TIENTS: Patient A: A 17 months old male presented with non-metastatic bilateral CPC. A de novo mosaic germline TP53 mutation was identified. After near-total resections, 16 months of standard chemotherapy were administered; 18 months later, localized tumor growth developed, again near-totally resected. Two cycles of re-induction chemotherapy were administered followed by three cycles of thiotepa/carboplatin with autologous hematopoietic cell rescue (AuHCR) and subsequently 21 months of sirolimus and thalidomide, continuing without residual or recurrent disease. Patient B: A 30 months old male presented with left lateral ventricular non-metastatic CPC. A de novo TP53 germline mutation was identified. Following subtotal resection, craniospinal irradiation with boost was administered followed by eight cycles of standard chemotherapy; 18 months later, localized recurrence developed; gross total resection was followed by 15 months of standard dose chemotherapies; four months thereafter, a second local recurrence developed, again gross totally resected. He then received one cycle of high-dose cyclophosphamide followed by three cycles of thiotepa/carboplatin with AuHCR. Subsequently he received sirolimus and thalidomide for 12 months, complicated by progressive pancytopenia. A small localized CPC recurrence was noted, gross totally resected, concomitant with myelodysplastic syndrome; he underwent an allogeneic matched unrelated donor marrow transplantation. CONCLUSIONS: Marrow-ablative chemotherapy with post-transplant targeted biological therapy may afford durable survival for select children with recurrent CPC.

RARE-32. PEDIATRIC METASTATIC SKULL BASE CHORDOMA WITH TP53 MUTATION – A CASE REPORT AND REVIEW OF THE LITERATURE

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Chordoma is an uncommon bone tumor arising from notochordal remnant, which accounts for 1–4% of all bone malignancies. It commonly occurs along the cranial-spinal axis, and skull base is one of most frequent sites, representing 35–49% of all chordoma cases. Surgical resection is widely accepted as the first choice of treatment. There are only limited number of reports about pediatric chordoma cases, and its biological behavior including genetic backgrounds were largely unknown. Here, we present a 5 year-old girl with a large aggressive skull base chordoma of 6 cm in maximum diameter, which eventually had multiple systemic metastasis. We initially tried chemotherapy based on the protocol for the osteosarcoma, but in vain. Because the tumor was highly vascularized on angiography, after embolization of the feeding arteries and bilateral internal maxillary arteries, endoscopic endonasal surgery was performed. The tumor was sufficiently removed, achieving effective mass reduction, and the residual tumors involving the lower cranial nerves and craniocervial junction were additionally treated with Gamma Knife radiosurgery. However, one month later, it showed systemic metastasis to bilateral cervical lymph nodes and lung. We tried chemotherapy with nivolmab and imatinib for this patient, whereas they showed the partial effect. The genetic analysis revealed somatic TP53 c.569C>T, (p.P190L) mutation in chordoma specimen. In the past literature, we found only one study of the adult chordoma cases, in which majority of the patients had somatic TP53 mutation (p.P72R). Further investigation with large number of the cases is essential to clarify the molecular biology of pediatric chordomas.

RARE-33. GIANT CELL TUMOR OF THE SKULL BASE WITH A HISTORY OF A SUCCESSFUL RESPONSE TO DENOSUMAB AND LATER DEVELOPING A SECOND TUMOR

