(OS) were 9.7% (95% CI:2.6–36.0%) and 13% (95% CI:4.5–37.5%), respectively. Three patients survived beyond five years. Nineteen patients relapsed in the following sites: local site (n=4), distal site (n=6), and local and distal site (n=9). Favorable OS prognostic factors were: GS (hazard ratio [HR]=0.30 [0.11–0.86], p=0.025), and HDCx/AuHCR (HR=0.40 [0.16–0.99], p=0.047). CONCLUSION: CSI and HDCx/AuHCR were statistically associated with improved survival. The overall poor outcomes and high PD rates are common in the late recurrence cycles and following consolidation chemotherapy warrants consideration of fewer induction cycles before consolidation and the intensification of consolidation with multiple cycles of marrow-ablative chemotherapy.

RARE-36. DSYMBRYOPLASTIC NEOEPITHELIAL TUMORS: A REVIEW OF CLINICAL AND MOLECULAR CHARACTERISTICS, AND OUTCOME IN A PEDIATRIC POPULATION AT A SINGLE CENTER
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BACKGROUND: Neuronal and mixed neuro-glial tumors of the central nervous system (CNS) are relatively rare. Dysplastic neuroepithelial tumor (DNET) is a benign, rare, slow-growing tumor, but in many cases is associated with intractable epilepsy. OBJECTIVE: To report the experience with DNET at a single free-standing children’s institution. METHODS: A retrospective chart review of 24 patients with confirmed DNET between 2001 and 2019 was performed. Data was collected on clinical characteristics, surgical management and molecular findings, and outcomes. RESULTS: Mean age at diagnosis was 10 years (range 2 to 19 years), with female predominance (54.2%). Most common presenting symptoms were seizures (79.2%) and headaches (12.5%). Loca- tion of the tumor (DNET) is a benign, rare, slow-growing tumor, but in many cases is associated with intractable epilepsy. Temporal (29.3%), parietal (16.7%), cerebellar (12.5%) and occipital (4.2%). A gross total resection was achieved in half the cases. Recurrence occurred in 4 patients (16.7%), all of whom had subtotal resections. The average follow up since diagnosis was 4.6 years (range 0.3 to 14 years). Nearly half the patients were seizure free after surgery, 75% of which had a favorable outcome after surgery, a subset of patients remains symptomatic. As molecular mechanisms in DNET are unknown, future aim is to describe the molecular characteristics of our DNET population, and correlate with outcomes.

RARE-37. NOONAN SYNDROME AND GLIONEURAL TUMORS: A CENTRAL NERVOUS SYSTEM CANCER PREDISPOSITION ASSOCIATION
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BACKGROUND: Noonan syndrome (NS) is associated with germline Ras signaling pathway mutations, RAS overactivation and increased tumorigen- genesis risk. Rosette-forming glioneuronal tumors (RFGT) are rare in- dolent tumors. We report the molecular profiling of two patients with NS and RFGT. Patient 1: A 22-year-old male with a history of NS was diagnosed with RFGT after partial tumor resection followed by focal irradiation. He was enrolled on a comprehensive genomic profiling study involving paired tumor-normal whole exome sequencing and RNA sequencing of the disease-involved tissue, revealing a germline PTPN11 alteration (p.Gly60Ala) concordant with NS, and a somatic deletion (p.Leu442 Thr434del) in PIK3R1 and a somatic variant (p.Lys656Glu) in FGFR1 with concomitant increased expression of PIK3R1 and FGFR1 by RNA-seqencing. The patient remains without evidence of disease 11 years since initial presentation. Patient 2: A 19-year-old male with persistent headaches, underwent a brain MRI revealing a germline PTPN11 alteration (p.Asn308Asp) resulting in clinical features of NS, including his mother and two siblings, enabling appropriate counseling. Two somatic variants were found in trans in PIK3R1 (p Thr434_Phe436del and p.Glu431_Asn433delinsAsp), and a somatic variant (p.Val658Met) in FGFR1, with resultant overexpression of PIK3R1. The patient is monitored with surveillance imaging. CONCLUSION: We report the molecular profiling of two patients with NS and RFGT; strongly suggesting their connection to RASopathies through the overactivation of the MAPK and PI3K/AKT/MTOR signaling pathways.

