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, lumbar spine location was commonest and 6 had leptomeningeal spread at diagnosis. All the patients underwent surgery and 11 received radiation following surgery. Eight patients below the age of 30 received radiation due to residual disease or metastases. Methylation profiling revealed 11,752 CpGs differentially methylated between the younger and older patients ($p < 0.05$), however only one CpG cg22496254 associated with gene NCAPG/DCAF16 (role in promoting mitosis) was detectable with FDR < 0.25 that overly methylated in the younger age group. This is a new finding in MPE. CONCLUSIONS: Spinal MPE is a rare spinal tumor. Our study though limited by numbers, showed younger patients had aggressive phenotype, most requiring radiation. Methylation profiling reaffirmed this finding and trend in the younger patients. Prospective studies in a larger cohort of patients with methylation profiling are needed to identify prognostic variables and new targets for treatment.

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

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BACKGROUND: Among the nine molecular subgroups of ependymoma identified, posterior fossa ependymoma A (PF-EPN-A) confers 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 distinct epigenetic 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. Genetic 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 nodular 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 with almost complete resolution of disease at 15 months. Unfortunately, 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 administered concomitantly with vorinostat therapy. This response highlights the importance of further studies evaluating this combination therapy and its potential use in this population.

EPEN-13. PRIMARY EXTRADURAL SACROCOCCYGEAL SUBCUTANEOUS MYXOPAPILAR EPENDYMOMA MISDIAGNOSED AS PILONIDAL CYST IN A 7 YEAR-OLD BOY: A CASE REPORT

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BACKGROUND: Ependymomas occur in the brain or spinal cord and rarely as an extraspinal variety at the sacrococcygeal region, separated from the spinal cord. This rare presentation is thought to originate from a group of heterotopic ependymal cells called the coccygeal medullary vestige. There are few reports of this occurrence in children. CLINICAL CASE: A 7-year-old male presented with a history of a soft mass arising in the sacrococcygeal area 3 years earlier, diagnosed as pilonidal cyst at primary level and treated with surgery twice, as this mass recurred the boy was sent to our hospital, a 3rd surgery was performed, all tumoral tissue was removed, no attachment with dural space was founded, pathology revealed myxopapilar ependymoma with positivity for PS100, EMA and Vimentin. After surgery a Follow up MRI of cranium and spine showed absence of disease, no radiotherapy neither chemotherapy was implemented. He has been on surveillance from 3 years now without recurrence. CONCLUSION: This report highlights the fact that pediatric ependymoma can have an extradural presentation and can be confounded with pilonidal cyst, total resection is needed to control the disease. Potential for recurrence or metastatic disease can continue 20 years from the time of primary tumor, so prolonged surveillance is important.

EPEN-14. GENERATION OF A C11ORF95-RELA FUSION TARGETING ANTIBODY AS A DIAGNOSTIC TOOL FOR SUPRATENTORIAL EPENDYMOMA

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Ependymomas account for 10% of paediatric brain tumours and arise in the ventricular walls of the central nervous system. Ependymomas were previously classified as one tumour type and all patients received similar treatment. However, recent genomic studies have identified nine different molecular subgroups of the disease, including one supratentorial subtype characterized by a novel fusion gene C11ORF95-RELA. When introduced into neural stem cells, this fusion is a potent driver of tumorigenesis and its presence in patient samples has previously also been shown to negatively correlate with overall survival. Accurate diagnosis of this subgroup is currently limited to sophisticated approaches such as break-apart FISH or RNA sequencing. Here, we report the generation of a C11ORF95-RELA Fusion-specific antibody that can be used for routine immunohistochemistry (IHC). Candidate antibodies were first selected using phage display and favourable leads were subjected to affinity maturation using ribosome display after a screening process involving immunoblotting and IHC. Further IHC-based screening of affinity-matured candidates using fusion-positive and -negative mouse tissue as well as human fusion-negative ependymoma tumour tissue produced one lead antibody. The antibody detects fusion-specific nuclear staining pattern on fusion-positive tissue and does not react with fusion-negative tissues. This candidate antibody is currently being tested on human fusion-positive ependymoma tissue. This accurate diagnostic tool holds great promise to transform the management of patients with supratentorial ependymoma.

EPEN-16. TRANSCRIPTIONAL REGULATORY CIRCUITRIES AS MOLECULAR TARGETS IN EPENDYMOMA

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Genomic sequencing has driven precision-based oncology therapy; however, genetic drivers remain unknown or non-targetable for many malignancies, demanding alternative approaches to identify therapeutic leads. Ependymomas comprise histologically similar tumor entities driven by distinct molecular mechanisms, such as fusion oncoproteins, genome-wide chromosomal instability, or disruption of DNA methylation patterns. Despite these differences, ependymomas resist chemotherapy and lack available targeted agents for clinical trial development. Based on our previous findings, we hypothesized that mapping chromatin landscapes and super enhancers (SE) could uncover transcriptional dependencies as targets for therapy (Mack, Pajtler, Chavez et al., Nature, 2018). To functionally test the requirement of these SE genes for ependymoma cellular growth, we designed a pooled RNA interference screen against 267 SE associated genes. Our screen was performed in one *C11ORF95-RELA*-fusion model and two PF-EPN-A models as controls in biological triplicates. As an indication that our screen was successful, positive controls scored among lead hits including *KIF11, BUB1B, PHF5A and MYC*. Importantly, we identified many subtype specific dependencies in both *C11ORF95-RELA*-fusion and PF-EPN-A models, thus revealing novel pathways that potentially govern subgroup-specific ependymoma cell growth. Further, several candidates detected across all ependymoma lines were also identified as pan-cancer dependencies or glioma/glioblastoma specific essential genes from the DepMap Cancer Dependency Gene Resource. Our findings reveal novel targets and pathways that are essential for ependymoma cell growth, which may provide new insight into therapeutic strategies against these aggressive brain tumors.

EPEN-17. FAVORABLE OUTCOME TO INTENSIVE CHEMOTHERAPY WITHOUT IRRADIATION IN INFANTILE METASTATIC EPENDYMOMA WITH A NOVEL MOLECULAR PROFILE: A CASE REPORT

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