EMBRYONAL TUMORS: TWO CASE REPORTS
Aaron Goldberg,1 Chenue Abongwa,1 Jody Pathare,1 Clay Hoeric,2 Michael Mukheon,1 Joffre Olaya,1 Amar Gajar,1 Krista Warren,1 Ramesh Patil,1 Lai,1 William Lowery,3
1Children’s Hospital of Orange County, Orange, CA, USA,
2University of California, Irvine, Irvine, CA, USA,
3St. Jude Children’s Research Hospital, Memphis, TN, USA,
4Miller Children’s Hospital, Long Beach, CA, USA

We report two cases of unusual extraneural metastasis in patients with embryonal tumors without central nervous system disease progression and prolonged survival. The first patient presented at 16 years of age with atypical teratoid rhabdoid tumor of the cervical spine. The tumor was confirmed to be a lytic tumor of the spine and the chest wall. He received chemotherapy, with or without HD-MTX, for 11 years, followed by single or tandem HDcx-AuHCR. The second patient presented at 36 years of age with a cervical spine metastasis and was treated with HD-MTX and IVENT-MTX. The patient was in CCR at 11 years of age. We discuss these cases and outline a potential treatment strategy.

INTRODUCTION: Patients with recurrent medulloblastoma have a poor prognosis with only around 8% of patients surviving at 5 years irrespective of salvage therapy used. We report on 29 patients from four institutions treated with a “MEMMAT” based antiangiogenic combination therapy. PATIENTS AND METHODS: From 11/2006 to 06/2016, 29 patients were diagnosed with a recurrent medulloblastoma (19 first, 10 multiple recurrences). The median age of recurrence was 10 years (range 1–27). Subgroup of medulloblastoma was available in 18 patients and was group 3 or 4 in all except two (one WNT, one SHH-infant). For their current relapse patients received an antiangiogenic combination therapy consisting of bevacizumab, thalidomide, celecoxib, temafibrate, and etoposide, alternating with cyclophosphamide and augmented with intravenous chemotherapy. RESULTS: As of 01/2020, 8/29 patients are alive at a median of 44 months after recurrence. 6/8 surviving patients are currently in CCR between 66 and 134 months after recurrence that prompted MEMMAT therapy. Median age at starting MEMMAT was 10 years (range 3–47). Subsequent relapses were treated with bevacizumab alone or with temafibrate and etoposide. Conclusions: Our results suggest that antiangiogenic metronomic chemotherapy has clinical activity in recurrent medulloblastoma. Further investigation with an international phase II study is ongoing (MEMMAT: ClinicalTrials.gov Identifier: NCT01356290).

MBCL-38. UNUSUAL EXTRANEOUS METASTASIS OF PEDIATRIC EMBRYONAL TUMORS: TWO CASE REPORTS

MBCL-41. LYMPHOHEMATOPOETIC TOXICITY IDENTIFIED IN PATIENTS WITH MEDULLOBLASTOMA RECEIVING CRANIOSPINAL IRRADIATION
Asutoku Watanabe, Yumiko Shimizu, Atsuhiko Ohta, Takashi Fukushima, Tomonori Suzuki, Ryo Nishikawa, and Ruyhei Tanaka; Saitama Medical University International Medical Center, Hidaka-shi, Saitama, Japan

BACKGROUND: Medulloblastoma (MB) is the most common malignant brain tumor of childhood. MB easily disseminates through the spinal fluid, Surgery followed by radiotherapy, applied to the entire craniospinal axis (CSI), and adjuvant chemotherapy, represent the treatment of choice for patients aged ≥3 years. Since the bone marrow of the skull and vertebal column are the major hematopoietic organs, we investigated the myelosuppressive effect of irradiation treatment in patients with MB retrospectively. METHODS: Medical records of newly diagnosed MB patients treated in our hospital from 2007–2019 were analyzed. Children 2 years old were excluded because they did not receive CSI to avoid potential neurotoxicity. RESULTS: Medical records of 18 patients (11 males and 7 females, aged 6–26, median 11 years) were reviewed. Eight patients were stratified as group 3 or 4 in all except two (one WNT, one SHH-infant). For their current relapse patients received an antiangiogenic combination therapy consisting of bevacizumab, thalidomide, celecoxib, temafibrate, and etoposide, alternating with cyclophosphamide and augmented with intravenous chemotherapy (etoposide and liposomal cytarabine). RESULTS: As of 01/2020, 8/29 patients are alive at a median of 44 months after recurrence. 6/8 surviving patients are currently in CCR between 66 and 134 months after recurrence that prompted MEMMAT therapy. Median age at starting MEMMAT was 10 years (range 3–47). Subsequent relapses were treated with bevacizumab alone or with temafibrate and etoposide. Conclusions: Our results suggest that antiangiogenic metronomic chemotherapy has clinical activity in recurrent medulloblastoma. Further investigation with an international phase II study is ongoing (MEMMAT: ClinicalTrials.gov Identifier: NCT01356290).

