the results of a transcriptome analysis of both pediatric and adult ACP to identify biological differences between these groups that may provide novel therapeutic insights, or support the assertion that potential therapies identified through the study of pediatric ACP may also have a role in adult ACP. Following bulk RNA sequencing, we explored the differential expression of adult ACP versus pediatric ACPs via geneset enrichment analysis. Preliminary studies identified a decreased cytokine gene expression profile in adult ACP that may correspond to a reduced senescence-associated secretory phenotype (SASP), which has recently been described in pediatric ACP. Additionally, our results corroborate our previous work demonstrating an increase in IL-6 expression and overall inflammatory response in pediatric tumors. To further characterize age-associated transcriptomic differences, we performed immunohistochemistry, immunoblotting, and other proteomic studies. Our results demonstrate phenotypic similarities and differences between pediatric and adult ACP.

CRAN-35. CHILDHOOD CYSTIC CRANIOPHARYNGIOMA: 15 YEARS OF INTRA-TUMORAL CHEMOTHERAPY WITH INTERFERON-ALPHA AT IOP/GRAACC/UNIFESP

Patricia Dastoli[1,2], Jardel Nicácio[1,2], Marcos Costa[1,2], Andrea Cappellano[1], Nasjla Silva[1], and Sergio Calvalheiro[1,2]; [1]Instituto de Oncologia Pediátrica GRAACC/UNIFESP, Division of Neuroncology, Sao Paulo, Brazil, [2]Universidade Feceral de São Paulo- Discipline of Neurosurgery, Division Neurosurgery Pediatric, Sao Paulo, Brazil

INTRODUCTION: Management of patients with craniopharyngioma is still controversial. Currently, quality of life is the goal for all treatment modalities. OBJECTIVE: To analyze the several treatment strategies and outcomes of craniopharyngioma patients in 15 years of experience in a single institution. METHODS: The authors retrospectively evaluated 110 craniopharyngeoma patients treated between 2002 and 2017, at IOP/GRAACC/UNIFESP. Data regarding initial clinical presentation, body mass index (BMI), endocrinological complications, treatment modalities and outcomes were collected. RESULTS: From 110 patients, 95 patients were eligible for evaluation. Fifty-five were boys and 40 girls. The mean age at treatment was 10,5 years. Fifty-six percent of children presented with signs and symptoms of intracranial hypertension, 24% with visual impairment and 38,5% with endocrinological disturbances. At the time of diagnosis, the mean BMI was 20,48kg/m^2. Thirty-eight children with cystic tumors were treated with intralesional interferon-alpha, 13 progressed to conventional treatment. Among the remaining 57 patients, 9 were treated with microsurgery, 29 with microsurgery + radiotherapy, 12 with endoscopic transesphenoidal surgery + radiotherapy and 7 recurrent patients with intracystic interferon-alpha. The mean BMI of patients treated exclusively with interferon-alpha was 22,88kg/m^2 (ideal weight) and with microsurgery + radiotherapy was 29,65kg/m^2 (overweight). After treatment, 61 cases (65%) progressed to panhypopituitarism. The mean follow up was 7,13 years. Eleven deaths occurred. Currently 41 patients are out of treatment. CONCLUSION: The interferon-alpha intracystic therapy has been shown to be effective in the control of predominantly cystic tumors, decreasing or delaying the morbidity from conventional treatment.

CRAN-37. MOLECULAR HETEROGENEITY OF SUBEPENDYMAL GIANT CELL ASTROCYTOMAS IN PATIENTS WITH MILD-PHENOTYPE TUBEROUS SCLEROSIS COMPLEX

Cody Nesvick, Amulya Nageswara Rao, Aditya Raghunathan, and David Daniels; Mayo Clinic, Rochester, MN, USA

Subependymal giant cell astrocytomas (SEGAs) are rare central nervous system tumors that are almost pathognomonic of tuberous sclerosis complex (TSC). These lesions are World Health Organization (WHO) grade-I tumors that arise in the lateral ventricles near the foramen of Monro and almost always contain detectable mutations in TSC1 or TSC2. We present three children with TSC in whom a diagnosis of SEGA was the presenting feature and who displayed a mild TSC phenotype. All patients underwent surgery for tumor resection. Following surgical resection, germline testing for TSC1 and TSC2 mutations was performed, and pathologic specimens subsequently underwent molecular analysis using chromosomal microarray and a 50-gene next-generation sequencing. There were two males and one female with an age range of 12 to 18 years. All patients had evidence of cortical tubers on brain MRI, and two had hypomelanotic macules; no other manifestations of TSC were present. None of these patients were found to have detectable germline mutations in TSC1 or TSC2. One tumor specimen contained copy-neutral loss of heterozygosity (cnLOH) of chromosome 9q34 affecting TSC1 and NOTCH1. The second specimen contained cnLOH of chromosome 16p12-13 affecting TSC2; DNA sequencing confirmed a known pathogenic mutation in TSC2 in this patient. A third specimen was cytogenetically normal but contained a missense mutation in NOTCH2 of unknown significance. These data demonstrate that SEGAs in patients with a mild TS phenotype are molecularly heterogeneous, sometimes containing genetic alterations in TSC1 or TSC2 but also containing mutations in other pathways associated with growth, development and neoplasia.