RARE-38. CLINICAL PRESENTATION OF MGA-NUTM1 FUSION TRANSCRIPT SARCOMA
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BACKGROUND: MGA-NUTM1 fusion gene tumor are recently de- scribed as new subtype of NUTM1-rearranged tumors. Regarding its rarity, standard treatment has not been reported. Here we described clinical pres- entation, radiologic finding, immunohistological profile, and treatment of a boy with MGA-NUTM1 fusion transcript sarcoma. CASE REPORT: A 5-year-old boy with 2-month history of progressive right hemiparesis and headache. Magnetic resonance imaging (MRI) revealed 7.8 x10.6 x 8.0 cm well de- fined heterogeneous enhancing mass at left fronto-parietal lobe. CT chest and abdomen, bone scan, MRI spine, and CSF studies were unremarkable. He underwent craniotomy with total tumor removal. Pathology demonstrated high grade spindle cell sarcoma. The immunohistological profile was positive for BCCR, NUT1, and TEL1, but negative for CD34, STAT6, desmin, SMA, actin sarcoma, EMA, PR, S100, SOX10, and S100. He received postoperative radiation therapy. Tumors were negative for BCOR, NUT1, and TEL1, but negative for CD34, STAT6, desmin, SMA, actin sarcoma, EMA, PR, S100, SOX10, and S100. He received postoperative radiation therapy.

RARE-39. MOLECULARLY CONFIRMED ATYPIAL CHOROID PLEXUS PAPILLOMA WITH INTRACRANIAL DISSEMINATION
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INTRODUCTION: Among choroid plexus tumors (CPTs), metastasis occurs more frequently as pathologic grading increases. There could be an underestimation of pathological diagnosis if disseminated CPTs are diagnosed with lower grade tumors such as choroid plexus papilloma (CPP) or atypical choroid plexus papilloma (aCPP). Thus, molecular diagnosis using genome-wide DNA methylation profiling be useful to clearly define a CPP entity among the tumor entity. Here, we report about a case of aCPP with intracranial dissemination that was molecularly diagnosed by methylation profiling. CASE DESCRIPTION: A 2-year-old girl presented with a history of vomiting, Brain magnetic resonance imaging showed a large tumor mass in the right lateral ventricle and diffuse enhancement surrounding her brainstem, which suggested dissemination. Gross total resection of the mass was performed. Intraoperative findings revealed multiple spot metastatic lesions on the inner wall of lateral ventricle and intraventricular dissemination. The radiographic diagnosis was aCPP with intracranial dissemination. The histological findings were consistent with a glioblastoma multiforme with a papillary pattern suggesting a neoplasm of epithelial origin, increased cellularity, several necrotic areas, and an intermediate number of mitoses. The CPT-SIOP-2000 treatment protocol was followed. Diagnosis was confirmed by molecular analysis, which showed a MGA-NUTM1 fusion transcript of S100 and MGA-NUTM1 fusion transcript of S100. The patient is currently free of recurrence with no chemotherapy. The patient is currently free of recurrence.
published classifier, and the tumor was classified as methylation class "plexus tumor, subclass pediatric A" with high confidence (calibrated score 0.96), which includes cases diagnosed as CPP and cCPPs. CONCLUSION: Our case indicates the clinical significance of molecular confirmation of diagnosis among CPPs, particularly lower grade tumors with dissemination.

RARE-40. CASE REPORT: LONG-TERM SURVIVOR OF A RARE, PEDIATRIC PRIMARY HISTIOCTIC SARCOMA (HS) OF THE CENTRAL NERVOUS SYSTEM (CNS) FOLLOWING COMPLETE RESECTION, CHEMOTHERAPY AND ALLOGENIC HEMATOPOIETIC CELL TRANSPLANTATION (ALLO-HCT) Daiana S Osorno1, Rolisa Abu-Arja1, Mohamed S. Abeld-Rai2, Jeffrey R. Leonard3, Eric A. Sribnick4, Jonathan L. Finlay5, David W. Ellison6, Jennifer Picarsich7, Samir Kahwash7, and Daniel R. Buse1. 1The Division of Hematology, Oncology, Blood and Marrow Transplantation, Nationwide Children’s Hospital; and the Ohio State University, Columbus, OH, USA, 2Department of Pediatric Neurosurgery, Nationwide Children’s Hospital and The Ohio State University, Columbus, OH, USA, 3Department of Pathology, St. Jude Children’s Research Hospital, Memphis, TN, USA, 4Department of Pathology, Cincinnati Children’s Hospital, Cincinnati, OH, USA, 5Department of Pathology and Laboratory Medicine, Nationwide Children’s Hospital and The Ohio State University, Columbus, OH, USA

