ETM2.22: TITLE: DEFINING THE CLINICAL AND PROGNOSTIC LANDSCAPE OF EMBRYONAL TUMORS WITH MULTIPLE ROSETTE (ETMR), A RARE BRAIN TUMOR REGISTRY (RBTG) STUDY

Sara Khan1, Palma Solano-Patez1, Tannu Suwal1, Salma Al-Karmi1, Mei Lu1, Ben Flo1, Maryam Fouladi1, Sarah Leahy1, Jean M. Muckley Levy2, Alvaro Lassaletta2, Eloy Rivas2, Alyssa Reddy2, G. Yancey Harper3, Naolin Gupta4, Michal Yalon-Orsen4, Laura Amariglio5, Hideo Nakamura5, Kuo-Sheng Wu6, Tai-Tong Wong6, Young-Shin Ra7, Milena La Spina7, Polidimilico Vittorino Emanuele8, Luca Massumi8, Anna Maria Buccelletti9, Jordana Tronchetti10, Richard G. Grundy11, Subhrendu Bandyopadhyay11, John Fangpiao12, David Scharnhorst12, Donna Johnston13, Lucie Lafay-Cousin13, Sandra Camelio-Pitragua14, Nabil Kabbara15, Amar Gajar16, Mahjoubat Bourt Aubarch15, Maria João Gol da Costa17, Derek Hanson18, Paul Wood19, Mayya Al-Hussami20, Nineen Amayri21, Yin Wang22, Daniel Catchpole23, Jean Michaud24, Anne E. Bendel24, Benjamin Ellezam25, Nicholas Gerber26, Ashley Plant27, Rubens Jefferies28, Christopher Dunham29, Christopher Moorertel30, Andrew Walter30, David Zigler31, Andrew Dosdghun32, Nicholas Gottardo32, Ahmet Demirtas33, Ramya Ramanujachar33, Eric Raabe34, Shapo Mary34, Peter Dirks35, Michael Taylor36, Hwang Eugene37, Holley Lindsey37, Tarik Tihan38, Jorgensen Mette39, Christine Dahl40, Sharon Low41, Amy Smith42, Lilo-Naz Hazzati43, Jesse Kresale44, Somers Gino44, Enrica Tan45, Division of Neuro-Oncology, Department of Pediatrics, Children's Hospital of Eastern Ontario, Ottawa, ON, Canada, 46Department of Laboratory Medicine, University of British Columbia, Vancouver, BC, Canada, 47Pediatric Hematology-Oncology, Department of Pediatrics, University of Minnesota Masonic Children’s Hospital, Minneapolis, MN, USA, 48Division of Pediatric Hematology/Oncology, Children’s Hospital, Wilmington, DE, USA, 49Children’s Cancer Institute, University of New South Wales, NSW, Australia, 50Department of Paediatrics, University of Otago, Christchurch, New Zealand, 51Kids Cancer Centre, Telethon Kids Institute, University of Western Australia, Perth, WA, Australia, 52Department of Hematology, Trakya University Medical Faculty, Edirne, Turkey, 53Paediatric Haematology and Oncology, Southampton Children’s Hospital, South Hampton, United Kingdom, 54Johns Hopkins School of Medicine, Sidney Kimmel Comprehensive Cancer Center, Division of Pediatric Oncology, Baltimore, MD, USA, 55The Hospital for Sick Children, Toronto, ON, Canada, 56Department of Oncology, Children’s National Medical Center, Washington, DC, USA, 57Center for Cancer and Blood Disorders, Phoenix Children’s Hospital, Phoenix, AZ, USA, 58Department of Pathology, University of California, San Francisco (UCSF), San Francisco, CA, USA, 59Great Ormond Street Hospital for Children NHS Foundation Trust, London, United Kingdom, 60Neurology Service, Department of Pediatrics, KK Women’s and Children’s Hospital, Singapore, Singapore, 61Orlando Regional Medical Center, Orlando, FL, USA, 62Paediatric Haematology/Oncology Service, KK Women’s and Children’s Hospital, Singapore, Singapore, 63Neuro Oncology Unit Department of Pediatric Hematology-Oncology and Hematology, Oncology, and Stem Cell Transplantation St Joan de Deu Hospital, Barcelona, Spain, 64Department of Human Pathology, Gunma University, Gunma, Japan, 65Department de Cancéropologie de l’Enfant et de l’Adolescent, Institut Gustave Roussy, Villejuif, Paris, France, 66PSI, Research University Institute Curie Research Center, Paris, France, 67APHM La Timone Hospital, Saint-Pierre, France

