expression (15% staining) was noted in 11 cases (31%). Another 11 cases (30%) showed absent PD-L1 expression. PD-1 expression (>1 cell/high-power field) was seen in 10/12 tumors (83%) without correlation with PD-L1. TMB of 5 mutations per megabase (mt/MB) was seen in all 30 tumors tested with the 592-gene panel, with 27% (n=8) exhibiting high TMB (17 mt/MB). No samples had high level MSI. Overall, 18/30 tumors (60%) had high PD-L1 or high TMB. Mutations in 25 genes were seen; the most frequent were MYD88 (26/30, 87%, all L265P), PIM1 (16/30, 60%), and CD79B (15/30, 50%, all Y196). Among 7 cases tested with RNA-sequencing, one recently reported ETV6-IGH fusion was found (Neuro-Oncology 2018;PMID:29432597). CONCLUSIONS: We found high TMB and PD-L1 expression in PCNSL with no correlation. Nearly two thirds of PCNSL tumors harbor favorable biomarker profiles with anticipated responsiveness to checkpoint inhibitors. Further studies are needed to validate initial results.

RARE-23. PRIMARY INTRA-AXIAL CENTRAL NERVOUS SYSTEM INFLAMMATORY MYOFIBROBLASTIC TUMOR, ALK NEGATIVE: A RARE ENTITY

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Inflammatory myofibroblastic tumor (IMT) is a rare spindle cell neoplasm with admixed inflammatory cells which occurs mainly in the abdomen or thoracic cavity of children or young adults. Primary CNS IMT is exceedingly rare with roughly 100 reported cases in the world literature, most of which are extra-axial and occur as a dural-based mass. Herein, we describe a rare case of intra-axial primary CNS IMT. A 24 year-old healthy female presented to the ER after falling and striking her head. A CT scan revealed an acute intraparenchymal hemorrhage, as well as a mass within the right frontoparietal lobe. Subsequent MRI was performed further characterizing the lesion as a 3.0 cm intra-axial tumor. Craniotomy was performed displaying a circumscribed neoplasm with relatively bland spindle cells arranged in fascicles with an admixed lymphoplasmacytic infiltrate. Mitotic activity was present but limited. Immunohistochemistry (IHC) was positive for TLE1 and vimentin but negative for GFAP, ALK, SMA, MUC4, KIT and β-catenin. Additional molecular testing by FISH for *ALK* (2p23) rearrangement was negative. We report a rare case of intra-axial primary CNS ALK Negative IMT. Approximately half of all IMTs harbor a clonal translocation that activates the anaplastic lymphoma kinase (ALK)-receptor tyrosine kinase at the 2p23 locus. As a result, *ALK* is overexpressed and can be detected by IHC or via molecular diagnostics (FISH, RNA sequencing or RT-PCR). Since this case was processed, additional novel anomalies involving rearrangements in *ROS1*, *RET*, *ETV6* and/or *NTRK3* genes have been described and could lead to promising therapeutic targets in the future.

RARE-24. OBJECTIVE RESPONSE AND CLINICAL BENEFIT IN RECURRENT EPENDYMOMA IN ADULTS: FINAL REPORT OF CERN 08-02: A PHASE II STUDY OF DOSE-DENSE TEMOZOLOMIDE AND LAPATINIB

