Japan, 4Department of Neurosurgery, Graduate School of Biomedical and Health Sciences, Hiroshima, Japan, 5Department of Neurosurgery, Osaka University Graduate School of Medicine, Osaka, Japan, 6Department of Pediatric Neurosurgery, Jichi Children's Medical Center Tochigi, Jichi Medical University, Tochigi, Japan, 7Department of Neurosurgery, Graduate School of Medicine, Yokohama City University, Yokohama, Japan, 8Department of Pediatric Neurosurgery, Osaka City General Hospital, Osaka, Japan, 9Department of Pediatric Hematology and Oncology, Osaka City General Hospital, Osaka, Japan, 10Division of Brain Tumor Translational Research, National Cancer Center Research Institute, Tokyo, Japan, 11Department 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 (ACP), 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 was 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 follow-up period was seven years. 26 patients with CPT (n=20) or aCPP (n=6) underwent gross-total resection without adjuvant therapy. Of the 24 patients are alive without recurrence. Four patients with CPC with (n=11) died of cancer. Five patients including three patients who received adjuvant chemotherapy achieved complete remission after a combination of initial surgery 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 died of leukemia. CONCLUSION: The excellent prognosis of CPC, 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 GERMINE AND TUMOR MUTATIONS LEAD TO POOR OUTCOMES

Mollin Hernando, Nicholas Foreman, Alexandra Suttman and Kuni 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. In follow-up post included demographic data, initial patient status, tumor mutation status, tumor mutation status, relapse status, and patient outcome. RESULTS: Six patients aged 8–13 years (median age 10 years) were identified for analysis. Four patients had p53/astrocytoma and two had NOTCH1/astrocytoma. One of the patients with p53/astrocytoma also had MNPS3A diagnosed very early at age 9. The combination of germline/tumor mutations is as follows: Neurofibromatous Type I (NF1)/BRAF V600E, NF1/CHExKRAS, NF1, Ki67kelator, ATM, MUTH, GCP3/BRAF-KIAA fusion, NF1/BRAF-KIAA (2 patients), and Marfan’s/BRCA2 fusion, NF1/BRAF-KIAA fusions and pilocytic astrocytomas. CONCLUSIONS: Patients with differing and complicated germline and tumor 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 Ramirez-Melo1, Regina M Navarro-Martín del Campo2,3, Manuel D Martinez-Albarraz1, Fernando Sanchez-Zubiera1, Ana I Orozco-Alvarado1, Luis A Arredondo-Navarros2,4, and Lorela Gutierrez-Oliva2; 1Hospital Civil de Guadalajara “Dr. Juan I Menchaca”, Guadalajara, Jalisco, Mexico, 2Hospital Civil de Guadalajara “Dr. Juan I Menchaca”, Guadalajara, Jalisco, Mexico, 3GAPNO, INTERNATIONAL, Mexico, 4Hospital Guadalajara “Dr. Juan I Menchaca”, Guadalajara, Jalisco, Mexico, 5Hospital Civil de Guadalajara “Fray Antonio Alcalde”, Guadalajara, Jalisco, Mexico

BACKGROUND: Primary central nervous system lymphoma (PCNSL) are very rare in children. PCNSL is a rare pediatric disease with a 2 month 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 and was treated with hypertensive shock. Subsequently a CT and MRI revealed a huge tumor mass in the left tempo-parietal region, infiltrating the white matter and crossing 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 and complete response after the 4th cycle. He returned to normal and a second MRI revealed complete remission. CONCLUSION: The fact that pediatric PCNSL may be effectively treated by a combination of HD-MTX and rituximab-based chemotherapy without irradiation. Lack of awareness of this rare entity may lead to extreme resections of brain, and potential permanent sequelae that were avoided in this illustrative case.

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

Amandeep Kalsi1,2, Jayson Neif1, Sarah Mann1, and Pierre Giglio2; 1Sarah Cannon Cancer Institute, Kansas City, MO, USA, 2HCA, Kansas City, MO, USA, 3UC Davis, Sacramento, CA, USA, 4Ohio 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 develop by Ewing's sarcoma carcino in a spine. A 28-year-old female 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 extracranial Ewing’s sarcoma with CD99 positivity. A comprehensive systemic and neuraxis work-up revealed no extraneural metastasis. Conclusively, this case emphasizes the importance of both surgical decompression to preserve neurological functions, followed by comprehensive chemotherapy regimens, reevaluation for local treatment, and adjuvant chemotherapy.
A RARE-34. UK CHILDREN'S CANCER AND LEUKAEMIA GROUP (CCLG); GUIDELINES FOR THE MANAGEMENT OF MENINGIOMA IN CHILDREN, TEENAGERS AND YOUNG ADULTS