We report an unusual case of a patient with primary CNS-HS a very rare neoplasm of histiocytic lineage with usually poor prognosis. An 8 year old boy presented with a one month history of headaches, nausea and vomiting. Phys- ical exam revealed left high-riding scapula, bilateral ptosis, and localized 2.4 cm posterior fossa (cerebellar) mass with restricted diffusion. The patient underwent a gross total resection of the mass. Initial post-operative lumbar puncture was positive for malignant cells. Pathology showed a few histiocytes in a myxoid background. Brain MRI revealed a localized 3-cm myxoid mass with abundant eosophinophil cytoplasm, moderately pleomrophic nuclei and numerous mitotic figures, consistent with CNS-HS with juvenile xanthogranuloma phenotype, as supported by positive IHC expression of CD68, Lymphoid Myeloid markers, and BRAF v600e mutation. He was treated with two cycles of clofarabine and cytarabine and triple intrathecal (IT) chemotherapy. He developed generalized seizures and MRI showed demyelin- ation consistent with IT methylprednisolone toxicity. MTX was then discontinued. He was then given two additional cycles of clofarabine and weekly intrathecal therapy prior to consolidation with an Allo-HCT using a 10/10 HLA alli- matched unrelated donor. His conditioning regimen included total body ir- radiation and cyclophosphamide. He did well post-transplant with peripheral blood chimerism at 1 year showing >95% donor cells. He remains disease-free with an excellent quality of life since August 2016. We report one of the few known survivors of this unusual and highly malignant entity.

RARE-41. SECOND MALIGNANCIES FOLLOWING TREATMENT FOR PRIMARY CENTRAL NERVOUS SYSTEM TUMORS IN PEDIATRIC PATIENTS: A SINGLE-INSTITUTIONAL RETROSPECTIVE REVIEW Nicholas Petel, Erik Dedekam, M. Shahrar Salamat, and Diane Puccetti, University of Wisconsin, Madison, WI, USA

Second malignant neoplasms following treatment for primary central nervous system (CNS) tumors in children are rare occurrences but may often have dire consequences, particularly, if thought to be induced by prior therapies. The authors retrospectively reviewed pediatric patients with primary CNS malignancies from the University of Wisconsin over the last 25 years (1994 – 2019) with any secondary malignant neoplasm and determined seven patients met criteria. Treatment modalities were reviewed with all patients receiving surgery, chemotherapy, and radiotherapy for treatment of their first malignancy. The second neoplasms found included 4 high-grade gliomas, 1 menin- giaioma, 1 thyroid carcinoma, and 1 myelodysplastic syndrome. The median latency time between diagnoses was 9 years (range 4 - 17 years). The outcomes varied according to histopathology of the second neoplasm with the high-grade gliomas, meningioma, and thyroid carcinoma demonstrating the worst outcomes. The high-grade glioma patients were thought to have been induced by prior radiation in most cases. The re- maining patients are still alive, at the time of this writing, and in follow up after treatment for their second neoplasm. Thus, long-term follow up is essential for children treated for a primary CNS tumor given the variety of second neo- plasms that could arise with differential consequences. In addition to our single institutional outcomes, we will also present an updated review of the literature of pediatric patients with primary CNS tumors and second malignancies.