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MBCL-46. TREATMENT OF RECURRENT WINGLESS-ACTIVATED MEDULLOBLASTOMA (WT-1 MB) INCORPORATING MARROW-ABLATIVE THIOTEGA AND CARBOPLATIN CHEMOTHERAPY (HDCTX) AND AUTOLOGOUS HEMATOPOIETIC PROGENITOR CELL RESCUE (AHUPCR): A DUAL REPORT
Micah K. Harris, Zachary N. Funk, Daniel R. Boss, Christopher P. Pearson, Jeremy Jones, Jeffrey Leonard, Rollu Aba-Aja, Jeffrey Auleta, Diana S. Osorio, Margaret Shatara, Stephan R. Paul, Jonathan L. Finlay, and Mohamed S. AbdelBaki; The Division of Hematology, Oncology, Blood and Marrow Transplant, Nationwide Children’s Hospital and The Ohio State University, Columbus, OH, USA, The Ohio State University College of Medicine, Columbus, OH, USA, Department of Pathology, Nationwide Children’s Hospital and The Ohio State University, Columbus, OH, USA, The Department of Radiology, Nationwide Children’s Hospital and The Ohio State University, Columbus, OH, USA, The Division of Pediatric Neurosurgery, Nationwide Children’s Hospital and The Ohio State University, Columbus, OH, USA, Section of Pediatric Hematology/Oncology, West Virginia University Healthcare Children’s Hospital, Morgantown, WV, USA

BACKGROUND: Wnt-MB infers an excellent prognosis, and metastatic disease is rare. However, specific treatment strategies and patterns of failure for patients with recurrent Wnt-MB are unknown. We report two cases of...
recurrent beta-catenin nucleopositive Wnt-MBs treated with an irradiation-sparing strategy, incorporating HDCx/AuHPCR. PATIENT 1: A 4-year-old female experienced local recurrence of a non-metastatic Wnt-MB nine months after gross total resection (GTR) followed by 18Gy craniospinal irradiation (CSI) with primary site boost to 54Gy, accompanied by weekly vincristine, followed by a maintenance regimen of nine cycles of cisplatin/vincristine alternating with cyclophosphamide/vincristine every third week. The recurrent tumor was followed by three cycles of HDCx/AuHPCR. She is disease-free over three years following relapse treatment. PATIENT 2: A 17-year-old male initially underwent GTR, followed by 23.4Gy CSI with 54Gy posterior fossa boost with concomitant weekly vincristine, followed by a maintenance regimen that included nine alternating cycles of vincristine/lomustine/cisplatin and cyclophosphamide/vincristine. Isolated right fronta! horn metastatic recurrence developed 19 months post-treatment; three cycles of irinotecan/temozolomide/bevacizumab and gamma-knife radiosurgery produced complete response. A second isolated metastasis recurrence on the left frontal horn occurred 13 months post-treatment, which was treated with two cycles of cyclophosphamide/etoposide followed by two cycles of HDCx/AuHPCR. MRI of the brain showed no residual tumor one month post-treatment. He currently awaits follow-up stereotactic radiosurgery. CONCLUSION: Patients with recurrent Wnt-MB may be treated with curative intent using a multidisciplinary approach that includes HDCx/AuHPCR, and minimization or avoidance of re-irradiation.