Elwira Szycht1, John Goodden1, Whitfield Gillian1, and Sarah Curry2,3

1Royal Marsden Hospital, London, United Kingdom, 2Institute of Cancer Research, London, United Kingdom, 3Leeds General Infirmary, Leeds, United Kingdom, 4Christie Hospital, Manchester, United Kingdom, 5Southampton Children's Hospital, Southampton, United Kingdom, 6Our Lady's Children's Hospital, Crumlin, Ireland

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 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 may lead to inappropriate management, toxicity, and survival. 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

Mohamed S. Abdel-Bakri1, Mohammad H. Abu-Awad2, Tom B. Davidson3, Jason R. Fungusaro4, Joseph R. Szczek, Ira J. Dunkel5, Girish Dhalla6, Sharon L. Gardner7,8, and Jonathan L. Finlay2,9

1National Cancer Center, Tokyo, Japan, 2Children’s Healthcare of Atlanta, Atlanta, GA, USA, 3Children’s Healthcare of Atlanta, Atlanta, GA, USA, 4Memorial Sloan Kettering Cancer Center, New York, NY, USA, 5The University of Alabama at Birmingham, Birmingham, AL, USA, 6The Stephen D. Hassenfeld Children’s Center for Cancer & Blood Disorders, New York, NY, USA

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 (HDCs/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 HDCs/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-HDCs/AuHCR) and four upon progression/recurrence. The 3-year progression-free survival (PFS) and overall survival

TIENTS: Patient A: A 17 months old male presented with non-metastatic bilateral CPC. A de novo mosaic germ line TP53 mutation was identified. After near-total resections, 16 months of standard chemotherapy were ad- ministered, and 18 months later, localized tumour growth developed, agaragmente, totally resected. Two cycles of re-induction chemotherapy were administered followed by three cycles of thiotepa/carboplatin with autologous hematopoietic stem 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 germ line mutation was identified. Following sub-total resection, craniospinal irradiation with boost was administered fol- lowed 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 re- currence 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 thalido- mide for 12 months, complicated by progressive pancytopenia. A small lo- calized CPC recurrence was noted, gross totally resected, concomitant with myelodysplastic syndrome; he underwent an allogenic 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

Akina Shimada1, Kazuhiko Karuozumi2, Kichihiro Kanamitsu3, Hisashi Ishida Ishida1, Kaori Fujitaira1, Kana Washio Washio1, Tadashi Kumatomo2, Chitose Ogawa2, and Masahiro Shin1

1Okayama University Hospital, Okayama, Japan, 2National Cancer Center, Tokyo, Japan, 3Tokyo University, Tokyo, Japan

Chordoma is an uncommon bone tumor arising from notochordal rem- nant, which accounts for 1–4% of all bone malignancies. It commonly oc- curs in cranial, cervical, and thoraco-lumbar regions; and might inadvertently be the first one of 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 metastases. 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 suffi- ciently removed, achieving effective mass reduction, and the residual tumors involving the lower cranial nerves and cranio-cervical junction were addition- ally treated with Gamma Knife radiosurgery. However, one month later, it showed systemic metastasis to bilateral cervical lymph nodes and lung. We tried chemotherapy with nolivomab 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 showed systemic metastasis to bilateral cervical lymph nodes and lung. 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 suffi- ciently removed, achieving effective mass reduction, and the residual tumors involving the lower cranial nerves and cranio-cervical junction were addition- ally treated with Gamma Knife radiosurgery. However, one month later, it showed systemic metastasis to bilateral cervical lymph nodes and lung. We tried chemotherapy with nolivomab 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 showed systemic metastasis to bilateral cervical lymph nodes and lung. We tried chemotherapy with nolivomab 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 showed systemic metastasis to bilateral cervical lymph nodes and lung. We tried chemotherapy with nolivomab 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 showed systemic metastasis to bilateral cervical lymph nodes and lung. We tried chemotherapy with nolivomab 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 showed systemic metastasis to bilateral cervical lymph nodes and lung. We tried chemotherapy with nolivomab 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 showed systemic metastasis to bilateral cervical lymph nodes and lung.