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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.

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OP 37

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

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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.

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NEUROBLASTOMA

OP 38

NEUROBLASTOMA IN A CASE OF CONGENITAL ADRENAL HYPERPLASIA

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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.

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BONE TUMOURS

OP 39

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

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Objective: Osteosarcoma and Ewing sarcoma are the most common bone sarcomas of the childhood. Kerbs von de Lungren 6 (KL-6) is a glycoprotein that is expressed on type 2 pneumocytes and bronchial epithelium. Serum KL-6 level can increase in many interstitial pulmonary diseas and lung cancers. Aim of the study is to evaluate the predictive value of serum KL-6 level on malign potential of pulmonary nodules in pediatric patients with bone sarcoma with pulmonary metastasis or with vague pulmonary nodules. **Methodology:** Blood samples were taken from patients with diagnosis of Ewing sarcoma or osteosarcoma at the time of diagnosis or first relapses. Control group was selected from 42 voluntary children without any chronic or acute diseases associated with lung. Serum of the blood samples were separated and frozen at -70 C° and KL-6 level was measured via ELISA method. Thorax computed tomography (CT) images of the patients were analyzed to interpret about pulmonary metastasis. **Results:** Total 47 patients were included in the study, 19 of the patients were with Ewing sarcoma and 28 with osteosarcoma.Thorax CT revealed pulmonary metastasis in 9 of the patients at first evaluation. KL-6 level of the these patients with pulmonary metastasis was greater than without metastasis (p;0.05) and control group (p;0.019). Patients with pulmonary nodule at any time had significantly higher serum KL-6 level at first evaluation than without metastasis (p; 0.04) and control group (p;0.017). **Conclusion:** In our study we found serum KL-6 level higher in patients with pulmonary nodules that relevant with pulmonary sarcoma metastasis than patients without metastasis and healthy control group. Our study also revealed that patients that had pulmonary metastasis during their follow-up also had higher KL-6 level at diagnosis. These results should be proven with more number of patients. Measuring KL-6 level may be used as a marker for early diagnosis of pulmonary sarcoma metastasis.

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RARE TUMOURS AND HISTIOCYTOSIS

OP 40

PROLONGED COVID-19 POSITIVITY AND CHEMOTHERAPY IN A PATIENT WITH NASOPHARENGEAL CARCINOMA

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Ankara Şehir Hastanesi

Case report: Nasopharyngeal carcinoma is a rare tumor that accounts for 1-3% of all childhood malignancies. A 16-year-old patient with refractory nasopharyngeal carcinoma, whose treatment has to be interrupted due to COVID-19 positivity.After 6 weeks because of disease progression, we started his chemotherapy altough he is still COVID-19 positive. We didn't see any complication. Prolonged COVID-19 positivity is thought to be associated with the infection of immortal malignant cells located in the nasopharynx

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OP 41

EVALUATION OF CLINICAL AND LABORATORY CHARACTERISTICS OF CHILDREN WITH RHABDOID TUMOR: A MULTICENTER STUDY

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Objective: Rhabdoid tumors, which are rare in childhood, are aggressive cancers. It can be particularly seen in 3 different anatomical regions, mostly in the central nervous system, kidneys, and soft tissue in early childhood. In this study, it was aimed to evaluate the clinical, radiological and pathological features of pediatric patients with rhabdoid tumors who were followed up and treated in 3 different pediatric oncology reference centers. **Methodology:** Erciyes University Faculty of Medicine, Kahramanmaraş Sütçü İmam University Faculty of Medicine, Health Practice and Research Hospital and Adana City Training and Research Hospital, 17 patients diagnosed with rhabdoid tumor between 2002-2021 were retrospectively analyzed. **Results:** Of the patients, 6 (35%) were female and 11 (65%) were male. Chemotherapy (Doxorubicin, Ifosfamide, Carboplatinum, Etoposide, Vincristine, Actinomycin-D, Cyclophosphamide) was administered to the patients at different times. Radiotherapy was applied to 8 (47%) of the patients. The tumor was in the brain in 8 (47%) of the patients, in the kidney in 4 (23%), in the skin in 4 (23%), and the liver in 1 (6%). **Conclusion:** In this study, the incidence of rhabdoid tumors was higher in males. This may be due to the small number of cases. The 2 years overall survival rates were 50% in brain tumors, 6% in kidney tumors, and 12% in others, according to tumor localization. The localization and stage of the tumor were determinants of the survival of the patients. More clinical studies are needed to improve survival and identify more effective treatment strategies in these tumors.

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PEDIATRIC LEUKEMIAS

OP 42

ACUTE ABDOMEN AND ITS OUTCOMES IN CHILDREN WITH ACUTE LEUKEMIA

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