans at 3 months post-surgery were negative for leptomeningeal metastases but showed further lymph node involvement with progression of pulmonary disease. Conclusion: Co-occurring extraneural and extracranial metastasis of ependymoma is a rare occurrence across all populations. To our knowledge, this would be the first published pediatric case of anaplastic ependymoma with lymph node, soft tissue, and pulmonary involvement. Treatment of ependymoma itypically local and the utility of chemotherapy remains unclear. Treatment options for extraneural mets is very limited, illustrating the need for new therapies and further studies directed at understanding the biology of these tumors and the factors that could influence their ability to metastasize to extraneural and extracranial sites.

## GERM CELL TUMORS

### GERM-01. RECURRENCE PATTERN AND SURVIVAL FOR RELAPSED INTRACRANIAL NON-GERMINOMATOUS GERM CELL TUMORS: A SINGLE-INSTITUTION EXPERIENCE

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Purpose: Intracranial non-germinomatous germ cell tumors (NGGCTs) have lower overall survival than germinoma because relatively higher recurrence usually occurs after first line therapy. Methods: Between January 2003 and December 2018, 111 consecutive patients diagnosed with NGGCTs reviewed. Those who progressed after first line therapy were included in this study. Data of first line treatment, salvage treatment, clinicopathological features and survival were collected and analyzed. Results: Totally, thirty patients (30/111, 27.0%) relapsed in our cohort, including 19 patients with accurate relapse information detail, and 11 patients who died of disease progression during follow up but without exact time and site of relapse. The median OS from diagnosis of the disease was 49.2 months (95% CI: 14.1 to 84.3 months) and 3-year OS was 54.3%. Patients who received both CSI and chemotherapy relapsed less than those who received reduced volume of radiotherapy or only CSI or only chemotherapy (22.5% vs. 45.5%, p=0.034). Of 19 patients who had detail information of recurrence time and site, the median time from diagnosis of disease to relapse was 9.5 months (2.2 to 72.1 months). Regarding to recurrence site, most patients relapsed in primary site (10/19, 52.6%) or distant intracranial (6/19, 31.6%). The recurrence site of other 3 patients were spinal (n=1), ventricular (n=1) and peritoneal (n=1). Conclusion: Protracted follow-up is recommended because late recurrence is not uncommon. Primary tumor site and distant intracranial are the most prevalent relapsed location. Patients who relapsed could benefited from both CSI and salvage chemotherapy.

### GERM-02. MANAGEMENT STRATEGY FOR CHEMO-REFRACTORY, PROGRESSIVE PEDIATRIC IMMATURE TERATOMA

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Background: Localized NGGCT, a heterogeneous entity, treated with chemo-radiotherapy harbors an overall survival of around 93% based. Here, we present a case of a child whose immature teratoma progressed on chemotherapy, while tumor markers remain within normal range. Salvage therapy for progressive immature teratoma is variable, and can include surgery, chemotherapy and/or craniospinal radiation. Case: 10 year old boy with precocious puberty presented to the local Emergency room with tonic-clonic seizures. Imaging showed a localized pineal mass, with mixed cystic and solid components. Biopsy results were diagnostic for mixed germ cell tumor with components of immature teratoma. Prior to the start of chemotherapy, serum and cerebrospinal fluid (CSF) tumor markers showed slight elevation of both alpha feto- protein (AFP) and beta-human chorionic gonadotropin hormone (β-HCG). The patient underwent two cycles of chemotherapy per ACNS1123, stratum 1, with normalization of tumor markers. Unfortunately, near the end of Cycle 2 of chemotherapy, patient presented with clinical signs of herniation, and was noted to have significant progression of pineal mass on imaging. Tumor was gross totally resected, with pathology conclusive for only immature teratoma. Pre- and post-operative tumor markers remained normal. Proton-beam craniospinal radiation was then administered. Patient is now almost 6 months off therapy, with unremarkable serum and CSF tumor markers, as well as serial imaging that remains negative for disease, with neurological development appropriate for age. Conclusion: This case highlights the unusual nature of progression of an immature teratoma with no elevation of tumor markers while on chemotherapy. While salvage chemotherapy, in the form of a metronomic regimen or high-dose consolidative regimen, can be considered, these do present quite a bit of short-term and long-term toxicity to the growing child. Craniospinal irradiation followed by close monitoring is a reasonable alternative, with less short-term toxicity, for an entity that is radio-sensitive.

### GERM-03. CLINICAL FEATURES AND OUTCOME OF CHILDREN WITH INTRACRANIAL NON-GERMINOMATOUS GERM CELL TUMORS: A POPULATION-BASED STUDY IN HONG KONG

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The incidence of central nervous system non-germinomatous germ cell tumor (NGGCT) is four times higher in Chinese children than in the Western population. Reports on the outcome of Asian patients are nonetheless limited. Here we aim to summarize the experience of treating pediatric NGGCT in Hong Kong. Leveraging a population-wide pediatric oncology database, Chinese children with NGGCT diagnosed from 2002–2020 (n=43) were retrospectively studied. The diagnoses of NGGCT were made either with elevation in tumor markers (AFP/hCG; n=19), or by histology with/without concomitant raise in tumor markers (n=24). Most patients were treated with a combination of chemotherapy (cisplatin/etoposide/bleomycin, carboplatin/etoposide or carboplatin/etoposide/ifosfamide) and radiation (craniospinal+boost, whole ventricular+boost, or focal). The male:female ratio was 37:6, and the median age of diagnosis was 11.2 years. Primary tumor locations were pineal in 18, sellar/suprasellar in 12, basal ganglial in 9, supratentorial in 3 and posterior fossa in 1. Three had metastasis. Among the patients diagnosed by histology (n=24), 12 had mixed GCT, 8 had malignant/immature teratoma, 3 had embryonal carcinoma, and 1 had yolk sac tumor. With a median follow-up of 8 years, 8 patients progressed (local=7, distant=1), and 9 patients died (progression=5, palliative treatment for congenital tumors=2, sepsis=1, procedural complication=1). The respective 5-year PFS and OS were 74.9±6.9% and 82.2±6.1%. In multivariate analysis, high serum hCG level and no radiation use were significantly associated with inferior PFS. Outcome did not differ according to chemotherapeutic reagents used or radiation fields. Four patients had growing teratoma syndrome. Long-term neuroendocrine sequalae were common. In conclusion, children with NGGCT had reasonable outcome after multi-modal therapy in Hong Kong. Effort should be made to minimize tumor and treatment-related toxicities. The role of tumor markers for risk-stratification within NGGCT needs to be further interrogated.

### GERM-04. PRIMARY INTRACRANIAL GERM CELL TUMORS ARE MORE PREVALENT AMONG PEDIATRIC PATIENTS OF ASIAN/PACIFIC ISLANDER RACE/ETHNICITY IN THE UNITED STATES

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Introduction: Primary intracranial germ cell tumors (GCTs) appear to be more prevalent among pediatric patients in eastern Asia than in the U.S. Herein we use cancer registry data to evaluate whether GCT prevalence differs by race/ethnicity among U.S. pediatric patients. Methods: Pediatric patients (age≤14) presenting between 2004–2017 with a primary intracranial GCT were identified by ICD-O-3 histological and topographical coding from the National Cancer Database (comprising >70% of cancers newly-diagnosed cancers in the U.S.), and categorized by NICHD age stages. Patients' age, sex, race/ethnicity, and overall survival, and tumor location