MBCI-48. OUTCOMES OF TREATMENT BASED ON THE ST. JUDE MEDULLOBLASTOMA-96 REGIMEN FOR JAPANESE CHILDREN WITH MEDULLOBLASTOMA

Junya Fujimura1, Tomonori Suzuki2, Yoko Watanabe2, Hidetaka Niiizuma3, Ryuta Saito4, Masayuki Kanazari5, Yukihiko Sonoda2, Atsuko Watanabe2, Ryuhei Tanaka3, Megumi Fujimura2, Akimi Yaguchi2, Taeko Ishii2, Takeshi Ooshima2, Akiko Tomita2, Akimi Kanituri2, Takaaki Yanagisawa2, Teiji Tominga6, Ryo Nishikawa7, and Hajime Arai1; 1Department of Pediatrics, Juntendo University Faculty of Medicine, Bunkyo-ku, Tokyo, Japan, 2Department of Neuro-Oncology/Neurosurgery, Saitama Medical University International Medical Center, Hidakaka, Saitama, Japan, 3Department of Pediatrics, Tohoku University Graduate School of Medicine, Sendai, Miyagi, Japan, 4Department of Neurosurgery, Tohoku University Graduate School of Medicine, Sendai, Miyagi, Japan, 5Department of Neurosurgery, Jikei University School of Medicine, Minato-ku, Tokyo, Japan, 6Department of Neuro-Oncology/Hematology, Saitama Medical University International Medical Center, Hidaka, Saitama, Japan, 7Department of Neurosurgery, Juntendo University Faculty of Medicine, Bunkyo-ku, Tokyo, Japan, 8Department of Neurosurgery, Jikei University School of Medicine, Minato-ku, Tokyo, Japan

Medulloblastoma is a type of malignant embryonal tumor in childhood that is considered to require multiagent chemotherapy followed by radical resection and craniospinal irradiation (CSI). However, the outcomes of chemotherapy for this tumor in Japan are unclear. Here, we performed a multicenter retrospective study to determine the prognosis of pediatric medulloblastoma patients in Japan treated with the St. Jude medulloblastoma-96 (SJMB96) regimen. Thirty patients with newly diagnosed medulloblastoma received treatment with the SJMB96 regimen at Juntendo University Hospital in Tokyo (n=10), Saitama Medical University International Medical Center in Saitama (n=10), and Tohoku University Hospital in Miyagi (n=10) from 2011 to 2018. All patients underwent tumor resection and CSI, with radiation doses of 23.4Gy for standard-risk patients (n=11) and 39.6Gy for high-risk patients (n=19). Six weeks after radiation therapy, patients received four cycles of high-dose chemotherapy with autologous peripheral blood stem cell transplantation according to the SJMB96 regimen. We found that 5-year overall survival was 80.8% among standard-risk patients and 74.2% among high-risk patients. No treatment-related deaths occurred. Eight patients who experienced recurrence died within 80 months of diagnosis. As these treatment outcomes are comparable to those previously reported outside of Japan, our findings indicate that this regimen is a therapeutic option for medulloblastoma patients in Japan.

MBCI-50. DISMAL OUTCOME OF HIGH RISK MEDULLOBLASTOMA TREATED WITH CHEMOTHERAPY FIRST APPROACH IN MALAYSIA

Shiao Wei Ouah, El Abdulk Rahman, M Mohd Ibrahim, Z Muda, Is Othman, MN Mohamed Umni, K Gunasagar, MP Ang, CB Goh, and KH Teh; Paediatric Haematology & Oncology Unit, Paediatric Department, Hospital Kuala Lumpur, Kuala Lumpur, Malaysia

INTRODUCTION: Patients with high risk medulloblastoma are treated either with high dose chemotherapy or hyperfractionated radiotherapy. Both approaches are not feasible in resource-limited countries. POG9031 trial has reported favourable outcome for high risk medulloblastoma using standard chemotherapy and radiotherapy only. Hence, we have adopted the protocol using chemotherapy first approach due to logistical reasons. OBJECTIVE: To review the outcome of children diagnosed with high risk medulloblastoma in Hospital Kuala Lumpur. METHODS: Patients diagnosed with high risk medulloblastoma between January 2015 and June 2018 treated using the chemotherapy first approach as per POG/9031 protocol were identified. Data was then extracted and analysed. RESULTS: Nine patients in total were identified, 3 boys and 6 girls. Median age at diagnosis was 9.3 years (2.6 – 15.9 years). Median follow up for survivors is 3.6 years. Five patients (55.6%) had macroscopic metastatic disease at diagnosis. All patients had significant residual disease post-op. Only 3 patients are disease free till last follow up, giving a 3 years event free survival of 16%. Of the 6 patients who had no residual disease, 4 have died, giving a 3 years overall survival of 46%. Patient with no metastasis at diagnosis (M0) fared better with 3 years event free survival of 38%, but 3 years event free survival for patients with macroscopic metastatic disease (M+) was 0%. CONCLUSION: Outcome of children with high risk medulloblastoma treated with chemotherapy first approach was dismal.

