INTRODUCTION: The efficacy of chemotherapy in recurrent ependymoma is unclear. We present results from the German HIT-REZ-studies. METHODS: 137 patients were analyzed regarding the treatment with chemotherapy at first recurrence, the time to progression or response (PFS) and to either time-point of death or last follow-up (OS). Tumor response evaluation was based on MRI and clinically; molecular data was available in 80. RESULTS: In our cohort, 96 patients (20 supratentorial, 73 infants <12 months) received chemotherapy during first recurrence: 89% (51.0%) temozolomide (TMZ) monotherapy, 12 (12.5%) HIT-SKK regimen. 9 (9.4%) carbotinib/etoposide (CE) and 26 (27.1%) other combinations. In 19.8% (26.3% in TMZ), chemotherapy was administered prior to surgery (chemoradiation). Overall, we observed progression in 78% (50% in TMZ). Total-gross resection was achieved in 86% without neoadjuvant chemotherapy and in 74% (69% in TMZ) with neoadjuvant treatment. Switching to temozolomide/etoposide (TE) after surgery and unresponsiveness to TMZ showed further progression in all cases of tumor-residuum after surgery. Switching to temozolomide treatment (1 year-PFS, treatment with HIT-SKK (50.0%-14.4%) or CE (55.6%-16.6%) was advantageous over TMZ (30.2%-67.7%). However, 5-year-OS was lower in CE (19.0%-16.8%) than in TMZ (39.8%-77.7%). Long-term control was seen in individual cases of TMZ, HIT-SKK and CE, with TMZ providing longest response of 72 months. CONCLUSION: Neoadjuvant TMZ has no significant advantage regarding PFS. However, in few cases chemotherapy prevented progression after incomplete resection. Difficulties in response evaluation and variability in therapies hinder conclusions. Supported by the German Children’s Cancer Foundation.

INTRODUCTION: Spinal myxopapillary ependymoma (MPE) is a rare pediatric tumor with a high frequency of local recurrence, even in patients previously treated for ependymoma who develop local findings even in the absence of CNS relapse. Salvage therapy with curative intent should be considered using a multimodal approach.

INTRODUCTION: Spinal myxopapillary ependymoma (MPE) is a rare histological variant of ependymoma classified as WHO grade 1 tumor. Further interrogation of the molecular and clinical profile is warranted.
Abstract

To better understand the biology and clinical phenotype. We summarize our institutional experience with spinal MPE including methylation-profiling. METHODS: A retrospective analysis of charts during the period of 2001 to 2019 of histologically proven MPE was done. We performed methylation profiling for 12 patients by Infinium MethylationEPIC Kit. RESULTS: 26 patients with spinal MPE were identified; median age of diagnosis was 34.2 years with a range of 11 to 59.9 years. Ten patients were below 30 years of age. Concomitant for radio-sensitization in 2 week intervals for a total of 6 weeks. Serial imaging after irradiation revealed decreased tumor burden within 18 months. MRI at 18 months exhibited mild interval growth of 2 lesions. CONCLUSIONS: To our knowledge, this is the first report of a clinical response in a pediatric patient with PF-EPN-A following irradiation administration concurrently with vorinostat therapy. This response highlights the importance of further studies evaluating this combination therapy and its potential use in this population.

EPEN-11. ONGOING RESPONSE IN A MULTIPLY RELAPSED METASTATIC POSTERIOR FOSSA EPNOMA A AFTER VORINOSTAT AND CONCOMITANT IRRADIATION

Hamza S. Quresh,1,2 Stephanie Toll,1,2, and Maxim Yankelevich1; 1Children’s Hospital of Michigan, Detroit, MI, USA; 2Central Michigan University, Mount Pleasant, MI, USA. Tumors and recurrent disease in the posterior fossa ependymoma A (PF-EPN-A) constitutes the worst prognosis. These tumors often relapse despite aggressive resection and irradiation, resulting in limited therapeutic options. Although the genomic profile of PF-EPN-A does not typically show any recurrent alterations; they demonstrate distinctive genetic changes which can be targeted with modulators such as histone deacetylase (HDAC) inhibitors. These inhibitors have shown efficacy in pre-clinical studies in both their anticancer and radiosensitizing properties. CASE: We describe a male diagnosed with a posterior fossa ependymoma at 3 years of age. After initial therapy with resection and focal irradiation, he went on to have a number of recurrences requiring multimodal therapy. Most recently, he developed diffuse intraventricular and leptomeningeal disease not amenable to surgical intervention. Genomic evaluation demonstrated a BCOR mutation and methylation profile was consistent with PF-EPN-A. He received 23.4 Gray craniospinal irradiation with a 30.6 Gray boost to the nodal lesions. Vorinostat was given concomitantly for radio-sensitization in 2 week intervals for a total of 6 weeks. Serial imaging after irradiation revealed decreased tumor burden within 18 months. MRI at 18 months exhibited mild interval growth of 2 lesions. To our knowledge, this is the first report of a clinical response in a pediatric patient with PF-EPN-A following irradiation administration concurrently with vorinostat therapy. This response highlights the importance of further studies evaluating this combination therapy and its potential use in this population.