meningiomas, the molecular profiles of childhood meningiomas have not been elucidated in detail. We analyzed 41 tumor samples from 37 pediatric meningioma patients (female: 17; male: 20; age range: 1-21 years). Anaplastic meningioma WHO grade II was the most frequent histological subtype (N=14, 38%). Most tumors were located at the convexity (N=18) or the skull base (N=15). Lack of SMO, AKT, KLF4/TRA27 mutations by Sanger sequencing (n=22) prompted whole genome sequencing of a subset (n=7). All tumors exhibited biallelic inactivation of NF1 (combined deletion and germline (57) or somatic (27) base exchanges/frameshifts).

Subsequently, tumor samples from all 37 patients were subjected to 450K DNA methylation profiling and targeted DNA sequencing using brain tumor specific gene panel. Loss of chromosome 18 was frequent (N=28, 76%), followed by loss of chromosome 1 (N=12, 32%) and chromosome 10 (N=7, 19%). Moreover, separation into three groups was evident: One encompassing all clear-cell meningiomas with enrichment for SMARCE1 mutations, a second dominated by atypical meningiomas, and a third group composed of benign meningiomas, as well as rare subtypes such as rhizoid meningiomas. When analyzed with 105 adult tumors, most of pediatric meningiomas (28/37) clustered into a separate methylation group both by unsupervised hierarchical clustering and t-statistic nearest neighbor embedding (t-SNE). These data suggest that pediatric meningiomas are genetically distinct from adult counterparts.

**NFM-12. ENHANCING BRAIN LESIONS IN CHILDREN WITH NEUROFIIBROMATOSIS TYPE 1**

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Children with Neurofibromatosis type 1 (NF1) are at an increased risk of developing glial tumors. Gadoilimum enhancement signals blood-brain barrier breakdown and implies tumor activity. In an attempt to determine the prevalence and outcome of enhancing brain lesions in children with NF1, we performed a retrospective analyses of all patients with NF1 followed in a Pediatric Neuro-Oncology department with periodic brain MRI. Of 64 children (31 males, 17 females), 17 (26.5%) who followed up included neuroimaging studies (optic pathway gliomas 62, other low grade gliomas 3, enhancing lesion 1, T2 hyperintensity), 17 (25.3%) showed evidence of enhancing brain lesions (median age at diagnosis 8 years). A total of 25 lesions were identified, mainly in the posterior fossa (60%). Four lesions (4 patients) were symptomatic. One symptomatic patient (mesencephalic lesion), underwent solely a ventriculostomy. Three symptomatic and 3 asymptomatic patients (based on imagiological progression) underwent chemotherapy (complete response obtained in 3 patients, partial response in 2, 1 patient under treatment). In patients with disease under chemotherapy, spontaneous regression was observed in 8 lesions (7 patients), the others remain imagiological stable.

Mean follow up time 4,5 years. In our experience, enhancing brain lesions are prevalent in children with NF1, and although gadolinium enhancement suggests glioma, the proliferative potential remains questionable. The best approach of this lesions is far from being determined. We admit that symptomatic lesions should be treated. In asymptomatic lesions, watchful surveillance may be appropriate and decision to treat must take in consideration multiple aspects, namely imagiological progression.

**NFM-13. LONG TERM FOLLOW-UP OF OPTIC PATHWAY GLIOMAS IN CHILDREN WITH NEUROFIIBROMATOSIS TYPE 1 – AN ONCOLOGY HOSPITAL EXPERIENCE**