RARE-42. PRIMARY INTRACRANIAL SARCOMA WITH DICER1 MUTATION - TREATMENT RESULTS OF A NEW MOLECULAR ENTITY Ronald V Daza1,2, Martin Mynarek3, Sandro Casavilca4, Antonio Wachtel Aptomizer6, Pamela Mora4, Christian Koechele2,3, Andreas Von Deimling4,5, Ulrich Schüller6,10, Raymundo Sernaque12, Gustavo Sarri11,13, Tatiana Negreiros11, Luis Ojeda11, Pamela Garcia-Corrochano11,5, Camilo Ejerique11,6, Jeffrey R. Leonard3, Jonathan L. Finlay5, David W. Ellison6, Jennifer Picarsich7, Samir Kahwash7, and Daniel R. Buse1. 1The University of Wisconsin, Madison, WI, USA, 2Department of Pediatric Neurosurgery, Nationwide Children’s Hospital and The Ohio State University, Columbus, OH, USA, 3Department of Pathology, St. Jude Children’s Research Hospital, Memphis, TN, USA, 4Department of Pathology, Cincinnati Children’s Hospital, Cincinnati, OH, USA, 5Department of Pathology and Laboratory Medicine, Nationwide Children’s Hospital and The Ohio State University, Columbus, OH, USA

Objective: We present an unusual case of a patient with primary CNS-HS a very rare neoplasm of histiocytic lineage with usually poor prognosis. An 8 year old boy presented with a one month history of headaches, nausea and vomiting. Physical exam revealed left high-riding scapula, bilateral ptosis, and localized 2.4 cm posterior fossa (cerebellar) mass with restricted diffusion. The patient underwent a gross total resection of the mass. Initial post-operative lumbar puncture was positive for malignant cells. Pathology showed a few histiocytes in a myxoid background. Brain MRI revealed a localized 3-cm myxoid mass with abundant eosinophil cytoplasm, moderately pleomrophic nuclei and numerous mitotic figures, consistent with CNS-HS with juvenile xanthogranuloma phenotype, as supported by positive IHC expression of CD68, Lymphoid Myeloid markers, and BRAF v600e mutation. He was treated with two cycles of clofarabine and cytarabine and triple intrathecal (IT) chemotherapy. He developed generalized seizures and MRI showed demyelin- ation consistent with IT methylprednisolone toxicity. MTX was then discontinued. He was then given two additional cycles of clofarabine and weekly intrathecal therapy prior to consolidation with an Allo-HCT using a 10/10 HLA alli- matched unrelated donor. His conditioning regimen included total body ir- radiation and cyclophosphamide. He did well post-transplant with peripheral blood chimerism at 1 year showing >95% donor cells. He remains disease-free with an excellent quality of life since August 2016. We report one of the few known survivors of this unusual and highly malignant entity.

Pineoblastomas have been thought to portend a poor prognosis, especially in younger children or those with metastases. Long term survivors after relapse for those with metastatic disease are rare. We report a young girl with a DICER1 mutation who survived recurrent metastatic pineoblastoma. She was initially diagnosed at the age of 3 with a localized pineoblastoma, underwent gross total surgical resection, and received six cycles of cisplatin-based chemotherapy. Despite achieving a complete response, she relapsed at primary site with widespread spinal metastasis. She then received cranial spinal radiation of 3600 Gy with proton beam, which boosted to primary to 5580 Gy, followed by chemotherapy with temozolomide, Imitoside and Avastin per COG ACNS0334 without radiation therapy. 16 months after completion of treatment, she relapsed at primary site with widespread spinal metastasis. She then received cranial spinal radiation of 3600 Gy with proton beam, which boosted to primary to 5580 Gy, followed by chemotherapy with temozolomide, Imitoside and Avastin per COG ACNS0382. She is now 3 years and 3 months from completion of treatment, is doing well clinically with stable imaging findings. No particular alteration was identified from the tumor molecular testing of her initial pineoblastoma. Of note, she was diagnosed with pleuroperitoneal blastoma soon after her initial diagnosis of pineoblastoma, and was found to have a DICER1 mutation (c.3062C>T; p.R688*) thought to be a nonsense mutation. While radiation therapy followed by chemotherapy was administered, Response from the recurrent tumor was minimal. We hypothesize that tumors lacking the molecular features of high grade glioma also has a positive impact on prognosis. In addition, we speculate that DICER1...