(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), local and distal sites (n=9). Favorable OS prognostic factors were CSI (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 rate during later induction 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. DYSEMBRYOPLASTIC NEUROEPITHELIAL 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. Dysembryoplastic 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, tumor location, surgical management, histopathological 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%). Location of the tumor was temporal (29.2%), frontal (25.0%), 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). Nineteen patients presented with seizures, of which 63.2% were seizure free after surgery. The samples with molecular genetic testing (microarrays or FISH), were all normal except one patient positive for BRAF V600E mutation. CONCLUSIONS: This is the first and largest review of pediatric DNETs in the last 10 years. Despite majority of patients having a favorable outcome after surgery, a subset of patients remains symptomatic. As molecular mechanisms in DNET remain unknown, future aim is to describe the molecular characteristics of our DNET population, and correlate with outcomes.

RARE-37. NOONAN SYNDROME AND GLIONEURONAL 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 tumorigenesis risk. Rosette-forming glioneuronal tumors (RFGT) are rare indolent tumors. We report the molecular profiling of two patients with NS and RFGT. PATIENT 1: A 22-year-old male with 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) consistent with NS, and a somatic deletion (p.Ile442_Thr454del) in *PIK3R1* and a somatic variant (p.Lys656Glu) in *FGFR1* with concomitant increased expression of *PIK3R1* and *FGFR1* by RNA-sequencing. The patient remains without tumor progression now nine months since irradiation. PATIENT 2: A 19-year-old male with persistent headaches, underwent a brain MRI demonstrated multiple abnormal signals in the pineal region and midbrain. He had a stereotactic biopsy revealing RFGT. He was enrolled on the genomic study revealing a germline *PTPN11* alteration (p.Asn308Asp) resulting in a new diagnosis of NS. Several family members were subsequently identified with clinical features of NS, including his mother and two siblings, enabling appropriate counseling. Two somatic variants were found in *trans* in *PIK3R1* (p.Thr454_Phe456del and p.Glu451_Asn453delinsAsp), and a somatic variant (p.Val695Met) 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 described as new subtype of NUTM1-rearranged tumors. Regarding its rarity, standard treatment has not been reported. Here we described clinical presentation, radiologic finding, immunohistological profile, and treatment of a boy with MGA-NUTM1 fusion gene tumor. CASE REPORT: A 13-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 defined 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 BCOR, NUT1, and TEL1, but negative for CD34, STAT6, desmin, SMA, actin sarcomeric, EMA, PR, S100, SOX10, BCL 6, and SABT2. The INI-1 showed nuclear expression and Ki-67 was positive in 50% of tumor nuclei. Molecular test for MGA-NUTM1 fusion transcript was positive, while SYT-SSX1, SYT-SSX2, and SYT-SSX4 fusion transcripts were negative. Four months after operation, MRI showed newly-seen two small enhancing foci at lateral and inferior aspects of the surgical cavity. He underwent re-surgery. Then focal radiation (54Gy and boost up to 60Gy at recurrent area) to the resection cavity was decided. Post-radiation chemotherapy including ifosfamide 3 g/m^2 and etoposide 150 mg/m^2 on Day 1–2, and carboplatin 500 mg/m^2 on Day 3, every 21–28 days was started. He has completed the first course of chemotherapy without any complication. CONCLUSION: MGA-NUTM1 fusion CNS sarcoma is rare. Treatment may require surgery, radiation and chemotherapy.

RARE-39. MOLECULARLY CONFIRMED ATYPICAL CHOROID PLEXUS PAPILLOMA WITH INTRACRANIAL DISSEMINATION

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INTRODUCTION: Among choroid plexus tumors (CPTs), metastasis occurs more frequently as pathological 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 may be useful to clarify malignant potential among thetumor 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. The pathological diagnosis was aCPP owing to the presence of a glandular structure 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 without radiation therapy, and the disseminated lesion was disappeared during the chemotherapy. Methylation data of the current case was entered into a recently