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BACKGROUND: Optic Pathway Gliomas (OPGs) are the most prevalent intracranial tumours in children with Neurofibromatosis type 1 (NF1). Although commonly indolent, OPGs may have an aggressive clinical course and their approach is often challenging. The aim of this study was to evaluate the outcome of OPGs diagnosed in patients with NF1 during childhood (median follow up to 22 years). METHODOLOGY: Retrospective review of demographics, neurological and ophthalmological evaluations, neuroimaging and treatments applied to all children with OPGs and NF1 presenting to an oncology hospital, 1991-2017. RESULTS: Of 62 children (31 males, 31 females, median age at diagnosis 4.0 years), 27(44%) were treated (based on clinical or imagiological progression), in 4 off which the initial decision had been watchful surveillance. Of the treated patients, all were submitted to chemotherapy, 4 underwent surgery, 1 radiotherapy. Visual acuity improved in 18 children and worsened in 3. Complete response in 1 patient, partial response in 19 and disease stability in 6. One patient died. One patient is currently under treatment, the remaining 25 have stable disease. Patients who didn’t undergo treatment remain neuro-ophthalmologically stable, 2 had spontaneous tumour regression. Mean follow up time is 7,9 years. CONCLUSION: Despite the complex management of patients with NF1 and OPGs, inherent to their variable natural history and difficulties in clearly defining neuro-ophthalmological progression and response to treatment in the pediatric population, our results suggest that careful surveillance and treatment is needed to improve quality of life and vision outcome for patients with NF1 and OPGs.

**NFM-14. IDENTIFICATION OF A THERAPEUTIC TIME WINDOW THAT IMPROVES VISION IN AN NF1-DEFICIENT OPTIC PATHWAY GLIOMA MOUSE MODEL**

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Optic pathway gliomas (OPGs) are gliomas that arise early during development in 12-25% of patients with Neurofibromatosis Type 1 (NF1). While chemotherapy is often used for patients who have growing tumors and/or vision loss associated with the tumor, a minority of patients have improvement in their vision following treatment. Thus, a new approach to treatment is needed to improve the vision of patients. In our laboratory, we have a NF1-deficient conditional knockout mouse model that develops optic pathway gliomas around 60 days of age (P60). These mice have associated axon degeneration, loss of retinal ganglion cells, and evidence of vision loss on behavioral testing, thus making this a good model to use in order to identify the optimal therapeutic time window that improves or prevents vision loss. In this study, we treated NF1-deficient mice with a Mek-inhibitor during different developmental time windows and measured response to treatment at different time points and compared vision loss to untreated control mice. Our results suggest that the timing of therapy initiation is critical for optimal outcomes in NF1-associated OPGs.

**NFM-15. BUILDING A NEUROFIBROMATOSIS THERAPEUTICS PROGRAM**

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Neurofibromatosis (NF) therapeutics is an emerging and growing field as more and more children are diagnosed with neurofibromatosis due to better awareness of the disease and diagnostic criteria. Patients with NF have an increased incidence of central nervous system (CNS) tumors and plexiform neurofibromas. Recent developments in the treatment of plexiform neurofibromas have significantly increased the number of patients seen for therapy. In order to provide high-quality care to these patients, a Neurofibromatosis Therapeutics Program (NFTP) should be developed and utilized. The program at Children’s Hospital Colorado has grown over the past few years and now includes a physician, nurse practitioner, nurse, and social worker. We collaborate closely with other disciplines that are vital in the care of the NF patient with plexiform neurofibromas and/or CNS tumors including: neurology, neurosurgery, neuroradiology, ophthalmology, orthopedics, plastic surgery, ophthalmology, neurology, dental, urology, dermatology, genetics, neuropsychology, and rehabilitation. Key players are identified in each subspecialty and then the NFTP is publicized to both primary care and subspecialty programs in order to identify patients who would be best cared for in this setting. Along with collaborating disciplines, the NFTP has developed protocols and standards of care for treatment, research, adverse reaction management, and data gathering to obtain prior authorizations for medications. The NFTP is actively conducting epigenetic research on plexiform neurofibroma tissue obtained during surgery. The NFTP is a strong and vibrant area of growth with the aim to provide high quality of care for the whole child and conduct lab and clinical research.

**NEUROSURGERY**

**NSRG-01. PRIMARY GLIOBLASTOMA IN THE PINEAL REGION OF A PEDIATRIC PATIENT: A CASE REPORT AND REVIEW OF THE LITERATURE**

Lee Hwang, Gabrielle Yeaney, Tanya Tekautz, Kaine Owwozulike, and Violette Lee Hwang

Glioblastoma, a very aggressive type of astrocytoma (WHO grade IV), is extremely rare in the pineal region particularly in the pediatric population. With only 5 pediatric cases reported since 1972, we present another report in the setting of technological advances in neurosurgical intervention. As in this case, patients usually present with non-specific symptoms such as headache, nausea, visual changes, and altered mental status. A complete