ETMR, an aggressive disease characterised by C19MC alterations, were previously categorised as various histologic diagnoses. The clinical spectrum and impact of conventional multi-modal therapy on this new WHO diagnostic category remains poorly understood as a majority of ~200 cases reported to date lack molecular confirmation. We undertook comprehensive multi-modal-pathologic studies to use large-scale uniformly-treated ETMR patients to improve disease recognition and treatment approaches. Amongst 623 CNS-PNETs enrolled in the RRTG registry, 159 primary ETMRs were confirmed based on a combination of FISH (123), methylation analysis (88), SNP and RNAseq (32) analyses; 91% had C19MC amplification/gains/fusions, 9% lacked C19MC alterations but had global methylation features of ETMR NOS. ETMRs arose in young patients (median age 26 months) predominantly as localized disease (MO-72%, M2-3% -18%) at multiple locations including cerebrum (60%) cerebellum (18%), midline structures (16%); notably 10% were brainstem primaries mimicking DIPG. Uni-and multivariate analyses of clinical and treatment details of curative regimens available for 110 patients identified metastatic disease (p<0.002), brainstem locations (p<0.005), extent of surgery, receipt of multi-modal therapy including high dose chemotherapy and radiation (p<0.001) as significant treatment prognosticators, while C19MC status, age and gender were non-significant risk factors. Analyses of events in all patients showed respective EFS at 3 and 12 months of 84% (95% CI: 77–91) and 37% (95% CI:20–41) and 4yr OS of 27% (95% CI:18–37) indicating despite intensified therapies, ETMR is a rapidly progressive and fatal disease. Our comprehensive data on the largest cohort of molecularly-confirmed ETMRs provides a critical framework to guide current clinical management and development of clinical trials.

GERM CELL TUMORS

GCT-02. THE LONG-TERM OUTCOMES AND SEQUELAE ANALYSIS OF INTRACRANIAL GERMINOMA FROM 187 PATIENTS IN THE SINGLE INSTITUTE: NECESSITY FOR THE ADAPTATION OF RADIOTherapy DOSE AND VOLUME

Joo Ho Lee1, Il Han Kim1, Keun Yong Eom2, Seung Ki Kim1, Kyeong-Chang Wang3, Tae Seog Hoo3, Hyoung Jin Kang3, and Hee Young Shin4, 1Seoul National University Bundang Hospital, Seongnam, Republic of Korea

PURPOSE: We aimed to refine the radiotherapy (RT) volume and dose determinant for disease failures and long-term sequelae in the intracranial germinoma. METHODS: The main treatment for intracranial germinoma was surgical therapy, followed by postoperative radiotherapy (CRT) (n=152) and chemotherapy (CRT) (n=152) during 1992–2015 in Seoul National Uni-
BACKGROUND: The optimal radiation field in patients with biframe germ cell tumors(GCTs) is controversial. METHODS: We retrospectively analyzed the medical records of 58 patients who were treated with radiotherapy for biframe GCTs. RESULTS: 27 patients were treated with focal radiotherapy(n=26) and 31 patients were treated with both focal and whole-brain radiotherapy(n=1). There was no difference in the recurrence rate between the two groups(P=0.446). CONCLUSIONS: Although the optimal radiotherapy field for biframe GCTs is controversial, our data suggest that both focal and whole-brain radiotherapy may be an appropriate treatment option for patients with biframe GCTs.

GCT-03. TREATMENT OUTCOMES, PHYSICAL DEVELOPMENT AND QUALITY OF LIFE OF PATIENTS WITH BIPOCAL GERM CELL TUMOR METHODS: We retro-

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GCT-03. TREATMENT OUTCOMES, PHYSICAL DEVELOPMENT AND QUALITY OF LIFE OF PATIENTS WITH BIPOCAL GERM CELL TUMOR METHODS: We retrospective-

GCT-06. DIAGNOSIS OF A RARE CASE OF RECURRENT GERM CELL TUMOR BY CSF PLACENTAL ALKALINE PHOSPHATASE PRESENTING WITH DIFFUSE INTRAAXIAL ABNORMALITY IN THE LOWER BRAINSTEM

INTRODUCTION: Germ cell tumors in the central nervous system (CNS) typically arise either at suprasellar and/or pineal region, and occasionally at basal ganglia. We report a case of diagnostically challenging, recurrent germ cell tumor presented with diffuse intraxial abnormality in and across the lower brainstem, which was diagnosed by the elevated placentalar alkaline phosphatase (PLAP) level in cerebrospinal fluid (CSF). CASE DESCRIPTION: A 28-year-old man had been treated by chemoradiotherapy at the age of 14 years for a left, thalamic GCT, followed by relapse at the age of 15 years. At the age of 20 years, he presented with a headache, nausea, and vomiting. MRI of the brain revealed diffuse hypointense signal abnormality in the lower brainstem, extending from the pons to the dorsal medulla oblongata. CSF analysis showed elevated PLAP level, suggestive of germ cell tumor. CONCLUSION: Despite the relatively low incidence, germ cell tumors of the brainstem should be considered in the differential diagnosis of extracranial origin PLAP-positive tumours. This series illustrates that the PLAP level in CSF can be a useful diagnostic tool for the diagnosis of recurrent germ cell tumors.