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 aCPPs. CONCLUSION: Our case indicates the clinical significance of molecular confirmation of diagnosis among CPTs, particularly lower grade tumors with dissemination.

RARE-40. CASE REPORT: LONG-TERM SURVIVOR OF A RARE, PEDIATRIC PRIMARY HISTIOCYTIC SARCOMA (HS) OF THE CENTRAL NERVOUS SYSTEM (CNS) FOLLOWING COMPLETE RESECTION, CHEMOTHERAPY AND ALLOGENEIC HEMATOPOIETIC CELL TRANSPLANTATION (ALLO-HCT)

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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. Physical examination revealed nystagmus and dysmetria. Brain MRI revealed a 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 rare malignant cells. Pathology showed a focally necrotic neoplasm, composed of nests and cords of large relatively uniform cells with abundant eosinophilic cytoplasm, moderately pleomorphic nuclei and numerous mitotic figures, consistent with CNS-HS with juvenile xanthogranuloma phenotype, as supported by positive IHC expression of CD163, CD68, CD14, fascin, and Factor XIIIa, while negative for CD1a, Lymphoid and Myeloid markers, and BRAFv600e mutation. He was treated with two cycles of clofarabine and cytarabine and triple intrathecal (IT) chemotherapy. He developed generalized seizures and MRI showed demyelination consistent with IT methotrexate toxicity; MTX was then discontinued. He was then given two additional cycles of cladribine and weekly intrathecal therapy prior to consolidation with an Allo-HCT using a 10/10 HLA allelic-matched unrelated donor. His conditioning regimen included total body irradiation 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

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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 meningioma, 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 glioma patients all deceased from progressive disease. The high-grade gliomas were thought to have been induced by prior radiation in most cases. The remaining 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 neoplasms 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

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OBJECTIVE: An unexpectedly high incidence of sarcomas of the Central Nervous System (SCNS) was recently observed in Peru. We describe clinical and biological characteristics of the disease. METHODS: Seventy pediatric patients with primary SCNS diagnosed between January 2005 and June 2018 were analyzed. DNA methylation profiling and gene panel sequencing was available from 28 and 27 tumors, respectively. RESULTS: Median age was 6 years (range 2–17.5), 66/70 patients had supratentorial tumors, 56 patients intratumoral hemorrhage at diagnosis. Three patients fulfilled clinical criteria of NF1; 35 had café-au-lait spots and/or freckling. DNA-methylation profiling classified 28/28 as "intracranial spindle cell sarcoma with rhabdomyosarcoma-like features and *DICER1* mutations". *DICER1* mutations were found in 26/27, *TP53* mutations in 22/27, and *RAS*-pathway gene mutations (*NF1, KRAS, NRAS*) in 19/27 tumors, all of which were somatic (germline control available in n=19 cases). Survival was analyzed in 57 patients with non-metastatic disease who received adjuvant therapy. Two patients had metastatic disease, eleven did not receive or abandoned treatment. Two-year OS was 66.3% (95%-CI: 54–81%), 2-year PFS 51% (38–67%). PFS was highest in patients treated with postoperative ICE chemotherapy followed by radiotherapy and ICE (2y-EFS 79% [59–100%], n=18) and worse after upfront radiotherapy followed by ICE (40% [19–85%]; n=10) or VAC (50% [28–88%], n=12) and radiotherapy only (21% [6–71%], n=11; p=0.008). CONCLUSION: Primary SCNS with *DICER1* mutation have an aggressive clinical course. A combination of chemotherapy and radiotherapy seems beneficial. A link to a cancer predisposition syndrome could not be established so far.

RARE-43. FAVORABLE OUTCOME OF A YOUNG GIRL WITH RECURRENT METASTATIC PINEOBLASTOMA ASSOCIATED WITH A DICER1 MUTATION

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Pineoblastomas have been thought to portend a poor prognosis, especially in younger children or those with metastases. Long term survivors after relapse, especially 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 high dose chemotherapy with autologous stem cell transplant 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 3600Gy with proton beam, with boost to primary to 5580Gy, followed by chemotherapy with Temozolomide, Irinotecan and Avastin per COG ACNS0821. 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 pleuropulmonary blastoma soon after her initial diagnosis of pineoblastoma, and was found to have a DICER1 mutation (c.2062C>T; pR688*) thought to be a nonsense mutation. While radiation therapy following recurrence is known to improve the outcome, more recent studies suggest that tumors lacking the molecular features of high grade glioma also has a positive impact on prognosis. In addition, we speculate that DICER1