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% (136/357) of cases, averaging 2.5 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 AFRF1-HTR2K (2/37) and TRX-PJRCA (2/37) fusions that were observed at much lower frequencies. The most frequently mutated genes were NOTCH3 (7/37), ATM (4/37), RASD1C (3/37), NFI1 (3/37), SLX4 (3/37) and NF1 (3/37). BRAF V600E mutations were observed in only 25 patients, while H3F1A (K27M) mutations were detected in 10 patients. Interestingly, we identified a GOPC-ROSI 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-ROSI 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 diagnostic and therapeutic decision addition, we identified a GOPC-ROSI 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 in 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 neuropsychological examinations showed no abnormalities. Brain MRI showed right fronto-cortical small tumor, with T1 low, T2 high, diffusion-weighting 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 finally diagnosed 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. Interestingly, we identified a GOPC-ROSI 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-ROSI 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 diagnostic and therapeutic decision addition, we identified a GOPC-ROSI 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.
LG3-03. MOLECULAR GUIDED THERAPY FOR A PEDIATRIC LOW GRADE GLIOMA: A CASE REPORT
Jaying VonBergen, Beth Armstrong, and Morgan Schmitt; Riley Hospital for Children, Indianapolis, IN, USA

Low grade gliomas are the most common type of central nervous system tumors among children. Despite the fact that they are not typically life threatening, low grade gliomas remain a significant clinical challenge. Case Study: Patient is a 4-year-old male who presented at 20 months of age with several weeks of ataxia, emesis, and head lift. Imaging revealed a right temporal lobe lesion; he was subsequently taken to surgery, where a gross total resection was achieved. Imaging 9 months post resection revealed recurrent disease within the right temporal region with leptomeningeal involvement. Rapid progression of disease led to referral for molecular guided therapy. This case illustrates a clear benefit of using molecular guided therapy in low grade gliomas.

LG3-04. A PHASE II RE-TREATMENT STUDY OF SELUMETINIB FOR RECURRENT OR PROGRESSIVE PEDIATRIC LOW-GRADE GLIOMA (PGLG): A PEDIATRIC BRAIN TUMOR CONSORTIUM (PTBC) STUDY
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The PBTC conducted a re-treatment study (NCT01089101) evaluating selumetinib. Eligible patients must have maintained stable disease (SD) or progressive disease (PD) on prior RT or chemotherapy. Patients were evaluable for outcomes with median follow-up of five years. Five-year radiation-free, progression-free and overall survival for patients with low-grade histology were 76% and 100%, respectively. One patient with high-grade glioma recurred 1.2 years after upfront chemoradiation and died soon after recurrence. CONCLUSION: IDH-mutant gliomas comprise a small proportion of pediatric gliomas. Incidence rate is higher in the second decade of life. Comparative analyses between pediatric IDH-mutant gliomas and adult historical cohorts are currently underway, evaluating outcomes, radiation therapy and frequency of malignant transformation.

LG3-05. MOLECULAR GUIDED THERAPY FOR A PEDIATRIC LOW GRADE GLIOMA: A CASE REPORT
Jaying VonBergen, Beth Armstrong, and Morgan Schmitt; Riley Hospital for Children, Indianapolis, IN, USA

Low grade gliomas are the most common type of central nervous system tumors among children. Despite the fact that they are not typically life threatening, low grade gliomas remain a significant clinical challenge. Case Study: Patient is a 4-year-old male who presented at 20 months of age with several weeks of ataxia, emesis, and head lift. Imaging revealed a right temporal lobe lesion; he was subsequently taken to surgery, where a gross total resection was achieved. Imaging 9 months post resection revealed recurrent disease within the right temporal region with leptomeningeal involvement. Rapid progression of disease led to referral for molecular guided therapy. This case illustrates a clear benefit of using molecular guided therapy in low grade gliomas.