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PURPOSE: Primary spinal low-grade gliomas (LGGs) are rare, can be difficult to treat, and can result in significant morbidity. The management of pediatric spinal LGGs remains controversial. METHODS: A national multi-centre retrospective review of spinal LGGs diagnosed in children less than 18 years of age between 1990–2015 was undertaken to examine the clinical features, pathological subtypes, and treatment outcomes. RESULTS: Forty-three patients from five institutions were included. The median age of diagnosis was 5.2 years. All patients were symptomatic at diagnosis. Forty-four percent of patients were diagnosed at least 6 months after symptoms developed. Two patients had metastatic disease at diagnosis. The most common histology was pilocytic astrocytoma (48.8%). Molecular information was available for 15/43 patients: 6 patients had *BRAF* fusions and 4 patients had *BRAF* V600E mutations. Gross-total resection was achievable in only 6 patients. Twenty-seven patients were treated with surgery-only and the others received chemotherapy and/or focal radiation. Eleven patients were irradiated. No patients were registered in clinical trials for first-line therapy. Twenty-three patients experienced relapse or progression. Patients were followed for a median of 8.3 years (range, 0.5–20.4 years). Five-year progression-free survival (PFS) and overall survival (OS) rates were 48.3% (95% CI, 32.3% to 62.5%) and 89.7% (95% CI, 74.6% to 96.1%) respectively. CONCLUSION: There is significant heterogeneity in surgical outcomes and treatment modalities of pediatric spinal LGGs. The PFS and OS rates remain suboptimal, likely due to tumor location. The low clinical trial enrollment rate highlights the paucity of available trials for spinal LGGs.

LGG-20. CLINICAL FEATURES AND TREATMENT RESULTS FOR PEDIATRIC OPTICO-HYPOTHALAMIC ASTROCYTOMA

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Current consensus for the treatment of optico-hypothalamic astrocytoma (OHA) is a chemotherapy-first policy, limiting the role of surgery for histopathological diagnosis and partial decompression. However, a subgroup of OHA patients show resistance to chemotherapy and have a worse prognosis. In this study, we retrospectively analyzed our clinical experiences of the treatment of patients with OHA in two university hospitals. We have extracted and analyzed the medical charts of 15 pediatric OHA patients treated in two university hospitals since 1990. NF-1-associated OHA patients were excluded. Patient ages ranged from 10 months to 21 years (median 7 years). Out of 15 cases, 12 patients had a tumor larger than 3 cm and classified as Dodge 3. The final histopathological diagnosis was pilocytic astrocytoma in 13 cases. Three patients with tumors classified as Dodge 1 or 2 show good prognosis only by biopsy or partial resection. However, regarding Dodge 3 tumor, patient prognosis is worse regardless of chemotherapy and radiotherapy. After the initial surgery, chemotherapy was administered in 11 cases and radiotherapy in 5 cases. Multiple surgeries are needed for tumor control in 7 patients. Four patients died of tumor progression or treatment-associated complications. When the initial tumor is large enough to cause neurological deterioration, a chemotherapeutic tumor suppressive effect might be limited in a subset of large OHA cases. Therefore, it is important to consider the proper timing of safe surgical decompression in the early phase when a large tumor does not respond to chemotherapy.

LGG-21. MR-GUIDED LASER INTERSTITIAL THERMAL THERAPY FOR UNRESECTABLE AND SYMPTOMATIC PEDIATRIC LOW GRADE GLIOMA

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BACKGROUND: Pediatric low-grade gliomas (LGG) not amenable to resection, while often indolent, represent a significant source of cancer-related morbidity and an unmet therapeutic need. Standardly, these patients are treated with sequential lines of chemotherapy, while delaying as long as possible radiation. Magnetic resonance-guided laser interstitial therapy (LITT) is a minimally invasive procedure that utilizes real-time MR thermography to ablate brain lesions. METHODS: A 15-year-old girl was diagnosed with a suprasellar, hypothalamic LGG, BRAF V600E mutation positive. The tumor was unresectable, and due to progressive vision loss and headaches, the patient underwent treatment. Despite sequential trials of thioguanine/procarbazine/lomustine/vincristine, carboplatin/vincristine, dabrafenib, and combination dabrafenib/trametinib, the patient continued to experience debilitating headaches, malnutrition, school absenteeism, and overall poor quality-of-life. Using real-time, sequential MRI-thermometry and the Neuroblate cooled directional laser catheter, the bulk of the enhancing tumor was heated to a killing temperature. RESULTS: At 1-year post LITT, the patient's symptoms were dramatically improved, including greatly improved headaches, malnutrition, school absenteeism, and overall quality of life. LITT was generally well tolerated, though the patient had slight progressive left homonymous hemianopia, thought secondary to LITT impact on the optic tracts. The tumor progressively shrank over the year post-LITT to a peak of 42% volume reduction. CONCLUSION: We report a case of a pediatric patient with an unresectable low grade glioma who underwent LITT with excellent clinical and radiographic effects. LITT should be considered for children with unresectable and morbid LGGs that fail to respond to more conventional therapies.

