The goal is to report our center experience treating a 12-year old child with unresectable optochiasmatic pilocytic astrocytoma using BRAF V600E inhibitor monotherapy.

Low grade gliomas (LGGs) are the most frequent pediatric tumors of the central nervous system. The outcome of the treated children is satisfactory, albeit at the expense of the poor quality of life due to neurologic impairment linked to the tumor localization and multimodal oncologic treatment, including chemotherapy application. Cytostatic agents are used with the aim of achieving better disease control and increasing survival.

In the modern era of pediatric oncology detecting BRAF V600E mutation in histopathological samples has become a standard procedure which enables BRAF inhibitor therapy commencement. LGGs are stable monogenetic tumors characterized by activated MAPK pathway during all the phases of the illness making these drugs an attractive therapeutic option in relapsed disease.

Taking into account tumor localization, in our patient, only partial surgical reduction of the tumor mass was feasible and followed by chemotherapy administration. Unfortunately disease relapse ensued requiring new neurosurgical procedure, again without the possibility of achieving a complete resection. After BRAF V600E mutation has been found in resected material, a compassionate use of dabrafenib has been initiated.

In our patient oral BRAF inhibitor monotherapy of incompletely resected low grade glioma was easy to implement, with good medication tolerance, resulting in satisfactory disease control and good quality of life.

306

ACUTE LYMPHOBLASTIC LEUKEMIA IN CHILDREN: 5-YEAR EXPERIENCE OF A SINGLE NATIONAL PEDIATRIC ONCOLOGY CENTRE

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Study aimed to analyze epidemiological and clinical characteristics, hematologic, immunophenotypic and cytogenetic parameters of paediatric acute lymphoblastic leukemia cases, aged 1 year and above. Acute toxicities and treatment responsiveness were also studied.

Medical records of all children treated for acute lymphoblastic leukemia at the Department of Oncology and Haematology in Children's Hospital Zagreb from beginning of January 2015 to the end of December 2019 were reviewed.

Infants aged less than a year were excluded.

: In five-year period 28 children were treated for acute lymphoblastic leukemia, of whom 2 infants. Most cases of ALL were diagnosed in 2018 (N=10), and the least in the previous year (N=2). Out of 26 patients 16 were female (61.54%), average age 5.25 years. Median number of leukocytes at the time of diagnosis was $35.36 \times 10^9/L$, hemoglobin level 83.96 g/L and platelet count $120.42 \times 10^9/L$. The most represented immunophenotype was „common’

(73.07%), T phenotype present in only one child. 26.92% of blood marrow samples were hyperdiploid by cytogenetic testing and in two samples hypodiploidy was verified. One child had bcr/abl positive (Ph+) disease.

All 26 children were treated according to ALL IC-BFM 2009 protocol, 12 (71.06%) belonging to intermediate risk group, while standard and high-risk groups were evenly

represented (N=7). 25 children achieved early complete remission (by the end of induction), 3 were eligible for allogeneic stem cell transplant (1 Ph+, 1 resistant, 1 relapse), that was performed accordingly. While all children were faced with febrile neutropenia at some point of the treatment, 7 (26.92%) were treated for microbiologically proven sepsis. Four patients were diagnosed with clinically-radiologically verified pulmonary fungal disease. Acute neurotoxicity was observed in 7 children (seizures, ischemia, PRES). Thromboembolic events were registered in 4 (15.38%) patients. Two children (7.69%) died during intensive chemotherapy; 2-year old boy in induction therapy due to septic shock, pancreatitis and multiorgan failure; 1.5-year old girl due to the same reasons at the beginning of re-induction. No further deaths were observed; all 24 children are alive and in remission.

Acute lymphoblastic leukemia is the most common childhood malignancy, occurring in early age, predominantly of „common’ immunophenotype and most frequently classified as intermediate risk. Serious adverse events during intensive chemotherapy are frequent but with adequate management usually successfully resolved. Survival rates are excellent and have reached more than 90% in developed countries. Our results are completely comparable with literature data.

307

HEPATIC SINUSOIDAL OBSTRUCTION SYNDROME IN CHILDREN TREATED FOR SOLID MALIGNANT TUMORS – DIAGNOSIS AND TREATMENT

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The aim of this report is to describe our center's experience regarding diagnosis and treatment of hepatic sinusoidal obstruction syndrome (SOS) in children treated for malignant solid tumor.

retrospective analysis of hospital records in 2014-2021 period was conducted. Data on patients treated for malignant solid tumor who developed hepatic SOS were extracted and additionally analyzed, with the focus on the type and staging of the tumor, oncologic treatment undertaken, SOS diagnostic criteria and therapy.

a total of 5 patients (1 boy, 4 girls), aged between 19 months and 8 years, developed hepatic SOS. Two patients suffered from nephroblastoma and one from nephroblastomatosis. Two patients (Ewing sarcoma, high risk neuroblastoma) developed SOS as a complication of autologous hematopoietic stem cell transplantation, during which both received abundant supportive care.

All patients had hepatomegaly, ascites, weight gain (>5%) and thrombocytopenia. Hyperbilirubinemia was registered in only one patient.

Clinical criteria were supported by radiological investigations, mainly abdominal ultrasound with Doppler. In one patient liver biopsy was performed due to the suspicion of metastatic liver disease.

Risk factors identified among our patients were cytostatic agents (actinomycin-D, busulfan), previous radiation therapy and supportive care (total parenteral nutrition, antifungal