Japan, [6]Department of Neurosurgery, Graduate School of Biomedical and Health Sciences, Hiroshima, Japan, [7]Department of Neurosurgery, Osaka University Graduate School of Medicine, Osaka, Japan, [8]Department of Pediatric Neurosurgery, Jichi Children's Medical Center Tochigi, Jichi Medical University, Tochigi, Japan, [9]Department of Neurosurgery, Graduate School of Medicine, Yokohama City University, Yokohama, Japan, [10]Department of Pediatric Neurosurgery, Osaka City General Hospital, Osaka, Japan, [11]Department of Pediatric Hematology and Oncology, Osaka City General Hospital, Osaka, Japan, [12]Division of Brain Tumor Translational Research, National Cancer Center Research Institute, Tokyo, Japan, [13]Department of Human Pathology, Gunma University Graduate School of Medicine, Gunma, Japan

BACKGROUND: Choroid plexus tumors (CPT) include choroid plexus papilloma (CPP), atypical choroid plexus papilloma (aCPP), and choroid plexus carcinoma (CPC). Because of their rarity, limited data are available on the current status of treatment and outcomes for pediatric CPTs. METHOD: We retrospectively reviewed clinical information on patients with CPT patients aged between 0 and 30 years at diagnosis and were treated in 8 institutions in Japan. RESULTS: Of forty-two cases initially diagnosed as CPT, 18 cases were reviewed by central pathologists. As a result, the diagnosis of CPC or aCPP in five cases were changed to other tumors including AT/RT and astroblastoma. The remaining 37 cases were subjected to analysis. Median age at diagnosis was two years (0 to 25) and the mean follow-up period was seven years. All 26 patients with CPP (n=20) or aCPP (n=6) underwent gross-total resection without adjuvant therapy. Of them 24 patients are alive without recurrence. Four patients of patients with CPC (n=11) died of cancer. Five patients including three patients experienced local relapse, achieved complete remission after resection of tumor plus chemoradiotherapy. All three patients with dissemination of CPC at diagnosis or relapse died of the disease. At least three patients were diagnosed with Li-Fraumeni syndrome: one died of medulloblastoma and one patient developed osteosarcoma. CONCLUSION: Compared with the excellent prognosis of CPP, the survival rates for CPC, especially disseminated CPC are unsatisfactory. Our results also underline the importance of considering genetic testing of TP53 for patients with CPC.

RARE-27. DOUBLE MUTATIONS: DIFFERENT GERMLINE AND TUMOR MUTATIONS LEAD TO POOR OUTCOMES

Molly Hemenway, Anan Nellan, Nicholas Foreman, Alexandra Suttman, and Kami Schneider; Univ of Colorado, SOM, Children's Hospital Colorado, Aurora, CO, USA

BACKGROUND: As genetic testing for both germline and tumor mutations has increased in completeness, complexity, and availability, more mutations and their impact on patient outcomes have been identified. METHODS: A retrospective review of pediatric patients who have identified germline mutations and a different tumor mutation was conducted. Data collected included demographics, tumor type, germline mutation status, tumor mutation status, relapse status, and patient outcome. RESULTS: Six patients aged 8–13 years old (median age 10 years) were identified for analysis. Four patients had pilocytic astrocytoma and two had pilomyxoid astrocytoma. One of the patients with pilocytic astrocytoma also had MPNST diagnosed very early at age 9. The combination of germline/tumor mutations is as follows: Neurofibromatosis Type I (NF1)/BRAF v600e, NF1, CHEK2/MYB-QKI, NF1, Klinefelter, ATM, MUTYH, GPC3/BRAF-KIAA fusion, NF1/BRAF-KIAA (2 patients), and Marfan's/BRAF-KIAA. The number of relapses per patient following initial diagnosis range from 3–7 with an average of 3.3. Four of the patients are alive and on therapy, which two are deceased. The two deceased patients both had NF1/BRAF-KIAA fusions and pilocytic astrocytomas. CONCLUSIONS: Patients with differing and compounded germline and tumor molecular genetic mutations have worse outcomes. These patients have more relapses and death when compared to those patients with one mutation, either germline or tumor. Broad molecular testing and germline testing for mutations is crucial in determining patient risk for poor outcomes.

RARE-29. PRIMARY CENTRAL NERVOUS SYSTEM NON-HODGKIN LYMPHOMA IN AN 11-YEAR-OLD BOY: A CASE REPORT

Jorge Luis Ramírez-Melo[1], Regina M Navarro-Martin del Campo[2,3], Manuel D Martinez-Albarran[1], Fernando Sánchez-Zubieta[4], Ana L Orozco-Alvarado[2], Luis A Arredondo-Navarro[5,3], and Lorelai Gutierrez-Oliva[5]; [1]Hospital Civil de Guadalajara "Dr. Juan I Menchaca", Guadalajara, Jalisco, Mexico, [2]Hospital Civil de Guadalajara "Dr. Juan I Menchaca", Guadalajara, Jalisco, Mexico, [3]GAPNO, INTERNATIONAL, Mexico, [4]Hospital Civil de Guadalara "Dr, Juan I Menchaca", Guadalajara, Jalisco, Mexico, [5]Hospital Civil de Guadalajara "Fray Antonio Alcalde", Guadalajara, Jalisco, Mexico

