are defined. Unique recommendations for DIPG response include an eight-week response duration, a twenty-five percent decrease for partial response, and the distinction of pontine and extra-pontine response for trials that use focal drug delivery. The recommendations presented here represent an initial effort to uniformly collect and evaluate response assessment criteria; these recommendations can now be incorporated into clinical trials to assess feasibility and corroboration with patient outcomes.

IMG-10. MRI-BASED RADIOMIC PROGNOSTIC MARKERS OF DIFFUSE MIDLINE GLIOMA

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BACKGROUND: Diffuse midline gliomas (DMG) are lethal pediatric brain tumors with dismal prognoses. Presently, MRI is the mainstay of disease diagnosis and surveillance. We aimed to identify prognostic image-based radiomics markers of DMG and compare its performance to clinical variables at presentation. METHODS: 104 treatment-naïve DMG MRIs from five centers were used (median age=6.5yrs; 18 males, median OS=11mos). We isolated tumor volumes of T1-post-contrast (T1gad) and T2-weighted (T2) MRI for PyRadiomics high-dimensional feature extraction. 900 features were extracted on each image, including first order statistics, 2D/3D Shape, Gray Level Co-occurrence Matrix, Gray Level Run Length Matrix, Gray Level Size Zone Matrix, Neighboring Gray tone Difference Matrix, and Gray Level Dependence Matrix, as defined by Imaging Biomarker Standardization Initiative. Overall survival (OS) served as outcome. 10-fold cross-validation of LASSO Cox regression was used to predict OS. We analyzed model performance using clinical variable (age at diagnosis and sex) only, radiomics only, and radiomics plus clinical variable. Concordance metric was used to assess the Cox model. RESULTS: Nine radiomic features were selected from T1gad (2 texture wavelet) and T2 (5 first-order features (1 original, 4 wavelet), 2 texture features (1 wavelet, 1 log-sigma). This model demonstrated significantly higher performance than a clinical model alone (C: 0.68 vs 0.59, p<0.001). Adding clinical features to radiomic features slightly improved prediction, but was not significant (C=0.70, p=0.06). CONCLUSION: Our pilot study shows a potential role for MRI-based radiomics and machine learning for DMG risk stratification and as image-based biomarkers for clinical therapy trials.

IMG-12. CHARACTERISATION OF MODELS OF *H3F3A*_G34R/V MUTANT PAEDIATRIC GLIOBLASTOMA *IN VIVO* USING MAGNETIC RESONANCE IMAGING

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Approximately 15% of paediatric/young adult cerebral hemispheric glioblastomas (pGBM) harbour G34R/V mutations in *H3F3A*, encoding the histone H3.3 variant. Development of novel therapeutic interventions demands models that accurately recapitulate this subset of disease and sensitive imaging methods with which to study tumours *in situ*. Three *H3F3A*_G34R primary-patient-derived cultures, alongside established cell-line KNS42 (*H3F3A*_G34V), were implanted orthotopically in immunocompromised mice. KNS42 (*TP53*_R342*) tumours were clearly detectable using T_2-weighted (T_2w)-MRI, enhanced following contrast agent administration, indicating impaired blood-brain barrier (BBB) integrity, and demonstrated minimal invasion. OPBG_GBM_001 cells (*TP53*_89-90X,*ATRX*_II2133-2144X) formed infiltrative tumours that were hyperintense on T_2w-MRI and demonstrated contrast-enhancement suggestive of heterogeneous BBB integrity. HSJD_GBM_002 cells (*TP53*_P278T,*ATRX*_R666*) spread diffusely throughout the brain with their full extent typically not discernible by T_2w-MRI, the BBB also remaining intact. No evidence of CHOP_GBM_001 tumour was detected by MRI 11months post-implantation. Immunocompetent syngeneic models using tumour cells induced by mutations modelling hemispheric pGBM (NRAS/shP53/shATRX±H3.3G34R) are being explored. Fast growing heterogeneous lesions with variable contrast-enhancement were identified; the H3.3G34R mutation conferred longer median survival (2 clones:25/28days, control:14days). These models have the advantage of an intact immune system and short latency for initial efficacy studies. Primary pGBM cells yield tumours that are more representative of the spectrum of clinical disease; variable hyperintensity on T_2w-MRI corresponding to cellular density, with diffusely infiltrative disease less clearly definable, a paucity of oedema and a range of contrast-enhancement. Pathological features including giant multinucleated cells, and mitotic figures were also evident.