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BACKGROUND: Ependymoma is a rare tumor for which the role of chemotherapy has not been established either for newly diagnosed or recurrent disease. We report on the first prospective adult clinical trial of chemotherapy for recurrent ependymoma. METHODS: Adult (> 18 years) were treated with dose-dense temozolomide (125–150 mg/m2, 7days on/7days off) and daily lapatinib (1250 mg/day). Efficacy was based on serial imaging of either brain or spine using MRI. Clinical benefit was evaluated using longitudinal assessment of symptom burden (MDASI-BT/MDASI-Spine) and performance status. Primary endpoint was progression free survival. Additional endpoints included 12-month PFS rate, objective response and clinical benefit (KPS and changes in proportion of moderate-severe disease-related symptom burden through cycle 6). RESULTS: 50 patients were treated with this regimen. Median age = 43.5, median KPS = 90, median number of prior relapse = 2. By tumor grade: III n = 19, II n = 16, I n = 8. 25 patients had spinal cord, 15 had supratentorial and 8 had infratentorial tumors. Treatment was well tolerated with a 10% Grade 3–4 myelotoxicity rate. Median PFS = 0.65 years (95%CI 0.46, 1.02). 1-year PFS=38%, with 1 complete and 4 partial responses. KPS improved in all but 1 patient and MDASI measures were completed on 86% of cases with reduction in moderate-severe pain in 53% and 71% of brain and spine patients respectively. Patients with spine tumors reported improvement in numbness (57%), weakness (24%), fatigue (23%)and autonomic dysfunction (bladder control (73%), sexual function (9%), and bowel function (36%)). CONCLUSIONS: These results demonstrate evidence of clinical activity, including objective responses and a nearly 40% stable disease rate at one year, with improvement in disease-related symptoms. This combination regimen was well tolerated and should be considered as a standard salvage regimen for adult patients with recurrent ependymoma.

RARE-26. MUTATIONS IN MAPK PATHWAY GENES ARE CHARACTERISTIC AND CONFIRMATORY OF MULTINODULAR AND VACUOLATING NEURONAL TUMOR OF THE CEREBRUM

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BACKGROUND: Multinodular and vacuolating neuronal tumor of the cerebrum (MVNT) is a rare, recently described neoplasm in the 2016 World Health Organization (WHO) classification with 14 cases reported to date. MVNTs usually present with epilepsy or are discovered incidentally. Median age of diagnosis is 44.9 years (22 to 71 years), and the most common location is the temporal lobe (~65%). METHODS/RESULTS: After reviewing 282 patients enrolled in our natural history study between September 2016 and August 2017, we uncovered two additional cases: the first patient is a 39-year-old woman and the other is a 57-year-old man. The tumors were localized in the left temporal and left frontal lobes respectively. Gross total resection was performed in both cases. Histological features were consistent with the 2016 WHO classification and other published reports. A CNS specific molecular panel revealed a BRAF p.Leu597Arg missense mutation in our first patient, a mutation that was recently reported in one other MVNT but has not been detected in any other primary CNS neoplasm within our database. The second patients lesion was found to have MAP2K1 p.Gln56Pro missense mutation. Our literature review found 2 cases with BRAF and 5 cases with MAP2K1 mutations in MVNT. MAPK pathway alterations reported in other glioneuronal tumors include BRAF p.Val600Glu mutations as well as BRAF fusions, neither of which have been reported in MVNTs to date. CONCLUSIONS: MVNT while rare, will likely be increasingly recognized. Our results suggest that the BRAF p.Leu597Arg missense mutation, (now described in two cases) and the MAP2K1 p. Gln56Pro missense mutation, may be useful diagnostic adjuncts to histopathological features and in differentiating this entity from more commonly epilepsy associated glioneuronal tumors.

RARE-27. CHIMERIC SPINAL CORD GLIOPROLIFERATIVE LESION FOLLOWING INTRATHECAL FETAL STEM CELL INFUSION

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BACKGROUND: Commercial stem cell therapy is available in select practices, particularly at non-U.S. sites, and is increasingly sought as a therapy by patients who are willing to travel for off-label treatment. To date, two cases of a spinal cord "glioneuronal lesion" or "glioproliferative lesion" have been reported. We report the third case, which by microdissection showed the glioproliferative spinal cord lesion to have mixed host/donor origin. CASE: A 73- year-old man presented with 3–4 years of progressive lower extremity weakness; he received intrathecal stem cell therapy (at least partly fetal in origin) in Russia, 7/2016. Symptoms began about 1 month after infusion. At presentation to our clinic in 11/17, neuroimaging showed nerve root enlargement with mild enhancement and clumping of nerve roots. Cerebrospinal fluid demonstrated significant elevation of protein (>1500mg/dL) and low glucose (32mg/dL). Multiple CSF samples were negative for malignancy or clonality. Extensive laboratory work-up for malignancy and inflammatory processes was negative. However, PET-CT showed intense uptake along the nerve roots of the cauda equina. Nerve root biopsy on 1/4/2018 showed a glioproliferative lesion with moderate cell density and