[4] *Acibadem University Department of Neurosurgery Division of Pediatric Neurosurgery*

Objective: Pediatric high grade gliomas(HGG) have dismal prognosis with median survival of 9-15 months after standard radiochemptherapy. Recent molecular investigations revealed a mismatch repair defect called Constitutional Mismatch Repair Deficiency (CMMRD), which induce pediatric HGG. In CMMRD, there are mutations at least one of the mismatch repair(MMR) genes in both tumoral and non-tumoral DNA. Patients generally have cafe au lait spots resembling the ones in NF-1. **Methodology:** Forty-four pediatric high-grade glioma cases operated in our clinic between 2015-2021 were included in the study. PMS2, MLH1, MSH6, MSH2 immunohistochemical antibodies were applied to the sections prepared from paraffin blocks with tumors of these 44 cases. Next generation Sequencing (NGS) Custom Panel for Brain Tumors was performed with DNA and RNA obtained from neoplastic tissue of 2 cases and germline NGS analysis was performed with DNA obtained from peripheral blood in 1 case. **Results:** MMR protein expression loss was detected in 11 (25%) cases. In 5 (45%) of these 11 cases, MMR protein loss was detected in both neoplastic and non-neoplastic tissue, and these cases were considered as CMMRD. NGS performed in 2 of these 5 cases revealed a hypermutant profile. At least one MMR protein loss was found only in the neoplastic tissue in 6 (55%) of 11 cases, and PMS2 deficiency was the most common. In 1 of these 6 cases, MSH6 deficiency was shown as germline by NGS. **Conclusion:** CMMRD and MMRD, are disorders with close relationship with pediatric high grade gliomas. Since CMMRD cases also may have cafe au lait spots, they should not be misdiagnosed as NF 1. Temozolomide induce more aggressive tumors in CMMRD ve MMRD, therefore its use is not suggested in those cases. Preliminary literature data advocate use of immunotherapy instead. All pediatric HGG cases should be evaluated for CMMRD and MMRD with molecular investigations to understand their biology.

https://doi.org/10.1016/j.htct.2021.10.1003

OP 37

IS METHYLATION STATUS SUBGROUPING REALLY A STRONG PROGNOSTIC FACTOR IN PEDIATRIC POSTERIOR FOSSA EPENDYMOMA?

Bahattin Tanrıkulu [1], Ayça Erşen Danyeli [2], Cengiz Canpolat [3], M. Memet Özek [4]

[1] *Acibadem University School of Medicine Department of Neurosurgery Division of Pediatric Neurosurgery*
[2] *Acibadem University School of Medicine Department of Pathology.*
[3] *Acibadem University Department Of Pediatrics, Division of Pediatric Hematology and Oncology*
[4] *Acibadem University Department of Neurosurgery Division of Pediatric Neurosurgery*

Objective: The effective treatment of posterior fossa ependymomas is surgery followed by radio-chemotherapy. Our aim is to evaluate the effects of sex, age, methylation subgrouping, extent of resection, radiation treatment (RT), MIB-1 index, grade, ATRX and H3K27M mutations on prognosis in pediatric patients with posterior fossa ependymoma (PFE). **Methodology:** This is a retrospective study. Forty-two children with PFE who had surgery in our institution between 1996 and 2018 were included. Formalin-fixed paraffin-embedded tumor samples were evaluated for H3K27me3 immunostaining, MIB-1 index, WHO grades, ATRX and H3K27M mutations.Samples with global H3K27me3 reduction were grouped as posterior fossa ependymoma group A (PFA), whereas tumor samples with H3K27me3 nuclear immunopositivity were grouped as posterior fossa ependymoma group B (PFB). **Results:** Mean age of patients was 4.4 years (range 0.71-14.51). Thirty-one patients (73.8%) were PFA, whereas 11 patients (26.2%) were PFB. WHO grades of PFAs were statistically higher in comparison to WHO grades of PFBs. There are no significant differences between PFAs and PFBs in terms of resection rates, disease recurrence and survival parameters.Patients with total surgical excisions had significantly better PFS and OS rates. **Conclusion:** Extent of surgical excision is the most important prognostic indicator in PFEs. Prognostic effect of methylation subgrouping may be minimized with more aggressive surgical strategy in PFAs.

https://doi.org/10.1016/j.htct.2021.10.1004

NEUROBLASTOMA

OP 38

NEUROBLASTOMA IN A CASE OF CONGENITAL ADRENAL HYPERPLASIA

Arzu Yazal Erdem, Sonay İncesoy Özdemir, Meriç Kamak Cihan, Derya Özyörük, Neriman Sarı, İnci Ergürhan İlhan

SBÜ Ankara Şehir Hastanesi, Çocuk Hastanesi

Case report: The majority of neuroblastomas are sporadic and not correlated with any specific constitutional germline chromosomal abnormality, inherited predisposition, or associated congenital anomalies. We report here a 1.5-year-old girl with a diagnosis of 21 hydroxylase deficiency and neuroblastoma. Neuroblastoma in a known case of congenital adrenal hyperplasia has rarely been reported. Based on our literature review, this is the fifth case report of congenital adrenal hyperplasia and neuroblastoma.

https://doi.org/10.1016/j.htct.2021.10.1005

BONE TUMOURS

OP 39

CAN SERUM KL-6 LEVEL BE USED AS A MARKER IN LUNG METASTASIS OF BONE SARCOMAS?

Turan BAYHAN [1], Neriman SARI [1], Selma ÇAKMAKCI [1], Seda ŞAHİN [1], Nazan ÇİLEDAĞ [2], İnci İLHAN [1]

[1] *Ankara City Hospital, Department of Pediatric Hematology and Oncology*