IMG-13. MRI-BASED RADIOMICS PROGNOSTIC MARKERS OF POSTERIOR FOSSA EPENDYMOMA

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PURPOSE: Posterior fossa ependymomas (PFE) are common pediatric brain tumors often assessed with MRI before surgery. Advanced radiomic analysis show promise in stratifying risk and outcome in other pediatric brain tumors. Here, we extracted high-dimensional MRI features to identify prognostic, image-based, radiomics markers of PFE and compared its performance to clinical variables. METHODS: 93 children from five centers (median age=3.3yrs; 59 males; mean PFS=50mos) were included. Tumor volumes were manually contoured on T1-post contrast and T2-weighted MRI for PyRadiomics feature extraction. Features include first-order statistics, size, shape, and texture metrics calculated on the original, log-sigma, and wavelet transformed images. Progression free survival (PFS) served as outcome. 10-fold cross-validation of a LASSO Cox regression was used to predict PFS. Model performance was analyzed and concordance metric (C) was determined using clinical variable (age at diagnosis and sex) only, radiomics only, and radiomics plus clinical variable. RESULTS: Six radiomic features were selected (all T1): 1 first-order kurtosis (log-sigma) and 5 texture features (3 wavelet, 2 original). This model demonstrated significantly higher performance than a clinical model alone (C: 0.69 vs 0.58, p<0.001). Adding clinical features to the radiomic features didn't improve prediction (p=0.67). For patients with molecular subtyping (n=48), adding this feature to the clinical plus radiomics models significantly improved performance over clinical features alone (C = 0.79 vs. 0.66, p=0.02). Further validation and model refinement with additional datasets are ongoing. CONCLUSION: Our pilot study shows potential role for MRI-based radiomics and machine learning for PFE risk stratification and as radiographic biomarkers.

IMG-14. DEVELOPING A PREDICTIVE GRADING MODEL FOR CHILDREN WITH GLIOMAS BASED ON DIFFUSION KURTOSIS IMAGING METRICS: ACCURACY AND CLINICAL CORRELATIONS WITH SURVIVAL

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PURPOSE: To develop a predictive grading model based on diffusion kurtosis imaging (DKI) metrics in children affected by gliomas, and to investigate the clinical impact of the model via correlations with overall survival and progression-free survival. MATERIALS AND METHODS: We retrospectively studied 59 children (33M, 26F, median age 7.2 years) affected by gliomas on a 3T magnet. Patients with tumor locations other than infratentorial midline were included. Conventional and DKI sequences were obtained. Mean kurtosis (MK), axial kurtosis (AK), radial kurtosis (RK), fractional anisotropy (FA) and apparent diffusion coefficient (ADC) maps were obtained. Whole tumor volumes (VOIs) were segmented semiautomatically. Mean DKI values were calculated for each metric. The quantitative values from DKI-derived metrics were used to develop a predictive grading model with penalized logistic regression (glmnet package, R). Elasticnet regularization was used to avoid model overfitting. Fitted model coefficients from each metric were used to develop a probability prediction of a high-grade glioma (HGG). Grading accuracy of the resulting probabilities was tested with ROC analysis. Finally, model predictions were correlated to progression-free survival (PFS) with a Kaplan-Meier analysis. RESULTS: The cohort included 46 patients with low-grade gliomas (LGG) and 13 patients with HGG. The developed model predictions yielded an AUC of 0.946 (95%CI: 0.890–1). Model predictions were significantly correlated with PFS (23.1 months for HGG vs 34.7 months for LGG, p<0.004). CONCLUSION: In our cohort, a DKI-based predictive model was highly accurate for pediatric glioma