2%). Among gliomas, 163/239 (68%) had *IDH1/2* mutations, which were significantly more enriched for *TP53* (121/163; 74%) and *ATRX* (90/163; 55%) co-mutations compared to IDH-wild type (IDH-WT) (q<0.0001). In comparison, IDH-WT gliomas (n=76) were enriched for alterations in *BRAF* (16/76 with SNV, 9/76 fusion; q<0.0001), *H3F3A* (14/76; q<0.0001), *PTEN* (10/76; q=0.038), and *CDKN2A* (16/76; q=0.038). Among 113 patients with high-grade gliomas (WHO grade 3/4), those with IDH-WT tumors (n=34) had a poorer prognosis that those with IDH-mutant tumors (n=79) (1 and 5-year OS of 68% and 28% vs. 97% and 76%, respectively) (p<0.0001). Further, a lower frequency of alterations in *TERT* (26/113; 23%), *EGFR* (6/113; 5%), and *PDGFRA* (4/113; 4%) were observed compared to previous studies for adult-only cohorts (~50-70%, 40%, and 10%, respectively). CONCLUSIONS: IDH-mutant gliomas were the most common tumor type seen in this AYA cohort. However, some alterations characteristic of adult high-grade gliomas (*TERT*, *EGFR*, *PDGFRA*) were not as frequently observed. Additional identification of molecular features that predict response to conventional and targeted therapies is underway.

PATH-16. NONINVASIVE DIAGNOSIS OF GLIOMAS THROUGH CSF CFDNA SEQUENCING IN PEDIATRIC AND ADOLESCENT AND YOUNG ADULT (AYA) PATIENTS

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PURPOSE: A subset of pediatric, adolescent and young adult (AYA) gliomas are located in the brainstem, eloquent locations, or present with diffuse/leptomeningeal disease, and are associated with high risk and low yield of biopsy. At the same time, accurate molecular diagnosis is necessary to direct optimal therapy. In such cases, analysis of cell free DNA (cfDNA) form cerebral spinal fluid (CSF) may represent a diagnostic alternative to biopsy. METHODS: We investigated the utility of CSF cfDNA sequencing through a stepwise approach, using clinically validated, targeted molecular assays. Testing was performed using a broad hybrid capture next generation sequencing assay (MSK-IMPACT) and subsequent targeted digital droplet PCR in a subset of cases. RESULTS: We analyzed 17 CSF samples from 17 pediatric (n=6) and AYA (n=11) glioma patients with primary or recurrent disease. Thirteen had tumors located within the brainstem, and four had leptomeningeal involvement. Somatic alterations were detected in 12/17 samples (71%). In 3/4 patients with leptomeningeal involvement, cfDNA testing revealed a BRAF fusion consistent with the diagnosis of diffuse leptomeningeal glioneuronal tumor (DLGNT). Among the 13 patients with brainstem involvement, four had somatic H3 K27M mutations, three had IDH mutations, and one had TP53 and ATRX mutations; five patients had no detectable mutations. CONCLUSION: In our analysis, we found that established glioma hotspot mutations were able to be detected within the CSF. We propose that in patients for whom tissue biopsy is high risk, not feasible, or tissue was nondiagnostic, CSF cfDNA sequencing has a substantial diagnostic yield and should be considered as a valuable novel diagnostic tool. Ongoing research is aiming to further increase the sensitivity of cfDNA testing, especially in patients with very low levels of CSF cfDNA.

PEDIATRIC NEURO-ONCOLOGY IN LOW/MIDDLE INCOME COUNTRIES

LINC-02. GAMMA KNIFE RADIOSURGERY AS AN EFFICACIOUS TREATMENT FOR PEDIATRIC CENTRAL NERVOUS SYSTEM TUMORS: A RETROSPECTIVE STUDY OF 61 NEOPLASMS

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PURPOSE: Brain tumors have an incidence of 1.15 to 5.14 cases per 100,000 children and are associated with significant morbidity and mortality. Radiosurgery has become a promising approach to manage these pediatric CNS tumors. The aim of the present study was to analyse the efficacy of radiosurgery in the treatment of a variety of pediatric tumors of CNS METHODS: This retrospective study was conducted from 1997-2012 at a single Neurosurgery centre. All paediatric patients (≤ 18 years of age) with CNS tumours who were treated with gamma knife radiosurgery (GKRS) and had a minimum follow up of 6 months were included in the study. Patients with lesions other than tumours were excluded. Clinical, radiological and GKRS planning data was collected and analysed in all patients. RESULTS: A total of 76 children with brain tumors had GKRS during the study period. Of these, 40 children (with 61 neoplasms) had follow up available and were included in the study. The mean age was 16 years (6-18 years). 17 patients received primary GKRS, 20 patients received secondary and 3 patients received both. The median tumor volume was 3.3cm3 (0.14 - 38.9 cm3). The mean dose was 12.56 Gy at 50% isodose line. The majority of the tumors were Meningioma (n=20) followed by Acoustic Schwannoma (n=17). The mean treatment time was 67.04 minutes. 33 tumors responded favourably to GKRS, 24 showed a stable size, 3 had no response while 1 progressed requiring surgery. CONCLUSION: GKRS has the potential to become an indispensable tool in the management of pediatric brain neoplasms