MBCI-51. POST-AUTOLOGOUS HEMATOPOIETIC CELL TRANSPLANTATION (AuHCT) PRACTICES FOR YOUNG CHILDREN WITH MALIGNANT BRAIN TUMORS

Mahvish Rahim1,2, Jeffrey Auletta3,4, Gurseh Dhali5,6, Jonathan Finlay1,6, and Scott Coven3,4; 1Indiana University School of Medicine, Indianapolis, IN, USA; 2Riley Hospital for Children, Indianapolis, IN, USA; 3Nationwide Children’s Hospital, Columbus, OH, USA; 4The Ohio State University, Columbus, OH, USA; 5Children’s of Alabama, Birmingham, AL, USA; 6University of Alabama at Birmingham, Birmingham, AL, USA

BACKGROUND: “Head Start” protocols have used autologous hematopoietic stem cell transplant (AuHCT) for infants and young children with recurrent brain tumors in order to avoid cranial irradiation. The post-AuHCT practice for children with a brain tumor diagnosis varies greatly. The goal of this research study is to explore practices and attitudes about post-AuHCT care for children with brain tumors. DESIGN: An anonymous REDCap survey link was provided to all site primary investigators and additional support personnel at “Head Start” institutions. The survey questions defined the role of the medical provider completing the form and explored the various practices relating to transition, management, communication and other aspects. RESULTS: Twenty of the 37 individual responders have been received so far. The majority report that prophylactic medicines were discontinued upon WBC recovery; however, management of discontinuation was split evenly between the neuro-oncology and stem-cell transplant teams. Nearly half of responders follow T-cell recovery following transplant without immunolympho guidance. Post-AuHCT vaccination practices are highly variable, with no clear consensus. Lastly, most responders reported adequate ease of transition and communication between the neuro-oncology and transplant teams. CONCLUSIONS: This work underscores the need for both multidisciplinary communication for children with brain tumors in the post-AuHCT period and for the development of standardized vaccination and other prophylaxis practices.

MBCI-52. ENDOCRINE PROFILE AFTER MEDULLOBLASTOMA TREATMENT

Miriam Pavon-Mengual1, Helen Curry2, Vrinda Saraf2, Zaianah Mohamed3, Helen Benshoff4, Daniel Ford5, Andrew Peet5, Jenny Adamski6, and Martin English7; 1Birmingham Women’s and Children’s NHS Foundation Trust, Birmingham, United Kingdom; 2University Hospitals Birmingham NHS Foundation Trust, Birmingham, United Kingdom; 3University Hospitals Birmingham NHS Foundation Trust, Birmingham, United Kingdom

BACKGROUND: Treatment of medulloblastoma has evolved substantially with more chemotherapy, risk-adapted dosing of radiotherapy (RT) and new RT techniques. We present the endocrine profile for our patients treated over a 20-year period. METHODS: The charts of patients treated for medulloblastoma between 1/1/00 and 31/12/19 were reviewed. 105 were available. Group 1 received chemotherapy alone, Group 2 received 23.4Gy whole CNS RT with PF boost to 54Gy, Group 3 received > 35 Gy whole CNS RT with PF boost to 54–59 Gy, Group 4 received PF RT to 54 Gy. All received chemotherapy according to national guidelines or clinical trials relevant at the time. RESULTS: Group 1 (MF 3:16, 7 survivors mean age 2 years range 1–7) had no endocrinopathies. At 5 years from diagnosis Group 2 (MF 15:13) and Group 3 (MF 35:14) had the following % RESULTS: Survival 77:61; Growth Hormone deficiency 92:100; Thyroid deficiency 75:81; ACTH deficiency 42:33. Girls were more likely to need sex hormone replacement than boys. Group 4 (MF 7:27, who aged 2) were all treated in the first decade. 3 survivors, one GH deficiency, one thyroxine deficiency, one both. CONCLUSIONS: There is a trend to earlier endocrinopathies in the group 5 vs group 2 patients, but it does not reach statistical significance. Girls are more likely to need sex hormone replacement than boys. This investigation provides a contemporary profile of...