CRAN-38. CLINICOPATHOLOGIC FEATURES OF SUPRASELLAR AND LATERAL CEREBRAL DESMOPLASTIC INFANTILE ASTROCYTOMA AND GANGLIOGLIOMA

Cody Nesvick, Ryan Naylor, Anton Wohl, Aditya Raghunathan, Laurence Eckel, Gesina Keating, and David Daniels; Mayo Clinic, Rochester, MN, USA

Desmoplastic infantile ganglioglioma (DIG; also known as desmoplastic infantile astrocytoma, DIA) is a rare, World Health Organization (WHO) grade-I central nervous system tumor that most often arises in very young children, often with rapidly enlarging head circumference. We reviewed the records of all infants diagnosed with pathology-confirmed DIG/DIA at Mayo Clinic from 1997 to 2016. Immunohistochemical staining was performed on tumor specimens to assess expression of IDH1 R132H, BRAF V600E, H3K27me3 and INI1. Seven patients were included in this study: age range was 2 - 9 months, and four were male. Six patients presented with rapidly enlarging head circumference, and one presented with isolated seizures. On immunohistochemical analysis, all tumors were negative for R132H-mutant IDH1. INI1 expression was retained in all specimens, and BRAF V600E expression was negative or only very focally positive. H3K27me3 expression was partly lost in four tumors. Three tumors were located in the suprasellar region, and four were located in the lateral cerebral hemispheres. All patients underwent surgery for tumor resection; a gross total resection was achieved in the lateral cerebral tumors, and a subtotal resection was achieved in the suprasellar tumors. At 2-year follow-up, none of the patients with a hemispheric tumor developed long-term sequelae from their surgery, but each patient with a supresellar tumor had a neurologic or endocrinologic deficit from tumor progression or surgery, including visual impairment, panhypopituitarism and/or hemiparesis. These results highlight the importance of tumor location in prognosticating these challenging tumors, suggesting that suprasellar location is associated with a poorer prognosis.

CRAN-39. BARIATRIC SURGERY FOR TREATMENT OF HYPOTHALAMIC OBESITY

Thomas Inge and Megan Kelsey; Children's Hospital of Colorado, Aurora, CO, USA

Craniopharyngiomas (CP) are neoplasms generally found in the area of the pituitary and hypothalamus, and treatment can lead to clinically significant endocrine, neurologic, behavioral and metabolic dysfunction. Severe obesity occurs in a high percentage of patients with CP, leading to metabolic comorbidities, impaired quality of life, and weight-related physical disabilities. This resultant hypothalamic obesity (HyOb) is often refractory to conventional lifestyle interventions and pharmacotherapy. Roux en Y gastric bypass (RYGB) and vertical sleeve gastrectomy (VSG) are bariatric operations for which there is an emerging experience in patients with HyOb. Although existing literature is limited, with heterogeneity of outcomes, evidence shows that RYGB can result in long term (≥2 years) weight loss of in excess of 20%, while weight loss after VSG is more modest at >10%. In this presentation, we will review the current evidence pertaining to the use of bariatric surgery in the treatment of CP-HyOb. We will also compare these results to those reported for other populations of HyOb, including Prader-Willi Syndrome and hypothalamic melanocortin signaling defects. While initial reports of bariatric surgery in CP-HyOb are promising, their limited scope makes it difficult to draw any substantial conclusions as to the long term safety and efficacy of bariatric surgery in CP-HyOb, and in particular which bariatric operations are most indicated for these patients. More robust, controlled, prospective studies with long term follow-up are needed to better define the role of bariatric procedures in the treatment of HyOb.

CRAN-40. A NATIONAL UK GUIDELINE FOR MANAGING PITUITARY ADENOMAS IN CHILDREN AND YOUNG PEOPLE UNDER 19 YEARS DEVELOPED ACCORDING TO THE AGREE II FRAMEWORK

Jo Blair[2], Márta Korbonits[1], Amy Ronaldson[1], Mary Dang[1], and Helen Spoudeas[3]; [1]Barts and the London School of Medicine, Queen Mary University of London, London, UK, [2]Alder Hey Children's NHS Foundation Trust, Liverpool, UK, [3]Great Ormond Street Hospital for Children NHS Foundation Trust, London, UK

Pituitary adenomas are usually benign tumours arising from the hormone-secreting cells of the anterior pituitary gland, resulting in excess hormone secretion and the development of characteristic syndromes, such as Cushing's disease, acromegaly and hyperprolactinaemia, and/or mass effects on surrounding vital structures causing for example visual disturbances and pituitary hormone deficiencies. In children and young people under 19 years (CYP), the management of pituitary adenomas is particularly challenging given their extreme rarity, more aggressive phenotype and likely genetic predisposition, as well as the lack of age- and pituitary-specific multidisciplinary teams in current decision-making and service provision. Hence, the UK sought to create national, high-quality, multi-professional guidance using AGREE II methodology