presented with an atypical teratoid/rhabdoid tumor (ATRT), 23.5% (21/90) demonstrated SYN, and 21% (19/90) extracranial MRT. RTPS1 was present in 84-, RTPS2 in six patients. In 77% (65/84) complete data on *SMARCB1* mutational status were generated. Methylation subgroup status was available in 59% (40/68) of ATRT or SYN. The 5-year overall- (OS) and event free survival rates of patients with RTPS1 were 19.8 ± 4.8% and 15 ± 4.2%, respectively. Age < 1 year at diagnosis (10.1±4.3% vs. 46.7±11.1%), presence of SYN (5.3±5.1% vs. 24.8±6%), histological diagnosis (ATRT vs. eMRT/RTK/SYN) (26.8±7.1% vs. 11.9±5.6%), localized disease (34.5±8 vs. 8.3±4.6%), and presence of PGV at C-terminal (33±8.6% vs. 9.4±5.3%) were significant prognostic factors for 5-year OS in univariate analysis. INTERPRETATION: In the largest cohort of patients with RTPS, predictors significant for positive outcome could be detected: age > 1 year, absence of SYN, histological diagnosis ATRT, localized disease and PGV located at C-terminal. In our research project, we aim to characterize the complete pheno- and genotype of patients with RTPS to develop a risk score including surveillance recommendation.

NFB-14. POST-OPERATIVE USE OF MEK INHIBITORS TO PREVENT REBOUND GROWTH FOLLOWING PARTIAL RESECTION OF PLEXIFORM NEUROFIBROMAS

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BACKGROUND: Plexiform neurofibromas (PNs) can cause significant morbidity leading to functional impairment, pain, and disfigurement. Management of PNs is challenging. Complete surgical resection is often not possible due to tumor growth along vital structures, and rebound growth is frequently experienced with partially resected PNs. The mitogen-activated protein kinase pathway has been implicated in the growth of PNs, and MEK1/2 inhibitors have been shown to be an effective treatment of PNs. OBJECTIVE: To describe our institutional experience using post-operative MEK1/2 inhibitors in the treatment of pediatric patients with PNs following subtotal resection (STR). METHODS: A single-institution retrospective record review. RESULTS: A total of 35 patients had STR of their PN. Fourteen patients underwent resection alone, ten patients received adjuvant mechanistic target of rapamycin (mTOR) inhibitors and eleven patients received adjuvant MEK1/2 inhibitors. The mean follow-up time was 5.1 years, but relatively shorter for patients receiving adjuvant MEK1/2 inhibitors. Mean time from resection to start of adjuvant therapy and mean duration of adjuvant therapy for patients in the mTOR inhibitor group was 3.3 weeks and 3.9 months, respectively, and for patients in the MEK1/2 inhibitor group was 3.1 weeks and 8.5 months, respectively. The number of patients in each group requiring additional treatment with surgical resection or medical therapy, was 11 of 14 patients (78.6%) in the resection only group, 7 of 10 patients (70%) in the adjuvant mTOR inhibitor group and 3 of 11 patients (27.3%) in the adjuvant MEK1/2 inhibitor group. CONCLUSIONS: A short course of MEK1/2 inhibitors following subtotal resection of PNs is effective in the short term in preventing rebound growth when compared to STR alone or adjuvant mTOR inhibitors. Treatment is well tolerated and should be considered as adjuvant therapy in pediatric patients. Long-term follow-up is necessary to judge the effectiveness of this approach.

NFB-15. "COGNITIVE IMPAIRMENTS IN CHILDREN AND ADOLESCENTS WITH NEUROFIBROMATOSIS"

