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SYNOVIAL SARCOMA ARISING IN THE RETROMOLAR TRIGONE: A RARE PRESENTATION

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Case report A 33-year-old female presented with complaints of lesion in the left retromolar trigone. CT scan demonstrated a soft tissue mass in left buccal space extending into the left infratemporal region. Biopsy was positive for Synovial Sarcoma. She received chemotherapy followed by wide local excision.Treatment with a total of 60 Gy radiotherapy was given. Patient tolerated the treatment well and remained disease free for 4.5 years. She developed recurrence and received palliative radiation for bleeding and pain control.

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RARE TUMOURS AND HISTIOCYTOSIS

PP 36

ASSOCIATION OF NF-1 AND MOYAMOYA SYNDROME : CASE REPORT

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Introduction: Neurofibromatosis type 1 (NF-1) is the most prevalent autosomal dominant genetic disorder. NF-1 vasculopathy is a significant complication of the disease. It affects both arterial and venous blood vessels of all sizes. Also Moyamoya syndrome is a cerebral vasculopathy . It is rarely detected with NF-1 in the pediatric group. Herein, we report of a 5 year-old male with NF1 and moyamoya syndrome. **Case Report:** A 6-month-old baby boy was brought by his family with the complaint of brown spots on the body. On physical examination, cafe au lait spots on the body, subcutaneous nodule in the occipital area and hypotonicity were found. He was examined considering neurofibromatosis, one of the neurocutaneous diseases. Abdominal ultrasonography, brain MRI, echocardiyography, electroencephalography were normal. There was no pathological evidence in eye examination. In genetic tests, NF1 p.GIn2217 gene was found heterozygous positive. patient was followed up with annual brain MRI. Hamartomatous lesions in left putamen and left thalamus posterior were detected in brain MRI when the patient was two years old. There was no pathological evidence on norological physical excamination. When he was three years old thickening and enhancement of the right optic nerve was found in MRI due to possible optic glioma. Only in the left cerebellar hemisphere, two millimeter-sized hamartomatous lesions in the white matter were found to have newly developed. Both internal carotid arteries (ICA) are thinned from the supraclinoid segment. Middle cerebral artery (MCA) M1 segment on the right and anterior cerebral artery (ACA) A1 segment on the left could not be selected. Many thin collateral vascular structures were selected in these localizations and were found to be significant in terms of Moyamoya disease. Regression was detected in right optic glioma. No predisposing factor was found in the examinations of the patient for thrombosis. Acetyl salicylic acid prophylaxis was started. The patient was taken under neurosurgery follow-up for revascularization surgery. The follow-up and treatment continues. **Conclusion:** Association of NF-1 and Moyamoya syndrome is rare, but carries a potential risk of clinicoradiological progression. Closed monitoring of children with neurofibromatosis type 1 enables early diagnosis of moyamoya syndrome. Revascularization surgery may effective way to prevent progression of clinical symptoms, but long-term results require close follow-up studies.

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PP 37

EVALUATION AND MANAGEMENT OF THYROID NODULES AT A TERTIARY CARE PEDIATRIC CANCER CENTER IN TURKEY

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Objective: The object of this study is to search the characteristics of children and adolescents with thyroid nodules and analyze our institutional experience in the management of thyroid nodules.The complaints of these patients, physical examination findings, diagnostic features, results of radiologic researches, choise of the most appropriate modality to these patients' thyroid nodule assessment, and the management of the pathology results were revealed. **Methodology:** Patients who applied to the pediatric endocrinology or oncology outpatient clinic of Ankara City Hospital with the diagnosis of thyroid nodule were examined. All patients who has pathology result as benign, atypia of undetermined significance/follicular lesion of undetermined significance (AUS/FLUS), follicular neoplasm/ suspicious for a follicular

neoplasm (FN/SFN), suspicious for malignancy (SM), and malignant were searched. **Results:** A total of 130 patients presented with thyroid nodules. Female male ratio was 1,95:1. The youngest patient was 68 months old. At admission there was no goiter in 71.5% of the patients on physical examination. Of all patients 36% of them underwent fine needle aspiration biopsy and 8 of the 76 patients who underwent biopsy were diagnosed with papillary thyroid cancer. One patient diagnosed with follicular thyroid cancer Patients that diagnosed cancer, 4.6% of them treated with radioactive iodine. **Conclusion:** Although most pediatric thyroid nodules are benign, distinguishing benign from malignant lesions is crucial. Interdepartmental communication and competence are very important in the follow-up of patients with thyroid nodules. Because of an increased risk of cancer in the pediatric population, diagnostic and therapeutic procedures for pediatrics need further research including multicenter studies to attain universal consensus regarding the diagnosis and management.

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TUMOR BIOLOGY, IMMUNOLOGY AND IMMUNOTHERAPY

PP38

TRAMETINIB EXPERIENCE IN A BRAF P.N 486 _P490DEL MUTATION POSITIVE LANGERHANS CELL HISTIOCYTOSIS

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Case report In Langerhans cell histiocytosis thyroid involvement is rarely seen. Here, we would like to present a 12-year-old male patient with lung, external auditory canal skin and lymph node involvement in diagnosis. Disease relapse occurred with thyroid involvement 19 months after remission. In molecular analysis, BRAF p.N 486 _P490del was detected and he reccived MEK inhibitor Trametinib monotherapy. He is still in remission for 16 months.

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PP 39

SERUM TOTAL OXIDANT AND ANTIOXIDANT STATUS IN CHILDREN WITH CANCER

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Objective: Oxidative stress has a potential role in carcinogenesis. Antioxidant enzymes have neutralizing effect both on cancer initiation, and progression. We aimed to assess the oxidant and antioxidant levels of pediatric cancer patients and to compare the levels in healthy controls. **Methodology:** The study involved 105 pediatric cancer patients (40 undergoing chemotherapy, 65 survivors) and 40 healthy children. The serum total oxidant status (TOS) and total antioxidant status (TAS) were measured. **Results:** The TOS and oxidative stress index were lower in pediatric cancer patients compared to the levels in the controls (3.73±1.35 vs. 4.21±1.72 μmol/L; p=0.08; 0.20±0.07 vs. 0.26±0.10; p=0.001, respectively). The mean serum TAS level was higher in patient groups compared to the level in the control (1.87±0.48 vs. 1.63± 0.32 mmol/L, p=0.001). The TAS level of children with cancer in survivors was still found to be significantly higher compared to the levels in the control group (1.85±0.45 vs. 1.63± 0.32 mmol/L, p=0.005). Radiotherapy, surgery, relapsed disease, presence of metastases and receiving enteral nutritional support caused no change in the TAS/TOS level. **Conclusion:** It has been revealed for the first time that the serum total antioxidant level increased during cancer treatment and didn't normalize after cessation of therapy for a long time.

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