BACKGROUND: Primary central nervous system lymphoma (PCNSL) are very rare in children. CLINICAL CASE: An 11-year-old male presented with a 2 months history with myoclonic movements in the upper right limb, and a sudden frontal headache, gait disturbance due to right hemiparesis and an ipsilateral convulsive episode. Upon admission he had critical condition, with hypertensive skull syndrome, Glasgow of 12, Karnofsky 40%, right hemiparesis, swallowing disorder, facial paralysis, and loss of photo motor reflex and unilateral amaurosis. A CT and MRI showed a huge tumor mass in the left tempo-parietal region, infiltrating the white matter and shifting the midline. A Tumor biopsy was done, and reported diffuse small cell non-Hodgkin lymphoma of high-grade, Burkitt type. Systemic lymphoma workup was negative. He received six cycles of chemotherapy based on high dose methotrexate, rituximab and triple intrathecal.After the second cycle an ophthalmologic evaluation was done, and found infiltration to the right retina, for which 6 cycles of intra vitreous chemotherapy with methotrexate were applied, he showed an excellent response, and recovered all his neurological functions except that right hemianopia persist. Control MRI showed partial response at 2nd cycle and complete response after the 4th cycle. No Radiation was performed. CONCLUSION: This report highlights the fact that pediatric PCNSL may be effectively treated by a combination of HDMTX and rituximab-based chemoimmunotherapy without irradiation. Lack of awareness of this rare entity may lead to extense resections of brain, and potential permanent secuelae that were avoided in this illustrative case.

RARE-30. A RARE CASE OF PRIMARY EWING'S SARCOMA OF THE CERVICAL SPINE

Amandeep Kalra[1,2], Jayson Neil[2], Sarah Mann[3], and Pierre Giglio[4]; [1]Sarah Cannon Cancer Institute, Kansas City, MO, USA, [2]HCA, Kansas City, MO, USA, [3]UC Davis, Sacramento, CA, USA, [4]Ohio State University, Columbus, OH, USA

Ewing sarcoma family of tumors predominantly affect the pediatric population in the long bones of the extremities or the pelvis, and only 8% of cases arise within the spine. Primary Ewing's sarcoma of the cervical spine is extremely rare and less than 30 cases have been reported in the literature thus far. Here we present a case of primary Ewing's sarcoma of the cervical spine in a 28-year-old female who presented with a three-month history of neck pain and right arm radiculopathy. MRI revealed a homogeneously contrast enhancing, eccentric mass with dural tail at C2-C7. After undergoing a hemilaminectomy, histopathology confirmed extraosseous Ewing's sarcoma with CD99 positivity. A comprehensive systemic and neuraxis work-up ruled out overt metastasis. We extrapolated data from children's cooperative group studies and IESS-II clinical trial to formulate a three phase treatment protocol as described below. To date, patient is in remission with no evidence of any residual disease in the cervical spine. In conclusion, although Primary Ewing's sarcoma of the cervical spine is extremely rare it should be considered a differential diagnosis in patients with neck pain and a spinal mass under the age of thirty. Less than 25% of EFT's present with overt metastasis and almost all have subclinical metastatic disease at the time of diagnosis, therefore, a comprehensive evaluation and systemic chemotherapy is recommended. We recommend a multidisciplinary approach of surgical decompression to preserve neurological functions, followed by compressed chemotherapy regimens, reevaluation for local treatment, and adjuvant chemotherapy.

RARE-31. RECURRENT CHOROID PLEXUS CARCINOMA IN THE SETTING OF LI-FRAUMENI SYNDROME: REPORT OF TWO CHILDREN MANAGED WITH INTENSIVE RE-INDUCTION AND MARROW-ABLATIVE CONSOLIDATION CHEMOTHERAPY WITHOUT IRRADIATION FOLLOWED BY MOLECULARLY-TARGETED BIOLOGICAL THERAPY

Margaret Shatara[1], Iwona Filipek[2], Maciej Ciołkowski[3], Paweł Kowalczyk[3], Diana S. Osorio[1], Mohamed S. AbdelBaki[1], Elizabeth A. Varga[1], Rolla Abu-Arja[1], Claire Heinerich[1,4], Daniel R. Boué[5], Christopher R. Pierson[5], Jeremy Jones[6], Catherine E. Cottrell[7], Elaine R. Mardis[7], Richard K. Wilson[7], Jeffrey R. Leonard[8], and Jonathan L. Finlay[1]; [1]The Division of Hematology, Oncology, Blood and Marrow Transplant, Nationwide Children's Hospital and The Ohio State University, Columbus, OH, USA, [2]Department of Oncology, Children's Memorial Health Institute, Warsaw, Poland, [3]Department of Neurosurgery, Children's Memorial Health Institute, Warsaw, Poland, [4]Universidade de Uberaba Medical School, Uberaba, Brazil, [5]Department of Pathology and Laboratory Medicine, Nationwide Children's Hospital, Columbus, OH, USA, [6]The Department of Radiology, Nationwide Children's Hospital, Columbus, OH, USA, [7]The Steve and Cindy Rasmussen Institute for Genomic Medicine, Nationwide Children's Hospital, Columbus, OH, USA, [8]The Division of Pediatric Neurosurgery, Nationwide Children's Hospital and The Ohio State University, Columbus, OH, USA

BACKGROUND: The optimal management for children with recurrent choroid plexus carcinoma (CPC), is not established. We report two children with germline TP53 mutations, whose CPC relapses were managed with marrow-ablative chemotherapy and oral biologically-targeted therapies. PA-