in 6 of 7 the samples (RYR1, SNX25, DSC2). Strikingly, there was one gene, ADGRL3, which was found to be consistently mutated in the entire disease progression process. Our findings therefore suggest that ADGRL3 might play roles in the disease progression of ependymoma patient.

EPEN-06. CHEMOTHERAPY OF RECURRENT EPENDYMOMA: LONG-TERM RESPONSE ONLY IN FEW CASES

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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 from first relapse to progression (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 infratentorial, 3 spinal) received chemotherapy during first recurrence: 49 (51.0%) temozolomide (TMZ) monotherapy, 12 (12.5%) HIT-SKK regime, 9 (9.4%) carboplatin/etoposide (CE) and 26 (27.1%) other combinations. In 19.8% (26.5% in TMZ), chemotherapy was administered prior to surgery (neoadjuvant), which resulted in tumor progression in 78% (85% in TMZ). Gross-total resection was achieved in 86% without neoadjuvant chemotherapy and in 74% (69% in TMZ) with neoadjuvant treatment. Switching to trofosfamide/etoposide (TE) after surgery and unresponsiveness to TMZ showed further progression in all cases of tumor-residuum after surgery. Regarding 1-year-PFS, treatment with HIT-SKK (50.0%±14.4%) or CE (55.6%±16.6%) was advantageous over TMZ (30.2%±6.7%). However, 5-y-OS was lower in CE (19.0% ±16.8%) than in TMZ (39.8%±7.7%) and HIT-SKK (42.9%±8.7%). 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

EPEN-07. PATTERNS OF EXTRANEURAL METASTASES IN PEDIATRIC SUPRATENTORIAL EPENDYMOMA: CASE SERIES AND REVIEW OF THE LITERATURE

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BACKGROUND: Ependymomas account for 10% of all malignant pediatric intracranial tumors. Standard therapy includes maximal safe surgical resection followed by involved-field radiation. Up to 50% of localized pediatric ependymomas recur. Extraneural metastases at time of recurrence are rarely reported. OBJECTIVE: To describe extraneural metastases of pediatric ependymomas. METHODS: Retrospective review of patients' medical records and literature review. RESULTS: Three patients with history of locally recurrent, supratentorial ependymoma developed extraneural metastases: one in a cervical lymph node, one with a scalp nodule, and one with a dural lesion. Each extraneural recurrence had similar histologic and molecular features as the initial diagnosis. The cervical lymph node recurrence was treated with multimodal therapy; she is without disease four years later. The isolated scalp nodule occurred at the exit site of a subgaleal drain placed during prior resection. Following nodule resection, he developed additional scalp and lymph node disease and is receiving palliative care. The isolated dural recurrence occurred at the exit site of a ventriculoperitoneal shunt placed following a previous resection. She died of progressive disease 18 months after dural lesion resection. Reports of lymph node, scalp, and dural metastases of ependymomas are exceedingly rare, and outcomes are poor. CONCLUSIONS: Extraneural manifestations of ependymoma are rare. Regional seeding from prior surgical procedures may play a role in metastatic spread. Extraneural metastases should be considered in children 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.

EPEN-09. IMPACT OF MOLECULAR SUBGROUP ON OUTCOME FOR INFANTS <12 MONTHS WITH INTRACRANIAL EPENDYMOMA - GERMAN EXPERIENCE FROM HIT2000, INTERIM-2000-REGISTRY AND I-HIT-MED REGISTRY

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BACKGROUND: For infant ependymoma (EP), decision for radiotherapy during first-line therapy is a dilemma. We analyzed therapy outcomes of EP patients younger than 12 months at diagnosis according to molecular subgroup. PATIENTS AND METHODS: Between 2001 and 2017, 30 patients with histological diagnosis of intracranial EP <12 months at diagnosis with DNA-methylation profiling available were registered in HIT-MED-studies/-registries. RESULTS: In 3/30, DNA methylation-based CNS tumor classification suggested a diagnosis other than EP or could not be assigned to a reference class. Of the remaining 27 tumors, 16 were classified as PF-A, 8 as *RELA*-fusion positive and 3 as *YAP*-fusion positive. Median age at diagnosis was 0.73 (0.30–0.99) years. After a median follow-up time of 5.36 (0.20–12.90) years, 59.3% experienced progressive disease (PD). 5y-PFS and -OS for the whole cohort were 38.2% and 73.1%. *RELA*- and *YAP*-fusion positive EP had significantly better OS than PF-A (5y-OS for PF-A: 55.9%; *RELA* 100%; *YAP* 100%; p=0.023). PFS was not significantly different. All but one patient with relapsed PF-A died despite multimodal salvage strategies. In contrast, patients with relapsing *RELA*- and *YAP*-fusion positive EP (n=5), survived with a combination of re-surgery and first or second local radiotherapy. CONCLUSION: In this cohort of infants <12 months, patients with PF-A had a significantly inferior OS compared to patients with *RELA*- and *YAP*-fusion positive EP. Salvage therapy was ineffective for patients with PF-A, whereas patients with can *RELA*- and *YAP*-fusion positive EP can be long-term survivors after PD. Therefore, subgroups-specific therapy should be discussed.

EPEN-10. SPINAL MYXOPAPILLARY EPENDYMOMA AND METHYLATION-PROFILING: THE MD ANDERSON CANCER CENTER (MDACC) EXPERIENCE

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