treat it. An eight years-old boy presented with decrease level of consciousness with headache, limb weakness, slurred speech, double vision, hemifacial weakness, paraplegia, and disequilibrium which worsened two weeks prior to admission. He was initially noted to have a medially inverted left eye two months prior to admission. On physical examination, there was a N.VI and N.VII palsy, hyperreflexia, paraplegia, and nystagmus. Head MRI demonstrated a solid with minimal enhancement after contrast administration in the left pons region extending into mesencephalon with cystic component and flat floor of fourth ventricle sign, causing hydrocephalus, suggestive diffuse intrinsic pontine glioma. He was given fractionated focal intensity modulated radiation therapy (IMRT) to the tumor along with 5 mm margins with a total dose of 54 Gy. 1.8 Gy fractions, given once daily for 5 days per week over a period of 6 weeks. Supportive care in the form of corticosteroids is used to treat the peri-tumoral edema. There was a short period of motoric improvement. But then the disease progressed with the latest head CT showed hydrocephalus formation with increased size of mass. He is treated with chemotherapy using Temozolomide, and currently alive after 3 months post diagnosis.

Keywords: childhood brain tumour, Diffuse Intrinsic Pontine Glioma, radiation therapy

DIPG-34. LIFE AND END-OF-LIFE AFTER DIFFUSE INTRINSIC PONTINE GLIOMA DIAGNOSIS

Idoya Serrano Pejenaute[1], Ricardo López-Almaráz[2,3], Aizpea Echebarria[2,3], Rosa Adán[2,3], Jimena De Pedro[2,3], Paula Gonzalez[2,3], Jesús Sanchez[4], Julio López-Bayón[4], Itziar Astigarraga[2,3], Miguel García-Ariza[2,3]; [1]Department of Pediatrics. Cruces University Hospital, Barakaldo, Basque Country, Spain. [2]Department of Pediatrics, Pediatric Hematology and Oncology Unit. Cruces University Hospital., Barakaldo, Basque Country, Spain. [3]Biocruces Health Research Institute, Barakaldo, Basque Country, Spain. [4]Department of Pediatrics, Pediatric Home Hospitalization and Pediatric Palliative Care Unit. Cruces University Hospital, Barakaldo, Basque Country, Spain

INTRODUCTION AND OBJECTIVES: Despite advances in pediatric oncology, Diffuse Intrinsic Pontine Glioma's (DIPG) life expectancy remains <10% at 2 years, and the mean survival time is 8-10 months from diagnosis. Its location associates multiple symptoms of complex management that condition their quality of life. The aim is to review our experience in the clinical course of the disease and the involvement of palliative care (PC). METHODS: Single-centre retrospective study of patients <18 years diagnosed with DIPG between 2011-2022. A review of the electronic medical record was carried out, recording demographic data, oncological treatments, PC and advance care planning (ACP). RESULTS: We registered 14 patients (2 alive). Median age at diagnosis was 6.4 years (range 3.5-11.9). Median survival from diagnosis was 9.6 months (range 0.5-18) and 3.8 months from progression (range 0.1-12). At diagnosis, 92% received oncological treatment (radiotherapy 75%, chemotherapy 41%, oncolytic viruses 21%, partial resection 8%). After progression, 4 patients received metronomic chemotherapy or re-irradiation and in the last month of life 2 received chemotherapy. In the last 3 months of life, 64% were admitted at least once for progression or end-of-life. In the last month, all received oral dexamethasone. Follow-up by PC team was provided in 64%, with first contact in the last month of life in 36%. ACP was recorded in 4 patients, all of them followed up for PC. Death occurred at home in 33%, hospital in 58%. Palliative sedation was used in 5 patients. CONCLUSIONS: Despite the known prognosis of DIPG, some patients continue receiving oncological treatment after progression and in the last month of life. Contact with PC teams occurred mainly at advanced disease stage. Median survival of children with DIPG is below one year, with multiple symptoms in the progression phase. Therefore, early follow-up by PC teams is recommended.

DIPG-35. PERSONALIZED TREATMENT FOR MOLECULARLY HETEROGENEOUS DIFFUSE MIDLINE GLIOMA, H3 K27-ALTERED PAEDIATRIC CASE

Nahla Ali Mobark[1], Musa Alharbi MD[1], Yasser S. Bayoumi[2], Aymn Albanyan[3], Ali Abdullah O. Balbaid MD[2], Wael abdel Rahman Aljabarat[1], Malak Abedalthagafi, MD[4]; [1]Department of Pediatric Oncology Comprehensive Cancer Centre, King Fahad Medical City, Riyadh, Saudi Arabia. [2]Radiation Oncology Department, Comprehensive Cancer Centre, King Fahad Medical City, Riyadh, Saudi Arabia. [3]Pediatric Neurosurgical Department, King Fahad Medical City, Riyadh, Saudi Arabia. [44]Genomics Research Department, Saudi Human Genome Project, King Fahad Medical City and King Abdulaziz City for Science and Technology, Riyadh, Saudi Arabia

