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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Current strategies for managing craniopharyngioma result in significant morbidity. Successful treatment with interferon alfa(INFα) after progression is reported in the literature. This retrospective review details our institutional experience with INFα in craniopharyngioma patients. Method: Between 2000-2021, we treated 81 craniopharyngioma patients. Twenty-two patients received 26 treatment courses of subcutaneous INFα. Twenty-three courses were evaluable for response. Results: Ten patients received upfront INFα after cyst decompression +/- ommaya placement. Progression free survival(PFS) ranged between 7-38mo. Three patients continue on treatment (10+, 12+, 14+mo); seven progressed (four on treatment (7, 9, 25, 38mo), three after treatment (13, 19, 32mo)). At progression, three underwent surgery alone, three underwent surgery and radiation, one resumed INFα. Thirteen patients received INFα after progression. Prior to INFα, eight patients had had surgery, five surgery and radiation. Two in each group had INFα, previously. PFS ranged between 5-82+mo. One patient remains on treatment (5+mo); four continue in follow-up without progression (23+,40+,64+,82+mo) with two patients avoiding radiation to date; eight progressed (three on treatment (6-8mo), five after treatment (16,24,26,46,71mo)). At progression, two underwent surgery alone, three underwent surgery and radiation, one received re-irradiation, two resumed INFα. While receiving INFα, two patients experienced serious adverse events (one intra-tumoral hemorrhage (not attributed to INFα), one suicidal ideation). Both recovered. One tolerated retreatment with INFα. Three additional patients stopped INFα for intolerance, but two received INFα at subsequent progression. No other unanticipated side effects were reported. Conclusion: INFα therapy in patients with both newly diagnosed and progressive craniopharyngioma delayed the need for aggressive surgical resection and/or radiotherapy in some cases. In some patients, INFα resulted in prolonged stabilization of disease delaying or avoiding radiation. Overall, INFα side effects were manageable. These results are encouraging regarding INFα therapy for patients with craniopharyngioma and warrant further evaluation with a clinical trial.

RARE-29. TRANSCRIPTOME CHARACTERIZATION OF PEDIATRIC ADAMANTINOMATOUS CRANIOPHARYNGIOMA AT THE CELLULAR LEVEL

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BACKGROUND: Adamantinomatous Craniopharyngioma (ACP) is a neurologically devastating brain tumor that affects children and adults. It is histologically heterogeneous with epithelial populations that are characterized by the nuclear accumulation of mutated β-catenin, and activated Wnt signaling. Current models suggest that ACP growth is driven through paracine mechanisms characterized by the senescence-associated secretory phenotype (SASP). However, detailed pathogenic mechanisms remain unknown. Improved definition of the various cellular phenotypes that compose ACP will inform and advance our understanding of this disease. METHODS: Single cell RNA-sequencing (scRNA-seq) and multiplex ELISA were performed on pediatric ACP tissue and cyst fluid, respectively. Reference scRNA-seq data was obtained from PanglaoDB. Preprocessing and standard analyses were conducted using Seurat software. Cellular phenotypes were annotated using the Human Primary Cell atlas. Differential expression and functional enrichment analyses were utilized to identify Wnt-signaling activation and epithelial subpopulations. Paracrine signaling was inferred via CellChatDB. SASP Atlas was utilized to query marker gene lists. Pseudotemporal ordering was performed using monocle3. RESULTS: ACP tissue is heterogenous and contains multiple distinct immune signatures. ACP tissue contains 2 unique epithelial subpopulations, which demonstrate canonical Wnt-signaling and SASP, respectively. Pseudotemporal ordering suggests the initial oncogenic event to be of epidermal character, with subsequent aggressive behavior from a separate epithelial cell population. CONCLUSIONS: Based on gene expression, cell populations that correspond to the histologically identifiable epithelial whorls and palisading epithelium can be identified. These subpopulations display unique functional signatures. Simultaneous and synergistic therapeutic targeting of these separate epithelial populations may lead to improved patient care.

