ETMR-22. TITLE: DEFINING THE CLINICAL AND PROGNOSTIC LANDSCAPE OF EMBRYONAL TUMORS WITH MULTI-LAYERED ROSETTES (ETMRS), A RARE BRAIN TUMOR REGISTRY (RBTC) STUDY

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ETMR, an aggressive disease characterised by *C19MC* alterations, were previously categorised as various histologic diagnoses. The clinical spectrum and impact of conventional multi-modal therapy on this new WHO diagnostic category remains poorly understood as a majority of ~200 cases reported to date lack molecular confirmation. We undertook comprehensive clinico-pathologic studies of a large molecularly confirmed cohort to improve disease recognition and treatment approaches. Amongst 623 CNS-PNETs patients enrolled in the RBTC registry, 159 primary ETMRs were confirmed based on a combination of FISH (125), methylation analysis (88), SNP and RNAseq (32) analyses; 91% had *C19MC* amplification/gains/fusions, 9% lacked C19MC alterations but had global methylation features of ETMR NOS. ETMRs arose in young patients (median age 26 months) predominantly as localized disease (M0-72%, M2-3 -18%) at multiple locations including cerebrum (60%) cerebellum (18%), midline structures (6%); notably 10% were brainstem primaries mimicking DIPG. Uni-and multivariate analyses of clinical and treatment details of curative regimens available for 110 patients identified metastatic disease (p=0.002), brainstem locations(p=0.005), extent of surgery, receipt of multi-modal therapy including high dose chemotherapy and radiation (P<0.001) as significant treatment prognosticators, while C19MC status, age and gender were non-significant risk factors. Analyses of events in all patients showed respective EFS at 3 and 12 months of 84%(95%CI:77–91) and 37%(95%CI:20–41) and 4yr OS of 27%(95%CI:18–37) indicating despite intensified therapies ETMR is a rapidly progressive and fatal disease. Our comprehensive data on the largest cohort of molecularly-confirmed ETMRs provides a critical framework to guide current clinical management and development of clinical trials.

GERM CELL TUMORS

GCT-02. THE LONG-TERM OUTCOMES AND SEQUELAE ANALYSIS OF INTRACRANIAL GERMINOMA FROM 187 PATIENTS IN THE SINGLE INSTITUTE: NECESSITY FOR THE ADAPTATION OF RADIOTHERAPY DOSE AND VOLUME

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PURPOSE: We aimed to refine the radiotherapy (RT) volume and dose determinant for disease failures and long-term sequelae in the intracranial germinoma. METHODS: The main treatment for intracranial germinoma was craniospinal RT only (n=51) during 1981–1992 and RT with upfront chemotherapy (CRT) (n=152) during 1992–2015 in Seoul National Uni-