LINC-03. A SINGLE CENTER RETROSPECTIVE ANALYSIS OF CURRENT THERAPY AND OUTCOME OF CENTRAL NERVOUS SYSTEM TUMORS IN CHILDREN OF BEIJING

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OBJECTIVE: To explore the clinical characteristics, therapy and outcome in children with central nervous system (CNS) tumors in Beijing, China. METHODS: Clinical data of 1003 patients with newly diagnosed CNS tumors under the age of 15 years admitted to Beijing Shijitan Hospital between January 2017 and December 2021 were retrospectively analyzed. The diagnoses were confirmed by pathology and/or molecular tests. RESULTS: 34 patients were lost to follow up. Medulloblastoma (n=472), ependymoma (n=79), ETMR (n=59) and pinealoblastoma (n=19) were treated according to HIT-2000/2017, 5-year progression-free survival (PFS) and overall survival (OS) were 76.4 ± 2.3%/ 84.8 ±1.9% in MB, 88.8±3.7% /89.5±3.8% in ependymoma, 49.9±7.6%/59.6±10.7% in ETMR, and 3y-PFS/OS of 11.1±10.5%, 90.0±9.5% in pinealoblastoma. The detailed histology of 59 cases of embryonal tumors included 40 cases of CNS embryonal tumor, 9 neuroblastoma, 7 ETMR, and 3 rare tumors (Intracranial mesenchymal tumor with FET-CREB fusion, CNS high-grade neuroepithelial tumor with BCOR alteration, and Melanotic neuroectodermal tumor of infancy). Among 19 cases of pinealoblastoma, two contain DICER1 mutations. Atypical teratoid/rhabdoid tumors (n=36) were treated by EU-RHAB-Protocol with 3y-PFS/OS of 42.2 ± 9.4 % and 62.8 ± 9.5 %. For low-grade gliomas (n=153, 114 cases of OPG and 39 other non-STR/NTR or progressive cases) under SIOP-LGG 2004, 5y-PFS/OS rates were 77.6±5.2% and 92.4±3.2%; Comparatively, among 63 cases of high-grade gliomas, 19 cases survived with median survival of 52 months, 44 cases died. Choroid plexus cancers (n=13) were treated according to CPT-SIOP-2009 with 5y-PFS /OS of 68.4 ± 13.1 % and 64.8±14.3 %. Among 109 germ cell tumors, 5y-PFS/OS were 93.8±2.8% and 95.5±2.2%, respectively. CONCLUSION: The prognosis of CNS tumors are related to pathology, molecular type and individualized therapy. By international multicenter cooperation and adopting advanced protocols, the survival rates of pediatric CNS tumors can be significantly improved.

LINC-04. OPTIC PATHWAY GLIOMAS IN CHILDREN: AN 18-YEAR EXPERIENCE IN A MIDDLE-INCOME COUNTRY

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BACKGROUND: Optic pathway gliomas (OPGs) have favorable survival outcomes, but can have significant morbidity, from the tumor or from its treatment. This retrospective study sought to describe the clinical presentation and outcomes of children with OPGs at a tertiary center in a middle-income country. METHODS: Patients younger than 18 years diagnosed with OPGs between 2002-2020 at the Hospital Civil de Guadalajara in Mexico, were included. Survival analyses and logistic regression were used to determine risk factors for progression. RESULTS: Thirty patients were identified with a median age of six years. Common clinical characteristics at presentation included: visual accuracy impairment (40%), headache (23.3%), nystagmus (20%), endocrine disturbances (16.6%), and diencephalic syndrome (13%). Neurofibromatosis-1 was present in 23.3% of the patients. Eight patients underwent resection at diagnosis, 7 of them had secondary complications. One died during surgery due to catecholamine-resistant shock, and the other developed neuro-infection and died due to septic shock after surgery. Five children developed neurological or endocrine sequelae. Fifteen total patients received chemotherapy as their first line treatment. The 5-year progression-free survival was 77.9±10% and overall survival was 89.5±5%. Patients who received chemotherapy or targeted therapy as the first line of treatment had higher PFS compared to those who received other therapies (p=0.03). The patients who received surgery as first-line treatment had a 3.1