LGG-22. EVALUATION OF IMMUNE AND GENOMIC CHARACTERISTICS IN PEDIATRIC OPTIC NERVE GLIOMA (ONG)

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Pediatric optic nerve glioma (ONG) is a rare, sight-threatening tumor. We previously reported clinical, radiologic, histopathologic, and molecular characteristics of pediatric ONG patients treated at Columbia University Medical Center between 2000–2017. Here we evaluate this cohort and one additional patient using quantitative multiple immunofluorescence (qmIF) and next generation sequencing (NGS) using the Columbia Combined Cancer Panel (CCCP). For qmIF, 4 micron immuno-blank slides were stained for CD3, CD8, CD68, CD163, HLA-DR, and Olig2. QmIF images were analyzed and data were processed in R studio and compared based on tumor mutation and treatment history. QmIF failed in 1 case and CCCP failed in 2 cases. CCCP confirmed KIAA1549:BRAF fusions in 2 patients, identified NF1 in 2 patients, and demonstrated both a KIAA1549:BRAF fusion and SETD2 mutation in the added case. Qualitative analysis showed immune infiltrate across cases included macrophages (CD68+, 1.6–6.5% of all cells) and T cells (CD3+, 0.4% to 1.5%). Non-cytotoxic T cells (CD3+CD8-) comprised 60.7–100% of the T cell compartment. There was no difference when comparing mutation groups. However, patients who previously received radiation had increased CD3+, specifically CD3+CD8- cells compared to non-irradiated patients ($p=0.01$ and $p<0.01$, respectively) while CD3+CD8+ and CD68+ cells were not different between groups ($p=0.49$ and $p=0.27$, respectively). In summary, qmIF analysis showed increased tumor infiltration by non-cytotoxic T cells in previously irradiated pediatric ONG patients compared to non-irradiated patients, while there was no difference in macrophages of cytotoxic T cells. This type of analysis may be useful in designing immunotherapeutic strategies for pediatric ONG.

LGG-23. EXCELLENT CLINICAL / RADIOLOGICAL RESPONSE TO BRAF INHIBITION IN A YOUNG CHILD WITH IN-OPERABLE SUPRA-SELLAR PILOCYTIC ASTROCYTOMA

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In-operable low grade gliomas (LGG) in the pediatric population continue to present a treatment dilemma. Due to the low-grade nature of these tumors, and variable response to chemotherapy / radiation, the choice of adjuvant treatment is difficult. Overall survival is directly related to the degree of surgical resection, adding complexity to these inoperable tumors. Current chemotherapeutic regimen for these inoperable tumors includes vincristine (VCR) and carboplatin (Carbo). With advancements in the molecular characterization of gliomas, the role of targeted therapy has come into question. We present a 2-year-old female with biopsy proven Pilocytic Astrocytoma (positive BRAF-V600E mutation) involving the hypothalamic/optic chiasm region. She presented with ataxic gait, bi-temporal hemianopia, obstructive hydrocephalus and central hypothyroidism, which progressed to altered consciousness, and right hemiparesis due to location/mass effect of the tumor. She was initially treated with chemotherapy (VCR/Carbo) but her tumor progressed at 6 weeks of treatment. As her tumor was positive for BRAF-V600E mutation, she was started on Dabrafenib monotherapy, resulting in dramatic improvement in her clinical symptoms (able to stand, improved vision), and a 60% reduction in tumor size at 3-months. At 6-months, follow up MRI showed slight increase in the solid portion of the tumor, with no clinical symptoms. We plan to add MEK inhibitor (Trametinib) and continue with Dabrafenib. Our experience and literature review suggests that LGG with