

history of thromboembolism. In our 1-year follow-up, the hemoglobin value was 17-17.5 g/dL. **Conclusion:** More than a hundred globin gene mutations associated with erythrocytosis have been described. Hemoglobin Andrew-Minneapolis mutation is one of them. Hemoglobin's affinity for oxygen has increased and EPO level is normal/increased. Due to the low number of cases, treatment recommendations were prepared based on polycythemia vera guidelines. Patients should be closely monitored in terms of hyperviscosity and thromboembolism, aspirin prophylaxis and phlebotomy are recommended according to symptoms. While investigating the etiology of polycythemia, hemoglobin electrophoresis is necessary, although it is very rare.

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## LYMPHOMAS

PP 79

### THE SMALLEST PRIMARY BONE LYMPHOMA

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**Case report:** Primary lymphoma of bone (PLB) is a rare malignant condition with lymphocytic infiltration of the bone; it accounts for 2–3% of all primary bone tumours in adults and children. Here we report a little girl with isolated PLB of B cell lineage focussing on diagnosis, evaluation and treatment strategy. Our case can help to get acquaintance with PBL, it should be taken into consideration as a different diagnosis for osteolytic lesions of bone.

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PP 80

### MRI FINDINGS OF BONE MARROW AT THE BEGINNING OF LEUKEMIA

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**Case report:** Pediatric ALL/lymphoma (LBL) is a clonal hematopoietic stem cell disorder which's highly aggressive. There is an overlap between ALL and LBL which shouldn't cause delay in the diagnosis of each other. We'll describe a patient who presented with leukemia symptoms such as fever, bone pain, who didn't have obvious atypical cells in his peripheral smear, BM aspiration and involvement in scintigraphy but had diffuse bone marrow (BM) involvement in the lower extremities in his MRI. BM biopsy showed ALL/LBL.

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PP 81

### THREE CASES WITH BURKITT LYMPHOMA PRESENTING WITH CHOLESTASIS

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**Case report:** Cholestasis secondary to neoplasm is rare in children. It is also rare in Burkitt lymphoma and may be cause to treatment delay. We report 3 cases diagnosed with Burkitt lymphoma with cholestasis. All patients had jaundice and high direct bilirubin levels. They were given LMB chemotherapy protocol. After COP chemotherapy, cholestasis disappeared rapidly in all patients. In conclusion, cholestasis at initial resolves rapidly with chemotherapy despite high liver function tests in Burkitt lymphoma.

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## BRAIN TUMOURS

PP 82

### HIGH GRADE GLIOMA OF CENTRAL NERVOUS SYSTEM: SINGLE CENTER TREATMENT EXPERIENCE

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**Objective:** To evaluate characteristics and treatment responses of patients with high grade gliomas (HGG) in our center. Medical files of patients with malignant CNS tumors between 1987-2020 were analyzed retrospectively. There were 44 patients with HGG. **Case report:** Diagnosis of patients as follows: 21 pons glioma, 2 anaplastic astrocytoma, 11 anaplastic ependimoma, 7 glioblastoma multiforme, 1 glioblastoma, 2 gliomatosis cerebri. The median age at diagnosis was 6,5 yrs (7 – 17 yrs), M/F:25/19. Age distribution: <5 yrs 12 patients, 5-10 yrs 18 patients, 10-18 yrs 14 patients. The most frequent complaints for pons gliomas: cranial nerve paralysis

(52%), visual impairment (48%), headache (38%), power loss (43%) and speech disorder (30%). **Methodology:** Surgery was performed to extrinsic component of mass in 3 patients of pons gliomas. For other HGG: 7 subtotal resection and 16 gross total resection had performed. 7 patients died before RT. And other 37 patients received radiotherapy. RT total doses varied between 50-60 Gy. 7 patients were not received chemotherapy, 3 of them died before chemo, and others received only RT. For other HGGs, platin based regimens used for the first line treatment. Temozolamide, bevacizumab, irinotecan as the other options. **Results:** Median progression free survival time was 6 mos (2 weeks-25 mos) for pons gliomas, for other gliomas median progression free survival time was 14 mos (0-74 mos). For pons gliomas: Event free survival rate for 6 mos was 75%, for one year 17%; one year, 18 mos, and two years overall survival rates were 84%, 52% and 10% respectively. For other HGGs: Event free survival rate for one year and two years were 57% and 17% respectively. One year and two years overall survival rates were 73% and 36% respectively. **Conclusion:** High grade glioma is a group of tumors in which still the helplessness experienced in treatment. Despite radiotherapy and chemotherapy, prognosis is very poor. The progression free and overall survival rates of patients were similar to literature. With new developments in molecular pathology, as the use of molecular target therapies, the progression free survival rates newly will improve.

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PP 83

#### ONC 201 PRACTICE IN DIFFUSE MIDLINE GLIOMA (H3K27M MUTANT)

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**Objective:** Diffuse Midline Gliomas (DMG), H3 K27M-mutant have the poorest prognosis among all pediatric high-grade gliomas, with a median survival of 9-11 months. Although radiotherapy (RT) is standard treatment for these tumors, unfortunately there has been no approved and effective treatment which completely diminishes the tumor yet. In our clinic, we started an up-to-date approach to manage DMG, which is adjuvant fractionated external beam radiotherapy along with ONC 201 after tissue diagnosis. **Methodology:** Between January 2016 and June 2021, a total of 11 patients with H3 K27M-mutant diffuse midline glioma, diagnosis confirmed by Next-Generation Sequencing

(NGS) were enrolled in study. All patients received ONC201 orally once a week following radiotherapy. Safety, and radiological evaluations were regularly assessed every 12 weeks. **Results:** Among the 11 patients, the median age of diagnosis was 5. Seven (63.6%) patients were male and 4 (36.4%) were female. Primary lesions were localized in the pons in 5 (45.5%) patients, unilateral thalamus (2 on the left, 1 on the right) in 3 (27.3%) patients, bilateral thalamus in 2 (18.2%) patients, and temporo-insular in 1 patient (9.1%). Median progression-free interval was 10 months and median overall survival was 16 months. **Conclusion:** Diffuse midline glioma has dismal prognosis. None of the treatment options made any dramatic changes in disease course during last 30 years. In our series, diffuse midline glioma patients who had ONC201 tend to have few months more progression free and overall survival (16 vs 11 months) in comparison to patients who had classical treatment in literature. As a neurooncology team, we strongly advocate to obtain tissue samples from diffuse tumors, to establish definite diagnosis and to perform NGS

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#### NEUROBLASTOMA

PP 84

#### PARATESTICULAR INVOLVEMENT IN NEUROBLASTOMA

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**Case report:** Neuroblastoma is 7-10% of all pediatric cancer cases. Primary testicular and paratesticular neuroblastoma is very rare in the literature. We aimed to present our experience with a 4-year-old patient with an abdominal and right paratesticular mass. The patient's imaging revealed extensive lung and bone metastases. In the diagnostic biopsy, the primary tumour consistent with poorly differentiated neuroblastoma and the right paratesticular mass biopsy revealed neuroblastoma metastasis.

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#### BONE TUMOURS

PP 85

#### PRIMARY EWING'S SARCOMA OF SPHENOID BONE EXTENDING TO BRAINSTEM; AN ORDINARY TUMOUR AT AN EXTRAORDINARY LOCATION AND INVOLVEMENT

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Ankara Şehir Hastanesi