RARE-30. NOVEL COLLISION TUMOR OF CRANIOPHARYNGIOMA AND EPENDYMOMA IN A PEDIATRIC PATIENT: A CASE STUDY

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Collision tumors are rare tumors comprised of two distinct histologies. In this case report, we discuss a suprasellar collision tumor consisting of adamantinomatous craniopharyngioma and supratentorial ependymoma in a pediatric patient. Case Presentation: Our patient was a two-year-old female with progressive craniopharyngioma status post cyst decompression with Ommaya reservoir placement, subcutaneous peginterferon, Ommaya taps, and subtotal resection. An MRI three months post-resection showed progression and treatment was started with subcutaneous interferon alfa. After eight weeks, she presented with new onset headaches and vomiting. MRI demonstrated tumor progression with associated obstructive hydrocephalus. She underwent a subtotal resection. Pathology revealed recurrent adamantinomatous craniopharyngioma and a 0.5cm ependymoma with classic histomorphology lacking anaplasia features. The ependymoma was positive for GFAP immunostain and EMA immunohistochemistry highlighted a 'dot-like' reaction. The Ki-67 proliferation index was very low (<1%). The limited diagnostic material precluded further genomic characterization of the ependymoma. The previous pathology was reviewed and no ependymoma was identified. Spine MRI was negative for metastatic disease. CSF cytology was negative for malignant cells. Following recovery from surgery, she received 54Gy (RBE) focal proton radiation. Eight months from completion of therapy, surveillance MRI shows stable residual tumor. Genetic work-up for cancer predisposition syndrome is in process despite no strong family history of cancer. Discussion: Due to the patient's young age at diagnosis, our initial treatment strategy was to delay radiotherapy and utilize other treatment options. Following diagnosis with a collision tumor, the patient proceeded to radiotherapy to manage both tumor components. The role of interferon in the development of a collision tumor in this patient in unknown, but we suspect it to be unrelated. Conclusion: To our knowledge, this is the first documented case of a suprasellar collision tumor comprised of craniopharyngioma and ependymoma. Discovery of the collision tumor impacted the patient's treatment plan.

RARE-31. RADIATION INDUCED MALIGNANCY ASSOCIATED WITH PEDIATRIC CRANIOPHARYNGIOMA: A SINGLE INSTITUTION EXPERIENCE

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BACKGROUND: Therapy for pediatric craniopharyngioma, a rare suprasellar tumor, includes surgical resection with consideration for intracranial radiation. Radiation is associated with increased risk of secondary malignancies. Between 2000 and 2021, 81 pediatric patients with craniopharyngioma were treated at our institution; 3 of 54 (5.6%) who received radiation therapy(RT) developed secondary malignancy within the treatment field. CASE DESCRIPTIONS: In all 3 cases, initial imaging demonstrated cystic/solid suprasellar mass and underwent resection; pathology revealed calcifications and wet keratin consistent with craniopharyngioma. None had known cancer predispositions. The first patient (male), presented at 4-years-old with headaches. He underwent subtotal resection (STR) with cyst fenestration(w/CF) and received 55.8Gy photon 3D-Conformal RT. Six-years later, the tumor progressed (edge of RT field). Patient underwent a second STRw/CF and fractionated RT(50.4Gy). Both pathologies were consistent with(c/w) papillary craniopharyngioma. Eight-years from first RT, progression occurred again within the RTfield; pathology revealed an (adeno)squamous carcinoma. The second patient, a 5-year-old female, presented with vision loss, underwent partial resection and received 54Gy focal proton therapy for adamantinonatous craniopharyngioma. Almost 5-years later, an unresectable right basal ganglia/globus pallidus mass was noted in the 30-54Gy field. Pathology was c/w anaplastic astrocytoma(AA). The third, a 9-year-old female was treated with 54Gy photon radiation and 7 years later had evidence of increasing mass. Pathology revealed high-grade-diffuse-glioma(HGDG). Molecular analysis of AA/HGDG both revealed PDGFRA amplification and CDKN2A/B homozygous loss. DISCUSSION/CONCLUSION: Malignant CNS tumors are reported following radiotherapy for a variety of primary CNS lesions. While radiation is a valuable therapy in achieving long-term disease control of pediatric craniopharyngiomas, it is important to understand the risk of developing secondary malignant neoplasms. Our report adds to the body of literature describing secondary malignancies post radiation therapy for the treatment of pediatric craniopharyngioma.