firmed the expression of the endogenous C11orf95-RELA fusion gene. These results suggested that a gene rearrangement is a primary mechanism to form the C11orf95-RELA fusion which is the direct driver of tumorigenesis. Our system to simulate a genomic event will provide significant insights into the understanding of the tumorigenic mechanism in ependymomas.

EPEN-35. PERITONEAL CARCINOMATOSIS OF ANAPLASTIC EPENDYMOMA: FIRST REPORTED CASE

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Peritoneal Carcinomatosis of anaplastic ependymoma is not a previously reported entity. The authors report on a child with multiple successfully treated brain and spine disease occurrences who subsequently develops carcinomatosis of the abdomen and no evidence of CNS recurrence. Ependymoma accounts for up to 10% of childhood CNS tumors diagnosed in the United States with a median age of 51–71 months. Typical locations are based on age. Disease is typically treated with surgical resection followed by radiation. The role of chemotherapy has not been proven but currently being examined with open clinical trials. We will describe patient's presentations, clinical treatment and recurrence with subsequent treatment and outcome at time of meeting.

EPEN-36. THE TREATMENT OUTCOME OF PAEDIATRIC SUPRATENTORIAL C11ORF95-RELA FUSED EPENDYMOMA: A COMBINED REPORT FROM E-HIT SERIES AND AUSTRALIAN NEW ZEALAND CHILDREN'S HAEMATOLOGY/ONCOLOGY GROUP

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AIM: Advances in molecular classification of paediatric ependymoma have been pivotal in improving risk stratification and understanding of this disease. *C11orf95-RELA* fused supratentorial ependymoma (ST-EPN) have been reported to have a poor outcome, with 10-year overall survival (OS) of 49% and progression free survival (PFS) of 19%. A cohort of patients from multiple international institutions with molecularly confirmed *C11orf95-RELA* fused ST-EPN were reviewed to assess their disease behaviour. METHOD: We reviewed patients with molecularly determined *C11orf95-RELA* supratentorial ependymoma diagnosed between 1999 – 2019. Demographic information, extent of surgical resection, use of radiotherapy and/or chemotherapy, disease recurrence, treatment at recurrence and clinical outcome data was collected. PFS and OS of all patients were estimated using Kaplan-Meier method. RESULTS: A total of 76 ST-EPN patients with *C11orf95-RELA fusion* were identified (median age: 7 years3 months, range: 5 months – 18 years7 months). 58 patients (76.3%) had complete surgical resection. 70 patients(92.1%) received radiotherapy. 55 patients(72.3%) received chemotherapy. The 10-year OS of *C11orf95-RELA* fused ST-EPN was 72.4% and PFS was 63.8%. In contrast, ST-EPN at a single institution with unconfirmed molecular status had an OS of 61.1% and PFS of 34.9%. CONCLUSION: Detailed molecular analysis identified distinct subgroups of patients with ST-EPN. Patients from this cohort with *C11orf95-RELA* methylation profiles had a significantly higher OS compared to previous reports and those with unconfirmed fusion status, emphasising the critical importance of complete molecular profiling to assist in treatment decision making. Complete molecular analysis in future prospective cohorts is essential for accurate risk stratification and treatment selection.

EPEN-37. TREATMENT OUTCOME OF RECURRENT EPENDYMOMA IN CHILDREN IN NORTHERN EGYPT

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INTRODUCTION: 1/3 of Ependymoma patients will develop recurrence with only 25% are long term survivors. Treatment is usually between surgery, radiotherapy or combinations. PATIENTS AND METHODS: Retrospective review of children with recurrent Ependymoma in northwest of Egypt between 2005 and 2019 in Alexandria School of medicine records. RESULTS: 27 patients were identified 19 of them after 2010. The median age is 9.7 years (1.5–19), with 16 males and 11 females. Pathology were 11 grade II Ependymoma and 16 anaplastic Ependymoma. 16 had gross residual disease after 1st surgery and 22 received radiotherapy initially at median dose of 53.5 Gy, 4 patients received suboptimal radiotherapy. The initial site was14 supratentorial tumors and 13 infratentorial. Median time to recurrence is 27.6 months(3–84), and recurrences were 17 local and 9 CSF disseminated, and one patient had recurrence at the scar with lung metastasis. At a median follow up of 56.6 months 14(51.8%) are still alive. Treatment was surgery only in 6(4 alive) radiotherapy alone in 2(1alive), combined in 15(9 alive) and 4 patients received neither. The best outcome were in patients with late local relapse treated with complete resection and CSI after 2010. Radiotherapy dose was between 54 to 57.3 Gy and one patient developed reirradiation injury at brain stem. 5 of the 14 living patients is having toxicity in form of hearing aids (4) and low TSH(1). CONCLUSION: Aggressive treatment of recurrent Ependymoma with surgery and radiotherapy is feasible and about half of the patients are salvageable.

EPEN-38. EZH2 INHIBITORY PROTEIN (EZHIP/CXORF67) EXPRESSION IS HIGHLY CONCORDANT WITH H3K27ME3 LOSS AND IS A PROMISING SURROGATE MARKER FOR POSTERIOR FOSSA TYPE A EPENDYMOMAS

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BACKGROUND: Gene expression and DNA methylation have identified 2 distinct clinicopathological subgroups among the WHO Grade II/III posterior fossa (PF) ependymomas (EPN), of which the PF-A molecular subgroup associates with poor outcome. OBJECTIVE: To analyse the utility of immunohistochemistry for H3K27me3, Tenascin C, EZHIP (Cxorf67), EZH2 and fluorescence-in-situ-hybridisation for chromosome 1q21 locus gain in the prognostic stratification of PF-EPNs. METHODS: All PF Grade II/III tumors were retrieved (2009–2019). Immunohistochemistry for H3K27me3, H3K27M-mutation-specific antibody, EZH2, EZHIP, Tenascin-C and fluorescence in-situ hybridisation for 1q21 locus was performed and compared with outcome. RESULTS: 71 PF-EPNs were included. H3K27me3 loss (PF-A) was seen in 65% (46/71) of cases, of which majority were positive for EZHIP (73%, 24/33) and Tenascin C (65%, 28/43). Minority showed chromosome 1q gain (19%, 8/42). An EZHIP negative PF-A tumor was immunopositive for H3K27M-mutant staining, while all others were negative. PF-A EPNs occurred at a median age of 4.5 years (range 1–53), were predominantly grade III (Grade III:II – 1.6:1), and 50% (10/20) of patients on follow-up experienced tumor progression. EPNs with retained H3K27me3 (PF-B) did not show EZHIP expression (0/20) or 1q gain; however, tenascin C expression was seen in 47% (8/25) of them. They occurred predominantly in adults, showed Grade II preponderance and only 2/11 patients on follow-up experienced progression. EZH2 expression did not correlate with H3K27me3 loss but positively correlated with EZHIP expression (p=0.015). CONCLUSION: H3K27me3 is a reliable surrogate for prognostic classification of PF-EPNs. EZHIP expression is highly concordant with H3K27me3 loss and is a valuable adjunct.

EPEN-39. CLINICAL STRATIFIED TREATMENT OF LOCALIZED PEDIATRIC INTRACRANIAL EPENDYMOMA WITH COMBINED LOCAL IRRADIATION AND CHEMOTHERAPY WITHIN THE PROSPECTIVE, MULTICENTER E-HIT TRIAL – THE MOLECULAR SUBGROUP MATTERS

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