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BACKGROUND: Giant cell tumor of bone(GCTB) is a rare neoplasm with unpredictable behavior, possible malignant transformations, and/or lung metastases. Surgery is usually the treatment of choice. In unresectable or metastatic cases, treatment with denosumab is a new treatment option. CASE PRESENTATION: A 14-years-old female presented with cachexia, dysphagia, diplopia, discoordination, strabismus, and multiple cranial nerve palsies in 10.2015. MRI revealed intra-extracranial mass arising from C2 vertebrae, compressing the medulla oblongata and the left cerebellar hemisphere, invading to the sphenoid bone and nasopharynges. Biopsy showed a GCTB. Surgical resection was done, which was incomplete because of tumor location (cranial nerve and vertebral artery involvement). Then local radiation therapy was performed 50.4Gy. During RT patient's condition declined and MRI showed disease progression. Treatment with denosumab 120mg q4w was initiated in 03.2016, which yielded successful results. Disease was under control for three years until 03.2019. Then she returned with clinical symptoms of diplopia and severe headache. MRI showed local tumor progression. Repeated biopsy revealed undifferentiated pleomorphic sarcoma, which could be either a malignant transformation of GCTB or a new tumor. The patient later underwent two cycles of chemotherapy with Ifosfamide/Doxorubicin. MRI after 2nd cycle showed marked tumor progression. The patient didn't receive any further treatment because of cachexia and died due to disease progression in 12.2019. CONCLUSION: To our knowledge, this is the youngest patient ever reported with a skull base tumor with such a clinical development, successful and long-time remission with denosumab and with such a chemotherapy-resistant malignant transformation or second cancer.

RARE-34. UK CHILDREN'S CANCER AND LEUKAEMIA GROUP (CCLG): GUIDELINES FOR THE MANAGEMENT OF MENINGIOMA IN CHILDREN, TEENAGERS AND YOUNG ADULTS

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Primary tumours of the meninges are rare accounting for only 0.4–4.6% of all paediatric tumours of the central nervous system. Due to the rarity of these tumours in children, and the consequent absence of collaborative prospective trials, there is no clear consensus on how the unique characteristics of paediatric meningiomas impact clinical status, management approach, and survival. Much of the evidence and treatment recommendations for paediatric meningiomas are extrapolated from adult data. Translating and adapting adult treatment recommendations into paediatric practice can be challenging and might inadvertently lead to inappropriate management. In 2009 Traunecker *et al.* published guidelines for the management of intracranial meningioma in children and young people on behalf of UK Children's Cancer and Leukaemia Group (CCLG). Ten years later we have developed the updated guidelines following a comprehensive appraisal of the literature. Complete surgical resection is the treatment of choice for symptomatic meningiomas, while radiotherapy remains the only available adjuvant therapy and may be necessary for those tumours that cannot be completely removed. However, significant advances have been made in the identification of the genetic and molecular alterations of meningioma, which has not only a potential value in development of therapeutic agents but in surveillance of childhood meningioma survivors. This guideline builds upon the CCLG 2009 guideline. We summarise recommendations for the diagnosis, treatment, surveillance and long-term follow up of children and adolescents with meningioma.

RARE-35. PINEOBLASTOMA IN CHILDREN SIX YEARS OF AGE OR LESS: FINAL REPORT OF THE HEAD START I, II AND III EXPERIENCE

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BACKGROUND: We report the outcomes of patients with pineoblastoma enrolled on the Head Start I-III trials. METHODS: Twenty-three children were enrolled between 1991–2009. Treatment included maximal surgical resection followed by five cycles of intensive-chemotherapy and consolidation with marrow-ablative chemotherapy and autologous hematopoietic cell rescue (HDCx/AuHCR). Irradiation following consolidation was reserved for children over six years of age or those with residual tumor at the end of induction. RESULTS: The median age was 3.12 years (range:0.44–5.72). Three patients withdrew from the protocols and two patients experienced chemotherapy-related mortality. Eight patients experienced progressive disease (PD) during induction chemotherapy. Ten patients received HDCx/AuHCR; eight experienced PD post-consolidation. Seven patients received craniospinal irradiation (CSI) with a median dose of 20.7 Gy (range:18–36 Gy) with boost(s) (median dose 27 Gy, range:18–36 Gy); three received CSI as adjuvant therapy (2 post-HDCx/AuHCR) and four upon progression/recurrence. The 5-year progression-free survival (PFS) and overall survival

(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