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BACKGROUND: Considering that paediatric high-grade gliomas (HGGs) are biologically distinct from their adult counterparts, the objective of this study was to define the landscape of HGGs in adolescents and young adults (AYAs). METHODS: We performed a multicentric retrospective study of 112 AYAs from adult and paediatric Ile-de-France neurosurgical units, treated between 1998 and 2013 to analyse their clinicoradiological and histomolecular profiles. The inclusion criteria were age between 15 and 25-years, histopathological HGG diagnosis, available clinical data, pre-operative and follow-up MRI. MRI and tumoral samples were centrally reviewed. Immunohistochemistry and complementary molecular techniques such as targeted/next generation sequencing, whole exome sequencing and DNA-methylation analyses were performed to achieve an integrated diagnosis according to the 2016 WHO classification. RESULTS: Based on 80 documented AYA patients, HGGs constitute heterogeneous clinicopathological and molecular groups, with a predominant representation of paediatric-subtypes (Histone H3-mutants, 40%) but also adult-subtypes (*IDH*-mutants, 28%) characterized by the rarity of oligodendrogliomas, *IDH*-mutant and 1p/19q co-deleted and the relative high frequency of "rare adult *IDH* mutations" (20%). H3G34-mutants (14%) represent the most specific subgroup in AYAs. In the H3K27-mutant subgroup, the non-brainstem diffuse midline gliomas are more frequent (66.7%) than diffuse intrinsic pontine gliomas (23.8%), contrary to children. We found that WHO grade has no prognostic value, but molecular subgrouping has major prognostic importance. CONCLUSIONS: HGGs in AYAs could benefit from a more personalized neuro-oncological management, driven by molecular subtyping rather than age group. Collaborative efforts are needed from paediatric and adult neuro-oncology teams to improve the management of HGGs in AYAs.

HGG-12. A CASE OF PEDIATRIC SPINAL HIGH-GRADE GLIOMA WITH NTRK1 GENE FUSION

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INTRODUCTION: Tumors arising from the spinal cord are uncommon, especially high-grade tumors in pediatric patients. We report a case of high-grade glioma in the spinal cord harboring *NTRK1* gene fusion, who received effective entrectinib therapy. CASE REPORT: A 5-year-old boy presented right hemiparesis and MR imaging revealed an intramedullary enhancing mass at the vertebral body level between C3 and Th1. He underwent microsurgical partial resection and the histological diagnosis was low-grade astrocytoma. After the first-line chemotherapy with vincristine and carboplatin, his right hemiparesis deteriorated and recurrent MR imaging showed growth of the tumor. He underwent microsurgical partial resection again and the histological examination was high-grade glioma with endothelial proliferation and necrosis. The chemoradiotherapy with temozolomide and focal irradiation of 50.4 Gy were given, and his neurological symptom slightly improved. One month later, he presented respiratory disturbance and required assisted ventilation with tracheostomy. MR imaging showed tumor progression invading upward to medulla oblongata. *NTRK1* gene fusion was detected in the previous surgical specimen by a gene panel testing, and he received entrectinib, a potent inhibitor of tropomyosin receptor kinase (TRK). Since then, no tumor progression has been demonstrated for several months by MRI and he has been stable neurologically. CONCLUSION: High-grade spinal cord tumors are rare and effective treatment strategies have not been addressed. Although the frequency of the gene fusion is very low in pediatric gliomas, identification of the driver gene aberration like in this case by a gene panel can provide potential targeted therapies for selected patients.

HGG-14. TREATMENT AND PROGNOSTIC FACTORS FOR PEDIATRIC GLIOBLASTOMAS--THE 10 YEARS EXPERIENCE FROM ONE SINGLE CENTER

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OBJECTIVE: We retrospectively analyzed the clinical features of pediatric glioblastoma patients in our center in the past 10 years. METHODS: From November 2009 to December 2018, patients with glioblastoma under 18 years were admitted to Guangdong Sanjiu Brain Hospital. Clinical and pathological features were summarized, and the curative effect was evaluated. RESULTS: A total of 31 pediatric patients were enrolled. The median age is 13.8 years (range 0.8–18), including 19 males and 12 females. To Sep, 2019, the median follow-up time was 18 months(Range 4–80 months). Among them,2 were lost to follow-up,13 died, 16 still survived, and the longest survivor survived for 80 months. The median survival time was 16.4 months, the 2-year survival rate was 38%. In the prognostic factor analysis, the median survival time of patients with surgical resection ≥90% was 18 months (95% CI 15.9–20 months), and for children with resection 90% was 11 months (95% CI 9.9–12 months), P=0.027, with significantly statistically difference. Multivariate analysis showed that tumor resection rate was an independent prognostic factor for survival. CONCLUSION: The prognosis of pediatric glioblastoma is still dismal. This study demonstrates that prognosis of such patients with GTR or near GTR is better.

HGG-16. EXOSOME-MEDIATED INTER-CLONAL INTERACTIONS IN PEDIATRIC GBM AND DIPG

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Pediatric Glioblastoma (pGBM) and Diffuse Intrinsic Pontine Glioma (DIPG) are highly heterogeneous brain tumors which we demonstrated are comprised by distinct sub-clones interacting in a functional network. Exosomes are known to mediate the crosstalk between tumor and its microenvironment. Based on this, we aimed to investigate the role of exosomes in mediating pGBM and DIPG inter-clonal communication. By using *optical barcoding* for single cell-tracking, we generated two bulk multicolor patient derived-cell lines (one DIPG H3.3K27M and one pGBM histone WT) from which we obtained two and five single cell-derived clones respectively. The sub-clones demonstrated significantly phenotypic differences in terms of morphology, growth, adhesion, migration and invasion properties. In particular, co-culture experiments, with the two most different clones for both cell-lines, confirmed the cell-cell interaction key role in driving their more aggressive phenotype. Furthermore, we found that pGBM and DIPG subclones release exosomes which are actively and differentially up-taken by individual clones. Analysis of the exosomal microRNAs showed a different profile between the two selected clones in each cell-line. In particular, we found a pool of five upregulated microRNAs in 1C5 clone (DIPG cell-line) strongly associated to Wnt-signaling and PI3K-AKT pathway. Similarly, a pool of five upregulated microRNAs for 5E2 clone (pGBM cell-line) were found associated with focal adhesion and PI3K-AKT pathway. Our study may provide novel therapeutic strategies by interfering with the exosome-mediated inter-clonal communication in pGBM and DIPG.

HGG-17. HIGH-GRADE GLIOMA IN VERY YOUNG CHILDREN; A SINGLE-CENTER 11-YEAR-EXPERIENCE

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