Diffuse midline glioma, H3 k27-altered (DMG) is a type of Paediatric- type diffuse high grade gliomas according to the 2021 WHO CNS tumors Classification. Diffuse intrinsic pontine glioma (DIPG) is another acceptable related term when it located in the pons with fatal prognosis. The combination of H3K27M with BRAF V600 mutations rarely reported in DMG although more commonly in Paediatric-type low grade gliomas (Diffuse low-grade glioma, MAPK pathway-altered). We present a twenty-month-old boy, previously healthy, presented with 2 weeks history of unsteady gait, drooling, cranial nerves palsy MRI imaging showed diffuse pontine mass with classic radiological features of DIPG. 2.6 x 1.6 x 3.2 (AP x TV x CC) with no evidence for spinal metastases. Patient underwent right retro sigmoid approach and open biopsy of lesion he received focal Radiation Therapy 54GY/30fx as stander of care of DIPG with mild neurogical improvement Pathological & molecular Diagnosis was Diffuse midline glioma, H3 k27-altered with Co-occurrence BRAF V600E mutation. Two months after end of radiation, he presented with vomiting, and neurological deterioration with new right-side hemiplegia. Imaging studies showed interval increase in the pontine lesion with increased edema causing narrowing of the fourth ventricle, no active hydrocephalus. He was started on combination therapy BRAF inhibitor Dabrafenib and MEK Inhibitor Trametinib as maintenance therapy the patient gradually showed Marked neurological and clinical improvement. A 6-month MRI after start of targeted therapy showed favorable treatment response with complete resolution of the previous diffusion restriction, reduced tumor volume on MR perfusion ,reduced perilesional edema otherwise almost stabilization of nonenhancing pontine lesion. The poor prognosis of recurrent DIPG is well known but our patient is clinically and radiologically stable with excellent quality of life and well tolerating the therapy . Our case show that personalized treatment approach that address molecular heterogeneity of H3K27M glioma are safe and feasible

DIPG-36. THE BRAIN-GUT-MICROBIOTA AXIS TO PREDICT OUTCOME IN PEDIATRIC DIFFUSE INTRINSIC PONTINE GLIOMA

Loris De Cecco[1], Veronica Biassoni[1], Elisabetta Schiavello[1], Andrea Carenzo[1], Maria Federica Iannò[1], Armando Licata[1], Manuela Marra[2], Maria Carollo[2], Luna Boschetti[1], Maura Massimino[1]; [1]Fondazione IRCCS Istituto Nazionale dei Tumori, Milan, Italy. [2]Istituto Superiore di Sanità, Roma, Italy

OBJECTIVE: Diffuse intrinsic pontine glioma (DIPG) is a rare childhood brain tumour with poor prognosis. Radiotherapy (RT) remains the only palliative intervention. In this scenario, it is essential to investigate tumor and patient microinvironment. Microbiota plays a critical role in human health and a functional link between the central nervous system and gut microbiota has been reported: the microbiota-gut-brain axis. With these premises, we have investigated the gut microbiota in DIPG. METHODS: A cohort of 18 patients was enrolled and we collected stool specimens at diagnosis (pre-RT) and after radiotherapy. Microbiota content was analysed through 16S rRNA sequencing on IONS5xl; base calling and demultiplexing were performed by Torrent Suite (ThermoFisher). Association to progression free survival (PFS) and overall survival (OS) were assessed by cox-regression univariate analyses and differential abundance (DA) analyses identified differences following RT. RESULTS: The Firmicutes/Bacteroidetes (F/B) ratio in pre-RT specimens has a median of 0.757 (range= 0.243-1.19). Having PFS as endpoint and at family classification level, an increased risk of disease progression was disclosed for Flavobacteriaceae and Bacillales with HR of 1.57 (p=0.00913) and 1.57 (p=0.215), respectively. The anaerobic bacteria Synergistaceae is found related to a decreased risk of progression with HR 0.662 (p=0.00718). These findings were also confirmed with OS as endpoint. DA analyses pointed out a number of families belonging to Firmicutes and Bacteroidetes phyla highly differentiated (log2|fold-change|>2) between pre- and post-RT. CONCLUSIONS: This study represents the first report of the microbiota-gut-brain axis role in DIPG patients. The F/B ratio, a parameter of normal intestinal homeostasis, show the presence of inflammatory patterns and some family members are associated to increased disease progression. These preliminary findings needs further validations. However, 16S microbiota has the potential ability to stratify patients and probiotic administration could restore a normal F/B ratio.

DIPG-37. EXPLORING THE ROLE OF THE EPIGENETIC FACTOR H2A.Z ACETYLATION IN DIPG

Yolanda Colino Sanguino[1,2], Laura Rodriguez de la Fuente[1], Dana Kisswani[1], Padraic Kearney[3,4], Evangeline Jackson[3,4], Ryan Duchatel[3,4], Holly Holliday[1,2], Nada Jabado[5], Maria Tsoli[1,2], David Ziegler[1,2], Matthew Dun[3,4], Fatima Valdes Mora[1,2]; [1]Children's Cancer Institute, Sydney, NSW, Australia. [2]UNSW, Sydney, NSW, Australia. [3]University of Newcastle, Callaghan, NSW, Australia. [4]Hunter Medical Research Institute, New Lambton Heights, NSW, Australia. [5]McGill University, Montreal, Quebec, Canada

Diffuse intrinsic pontine glioma (DIPG) is the most aggressive brain tumor found in children with a peak incidence of 5-7 years of age, with median survival after diagnosis less than one year. In more than 60% of DIPG cases, a recurring somatic mutation in the H3F3A gene, that causes a lysine 27 to methionine substitution is seen in the histone variant H3.3 (H3.3K27M). Wildtype histone variants H2A.Z and H3.3, are frequently found in the same nucleosome and cooperate to regulate transcription. We