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Recurrent pediatric high-grade glioma (pHGG) is the leading cause of cancer-related mortality in children. Immunotherapy is a successful treatment approach for a growing number of cancers and is being investigated as a treatment strategy for pHGG. Immunotherapy has shown the most benefit in tumors with increased infiltrating T cells at baseline. Our recently published results revealed that neoadjuvant checkpoint inhibition in recurrent adult glioblastoma was associated with upregulation of a T cell and interferon-γ-related gene expression signature (Tcell-IFNγGES) and was correlated with a significantly extended overall survival (OS). In this study, we examined the immune landscape in recurrent pHGG and the association of Tcell-IFNγGES in the tumor with survival. We analyzed tumor RNAseq data collected at time of recurrence from a historical cohort of 42 pHGG patients from the Children's Brain Tumor Tissue Consortium. We found a significant transcriptional enrichment of Tcell-IFNγGES in 54% of the tumors. The survival of patients with high Tcell-IFNγGES was observed to be significantly higher than patients with low Tcell-IFNγGES, (log-rank p=0.05). The 3-year OS for patients with low *versus* high Tcell-IFNγGE was 28.5% (95%, CI:13.7%-59.5%) compared to 50.2% (95%, CI:33.1%-76.1%). When patients were stratified by age, gender and race, low Tcell-IFNγGES was found to be a poor OS prognostic factor (hazard ratio=2.4 (1.14–5.14), p=0.02). This indicates a strong relationship of decreased Tcell-IFNγGES and increased risk of death. Future investigations are necessary to validate these findings, and to explore the value of Tcell-IFNγGES as a predictive biomarker for response to immunotherapy in pHGG.

IMMU-31. PNOC007: H3.3K27M SPECIFIC PEPTIDE VACCINE COMBINED WITH POLY-ICLC FOR THE TREATMENT OF NEWLY DIAGNOSED HLA-A2+ H3.3K27M DIFFUSE MIDLINE GLIOMAS (DMG)

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OBJECTIVE: To assess safety and efficacy within a multi-center trial the H3.3K27M specific peptide vaccine with poly-ICLC in HLA-A02.01$^+$ patients diagnosed with H3.3K27M$^+$ DMGs. METHODS: After focal radiation therapy, participants 3–21 years of age were enrolled into two strata. Stratum A: newly diagnosed diffuse intrinsic pontine glioma (DIPG); Stratum B: other DMGs. H3.3K27M vaccine was administered with poly-ICLC IM every 3 weeks for 8 doses followed by every 6 weeks for a total of 96 weeks. Immuno-monitoring of peripheral blood mononuclear cell (PBMC) and imaging occurred every 3 months. Modified iRANO criteria were applied. PBMC samples were evaluated by mass cytometry. RESULTS: From November 2016 until March 2019, 19 eligible patients (median age 11, range 5–17 yrs; 53 % female) were enrolled in Stratum A and 10 eligible patients (median age 13, range 7–18 yrs; 60 % female) in Stratum B. Treatment was well tolerated (7 grade 3; 0 grade 4 related toxicities). Median number of vaccines per participant was 6 (range 1–11). Overall survival at 12 months was 40% (95% CI 22–73%) for Stratum A and 39% (95% CI 16–93%) for Stratum B. Among the 19 subjects with longitudinal immune cell assessments, 7 exhibited an expansion of K27M-reactive CD8+ effector memory T-cells correlating with prolonged survival (p=0.028). CONCLUSION: H3.3K27M specific vaccine in combination with poly-ICLC is well tolerated. CyTOF-based immune monitoring of PBMCs facilitates sensitive high-throughput analysis. Further investigation is warranted to determine if this may be predictive of clinical outcomes.

LOW GRADE GLIOMA

LGG-01. CLINICAL MANAGEMENT AND GENOMIC PROFILING OF PEDIATRIC LOW-GRADE GLIOMAS IN SAUDI ARABIA

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Pediatric Low Grade Gliomas (PLGGs) display heterogeneity regarding morphology, genomic drivers and clinical outcomes. The treatment modality dictates the outcome and optimizing patient management can be challenging. In this study, we profiled a targeted panel of cancer-related genes in 37 Saudi Arabian patients with pLGGs to identify genetic abnormalities that can inform prognostic and therapeutic decision-making. We detected genetic alterations (GAs) in 97% (36/37) of cases, averaging 2.51 single nucleotide variations (SNVs) and 0.91 gene fusions per patient. The *KIAA1549-BRAF* fusion was the most common alteration (21/37 patients) followed by *AFAP1-NTRK2* (2/37) and *TBLXR-PI3KCA* (2/37) fusions that were observed at much lower frequencies. The most frequently mutated) genes were NOTCH*1 3* (7/37), *ATM* (4/37), *RAD51C* (3/37), *RNF43* (3/37), *SLX4* (3/37) and *NF1* (3/37). BRAF V600E mutations were observed in only 2/37 patients, while H3F3A (K27M) histone mutations were not detected. Interestingly, we identified a GOPC-*ROS1* fusion in an 8-year-old patient whose tumor lacked *BRAF* alterations and histologically classified as low grade glioma. The patient underwent gross total resection (GTR) currently he is disease free. To our knowledge this is the first report of *GOPC-ROS1* fusion in PLGG which may represent a genomically-distinct subgroup of pLGGs that could be targeted with oral target therapy crizotinib. Taken together, we reveal the genetic characteristics of pLGG Saudi patients can enhance diagnostics and therapeutic decisions. In addition, we identified a GOPC-*ROS1* fusion that may be a biomarker for pLGG. Our study proves the possibility of using genetic profiling to guide optimal treatment strategies for pLGG in Saudi population

LGG-02. A BRAIN TUMOR DIAGNOSED AFTER TRANSITION TO THE DEPARTMENT OF ADULT NEUROSURGERY FROM THE DEPARTMENT OF PEDIATRICS

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The patient was a 17-year-old boy with a history of 4 non-febrile convulsions at 15 and 16 years of age. He visited the Department of Pediatrics at a pediatric hospital. An electroencephalogram showed right frontal spike discharge. MRI was performed and judged to show no abnormality. The pediatric doctor diagnosed him with epilepsy. At 17 years old, he was referred to our Department of Adult Neurosurgery for transition. Physical and neurological examinations showed no abnormalities. Brain MRI showed right frontal cortical small tumor, with T1 low, T2 high, diffusion-weighted imaging low, and partial contrast enhancement. We diagnosed him with a brain tumor and symptomatic epilepsy. We surgically removed a right frontal cortical tumor. A pathological examination finalized the diagnosis of dysembryoplastic neuroepithelial tumor. MRI confirmed the total removal of the tumor. Anticonvulsant was started before surgery. No epileptic seizure was observed, so the anticonvulsant medication was gradually tapered and stopped at two years after the surgery. No epilepsy nor recurrence has been observed thus far. The problem with the initial management of this case at the Department of Pediatrics in the pediatric hospital was that the brain tumor was missed despite an MRI examination. Had the transition not happened, this brain tumor might not have been diagnosed. A brain tumor is a rare disease, and epilepsy is a common disease. However, in cases of non-febrile convulsion, a brain tumor should be considered. Collaboration within a single department, hospital and local area should be established.

LGG-03. INCIDENCE AND OUTCOME OF PEDIATRIC IDH-MUTANT GLIOMA

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INTRODUCTION: The incidence of *IDH* mutations in pediatric glioma is unclear. Recent publications suggest rates ranging between 0–20%.