ment) resulted in endocrine preservation of all patients and a significantly longer EFS when compared to upfront surgical resection in this single institutional retrospective review. Further analyses will elucidate the implications of ORI with respect to ophthalmological, vascular and neurocognitive outcome.

RARE-24. THE USE OF NOVEL *IN VITRO* MODELS TO STUDY ADAMANTINOMATOUS CRANIOPHARYNGIOMA DISEASE BIOLOGY AND DRUG RESPONSE

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BACKGROUND: Challenges around the design and investigation of cell culture models of adamantinomatous craniopharyngioma (ACP) have arisen from the cellular heterogeneity of these tumors, with populations that harbor disparate requirements in culture. Novel approaches to *in vitro* modeling of ACP are needed. METHODS: Intraoperatively collected tumor specimens were mechanically digested and plated under conditions tailored to the cell population of interest. ACP tumor-derived fibroblasts and epithelial cells were isolated using serum-containing and keratinocyte-specific media respectively. ACP-derived epithelial cells were immortalized via SV40 virus transfection and puromycin treatment for stable cell-line generation. Cell line validation included immunofluorescence with markers appropriate for the cell population of interest. RNA sequencing of cell lines was compared to ACP transcriptome reference data. Cell typing was conducted using short tandem repeat sequencing. RESULTS: ACP fibroblasts and ACP epithelial cells maintained spindle-like and cobblestone morphologies respectively, even after 4 passages. Immunofluorescence staining confirmed high levels of Vimentin expression in ACP-derived fibroblasts, and panCK and B-catenin in ACP-derived epithelial cells. Point mutation in exon 3 of the CTNNB1 gene was identified in ACP-derived epithelial cells. CONCLUSION: Initial limits related to cell line development in ACP may be addressed through the isolation and culture-specific ACP cell populations. This experience demonstrates the maintenance of validated markers of the cell populations of interest *ex vivo*. While preliminary, such cell lines offer promise as tools for the identification and study of potential therapeutic vulnerabilities in ACP.

RARE-25. PRIMARY INTRACRANIAL EWING SARCOMA IN A CHILD: CASE REPORT

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Ewing sarcoma is a rare childhood tumor which accounts for 3% of all pediatric malignancies. More so, primary intracranial involvement with meningeal attachment is even rarer, accounting for only 1% of all Ewing sarcoma. We report a case of a 5-year-old boy who presented with headache, vomiting, and left-sided weakness that rapidly progressed over a period of three months. Cranial MRI showed a 7.1 x 6.7 x 8.6 cm multilobulated, heterogeneously enhancing, mixed solid and cystic extra-axial tumor compressing the frontoparietal lobe and causing significant midline shift. It was attached to the falx and infiltrated the middle third of the superior sagittal sinus. We performed a large right frontoparietal craniotomy to excise the tumor. Because of massive bleeding from the tumor, only a subtotal resection was possible. The bone flap was left out. The patient was discharged fully awake but with right hemiplegia on the fourteenth post-op day. Histopathologic examination revealed a spindle cell neoplasm that exhibited diffuse membranous staining for CD99. Fluorescence in-situ hybridization confirmed EWSR1 gene rearrangement, consistent with Ewing sarcoma. Three months after his surgery, the patient subsequently received 56 Gy of radiation therapy. At twelve months post-op, he remains fully awake and is back in school. He has residual left hemiparesis, but with antigravity movement. A multidisciplinary team involving Pediatric Oncology, Pediatric Neurology, Neurosurgery, Pathology, Radiation Oncology, and Rehabilitation Medicine is essential for patients with rare central nervous system tumors, to maximize effective treatment strategies despite limited resources.

RARE-26. EVALUATING THE CLINICAL UTILITY OF DNA METHYLATION PROFILING FOR CHOROID PLEXUS TUMORS

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INTRODUCTION: Choroid plexus tumors (CPT) are rare, potentially aggressive CNS tumors with defined histologic criteria for grading. In recent years, several patients within our practice have demonstrated discordance between histological diagnosis and clinical behavior. DNA methylation profiling has emerged as a potential diagnostic adjunct for aiding clinical planning and treatment approach. In this study, we sought to retrospectively evaluate the clinical utility of DNA methylation profiling within our cohort of patients with CPT. METHODS: We performed a retrospective chart review of all patients with choroid plexus tumors treated at Dana-Farber / Boston's Children's Cancer and Blood Disorder Center between 1990-2021, evaluating the histology, treatment approach, and clinical outcome. Available tissue samples were sent to the National Institute of Health for DNA methylation profiling. RESULTS: Seventeen patients with CPT were identified. Median age at diagnosis was 1.8 years (range: 0.4-27.7). Histologic diagnosis included choroid plexus papilloma (CPP; n=4), atypical choroid plexus papilloma (aCPP; n=5), and choroid plexus carcinoma (CPC, n=8). DNA methylation in an initial subset placed these tumors with the pediatric type A (n=5), pediatric type B (n=6), and adult (n=1) subgroups. For one patient, methylation profiling returned as unclassifiable (possibly representing an alternative diagnosis). Discrepancies with the histologic grade were noted in several cases: one patient diagnosed with CPP grouped with pediatric type B CPT on methylation analysis, had rapid recurrence, and a diagnosis of CPC was made on a re-resection specimen; another patient with aCPP with concerning features was classified as pediatric type A by methylation, and is without evidence of disease after initial complete resection. Survival outcomes based on histologic diagnosis and molecular subgroups are compared and reported. CONCLUSION: DNA methylation profiling is a useful tool for the diagnosis of CPT and may have the potential to guide clinical planning and management.

RARE-27. TREATMENT AND OUTCOMES IN ATYPICAL CHOROID PLEXUS PAPILLOMA: A SINGLE INSTITUTION EXPERIENCE

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BACKGROUND: Atypical choroid plexus papillomas (aCPP) are rare central nervous system (CNS) tumors often occurring in very young children. While surgical resection has been a mainstay of therapy, there is no consensus and limited data on the treatment of relapsed or metastatic tumors. METHODS: Retrospective review of the treatment and outcome of patients diagnosed with aCPP since 2011 was performed. RESULTS: Of the seven patients, 4 were male and 3 were female with a median age of 3 years at diagnosis (range: antenatal to 18 years old). All non-metastatic patients (six) were treated with surgery and all achieved gross total resection. Two patients had diffuse leptomeningeal contrast enhancement on diagnosis MRI that resolved after resection of primary tumor alone. One patient developed local relapse underwent re-resection with a GTR then was treated with 4 cycles of chemotherapy based on CPT-SIOP-2000 protocol (carboplatin, etoposide) and has not had further relapse in 24 months. One patient had metastatic disease at the time of diagnosis. They were treated with adjuvant chemotherapy, which stabilized disease for 36 months until they had progression. Additional four cycles were given and has again stabilized disease now 8 months from completion of that therapy. One non-metastatic patient died of unknown causes 28 months from diagnosis. CONCLUSIONS: Surgical resection remains the standard of care for patients with aCPP. However, chemotherapy based on the SIOP backbone may be useful to reduce the need for or to delay radiation therapy in select patients in the relapsed or metastatic setting.

RARE-28. THE USE OF SUBCUTANEOUS INTERFERON IN PATIENTS WITH CRANIOPHARYNGIOMA: AN INSTITUTIONAL RETROSPECTIVE REVIEW

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