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INTRODUCTION-BACKGROUND: Cognitive, learning and/or behavioral disorders are common (up to 70%) complications of neurofibromatosis (NF). The first multidisciplinary-clinic for neurocutaneous-disorders was established at the Aghia Sophia Children's Hospital in Athens, Greece, in 2016. Since then, more than 200 children and adolescents with NF have been examined. SCOPE: Acknowledging and indicating awareness on the devastating life-long consequences (poor academic performance, behavioral problems, and limited career prospectives) that can result from cognitive impairment, a research collaboration with educational specialists was recently implemented to examine the neurocognitive functions of children and adolescents with NF. MATERIALS: Children and adolescents aged 7-14 years who suffer from NF type I or type II, were eligible for study entry. The third edition of the Wechsler Intelligence Scale (WISC-III) was used to measure participants' cognitive function. RESULTS: Preliminary results of this ongoing study are presented. Patients' recruitment was limited by the coronavirus disease 2019 (COVID-19) restrictions. At this stage, the research involved 10 participants suffering from NF, with mean (± SD) age of 11.55 (± 1.80) years and a male-to-female ratio of 1. The mean (± SD) full-scale intelligence quotient (IQ) was 85.50 (± 18.80), corresponding to the 0.3rd to 73th percentile range. The mean (± SD) scores of performance IQ and verbal IQ were 84.90 (± 17.43) and 89.40 (± 17.23) respectively, corresponding to the 1st to 73rd percentile range for both subscales. CONCLUSION: Significant cognitive deficits, according to the percentile scores of WISC-III, were demonstrated in the small number of children and adolescents suffering from NF (type I or type II). Cognitive assessment, as part of the multidisciplinary approach of these patients is warranted, to aid timely educational interventions and improve patient learning outcomes.

NFB-17. "OPTIC PATHWAY FINDINGS IN CHILDREN WITH NEUROFIBROMATOSIS TYPE-1 (NF-1)

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BACKGROUND/OBJECTIVES:Optic-pathway-glioma(OPG) represents the most common central-nervous tumor in children with neurofibromatosis type-1(NF1), occurring at an incidence of 15-20%. It is estimated that 1/3 of NF1 patients(pts) with OPG will need treatment. DESIGN-METHODS:We performed a retrospective-review of all NF1-pts examined in the First Hellenic Multidisciplinary-Clinic – Center of Expertise for Neurocutaneous-disorders. Gender, age, MRI-radiological and ophthalmological findings, presence of OPG, management and outcome were analyzed. RESULTS:Since the establishment of the Clinic in 2016, 198pts with clinical diagnosis of NF1 based on NIH1988-criteria were evaluated and of them, 165(73 females, median age:5.5y, range:0.3-17.1y), who had imaging studies were included in this analysis. Eighty three pts(50.3%) had NF1-positive genetic-testing and 45NF1-family-history(27.3%). Imaging-findings from optic pathway were found in 55/165pts(28females). Percentage of pts with findings were 51.7% for <3y, 45.4% for 3-5y, 34.7% for 5-10y and 8.5% for >10y, respectively. The median age of their first brain-MRI imaging was 2.82y. Upon 1stMRI-imaging, 70.9% presented thickness of the optic nerves(ON)(25,4%bilateral, 20% optic chiasm,18.1% right ON, 10.0% left ON), 14.5%ON-tortuosity, 38.1%OPG(43,5% in the optic-chiasm) and 34.5% contrast enhancement. Of notice, 14pts presented an OPG after a median follow-up time of 1.79y. According to LGG2004-protocol indications for treatment, only 15/55pts had to be treated(27,2%, 5pts with family history, 33.3% between 5-10y). Severe vision-loss with need for immediate start of treatment upon 1stMRI imaging was found in 4pts, of whom 75% had family-history and first evaluation after the 5th year of age. Of notice, only 2pts<3y had to receive treatment, one with family-history and one with symptoms(diencephalic syndrome). CONCLUSIONS: Pts with NF1 should be followed by a multidisciplinary-team. Management should be individualized and imaging-studies can be limited to patients at high-risk. Positive family-history may be a negative prognostic factor for OP lesions.

NFB-18. INTEGRATION OF SINGLE-NUCLEI RNA-SEQUENCING AND SPATIAL TRANSCRIPTOMICS TO DEFINE THE COMPLEX TUMOR MICROENVIRONMENT OF NF1-ASSOCIATED PLEXIFORM NEUROFIBROMA AND HIGHLY-AGGRESSIVE MALIGNANT PERIPHERAL NERVE SHEATH TUMORS

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