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PO054/#1571 | Publication Topic: *AS05 - SIOP Scientific Program/AS05.c - Lymphomas*

Successful Treatment of an Atypical Case of Cd10+ Disseminated Marginal Zone Lymphoma in a Child: Case Report and Review of the Literature

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Background and Aims: Marginal Zone Lymphoma (MZL) in children is extremely rare and distinct from adult cases. Management guidelines for disseminated MZL in children are lacking and risk of relapse is high. Here we describe an unusual case of disseminated CD10+ MZL in a child with aberrant clinicopathology findings, and its successful treatment.
Methods: Case report and literature review.
Results: A previously healthy 17-year-old girl presented with 6 months of severe fatigue, progressive abdominal pain, pallor, enlarged lymph nodes (LN) and significant splenomegaly. Initial lymphocytosis at 19.3 x 10*9/L rapidly quadrupled while anemia remained stable. PET-CT showed extensive splenic, LN and bone marrow involvement. There was no central nervous system spread. Pathology from LN biopsy revealed an unusual indolent mature B-cell lymphoma, CD10+ BCL2+ by immunohistochemistry, BCL6 and BCL2 negative by FISH, and with plasmacytic differentiation, most consistent with disseminated MZL. Patient had Naive B cell lymphopenia prior to treatment, no autoimmunity disease, no HCV or EBV infection and germline panel for inborn error of immunity was negative. Rituximab 375mg/m^2 4 weekly doses was used for cytoreduction. Despite giving the first dose over 3 days with premedication to decrease risk of cytokine release syndrome (CRS), patient experienced significant CRS after the 2mg of Rituximab. She tolerated other doses well and reached partial response. She received 6 cycles of R-CVP and achieved complete remission (CR) after cycle 3. Due to high risk of relapse in MZL during the first 2 years after treatment, she completed 2 years of maintenance with Rituximab q2months. She continues to be in CR now 2 months after the end of treatment.
Conclusions: Despite the challenging diagnostic of this atypical CD10+ disseminated MZL in a child, treatment including cytoreduction with Rituximab, R-CVP chemotherapy followed by Rituximab maintenance was well tolerated and successful.

PO055/#860 | Publication Topic: *AS05 - SIOP Scientific Program/AS05.c - Lymphomas*

A Single Center Experience of Pediatric Burkitt Lymphoma

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Background and Aims: To evaluate the demographic, clinic, pathological characteristics and treatment responses of patients diagnosed with Burkitt lymphoma (BL) in our institution.
Methods: The files of 40 patients who were followed up and treated with a diagnosis of BL in our center between 1988 and 2023 were retrospectively reviewed. Demographic data, clinical features, treatment outcomes, event-free survival (EFS) and overall survival (OS) rates of the patients were evaluated.
Results: Among 137 patients diagnosed with non-Hodgkin lymphoma,40 were diagnosed with Burkitt lymphoma. Files of 39 of these patients were eligible. The median age at diagnosis was 8 years (range: 2–17 years), with a male-to-female ratio of 3.3. Twenty-eight percent of patients (n:11) were <5 years old,38% (n:15) were between 5-10 years old, and 33% (n:13) were over 10 years old. The median time from symptom onset to diagnosis was 3 weeks (range:0–24 weeks), with abdominal pain and abdominal swelling being the most common complaints. Tumor lysis syndrome was present at diagnosis in 36% of patients. Stage distribution was as follows: Stage II 13% (n:5), Stage III 72% (n:28), and Stage IV 15% (n:6). When looking at risk groups, 14 patients (36%) were classified as risk group 2, 22 patients (56.3%) as risk group 3, and 3 patients (7.7%) as risk group 4.Twenty-eight percent of patients had a primary site except abdomen. After 2019, all patients were treated according to the NHL BFM 2012 protocol. Rituximab was used in 33% of patients (13/39). It was noteworthy that five of 13 patients receiving rituximab developed secondary immunodeficiency requiring IVIG. The median follow-up time was 3 years (range:4 months–20 years). The five-year event-free survival (EFS) was 88%, and the ten-year EFS was 81%. The five and ten-year overall survival (OS) rates were both 85%.
Conclusions: Our treatment results were acceptable and same as the literature. Despite chemotherapy and targeted therapies, the chance of survival is low in patients with progressive disease or relapse.

PO056/#277 | Publication Topic: *AS05 - SIOP Scientific Program/AS05.c - Lymphomas*

Granulocyte Colony Stimulating Factor as Primary Prophylaxis to Reduce Acute Mortality During Induction Chemotherapy in Childhood B Cell Non-Hodgkin Lymphoma

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Background and Aims: Pediatric NHL achieve complete remission in high-income countries; however, mortality is high in low income countries due to Infections up to 19.7%. Use of Granulocyte Colony Stimulating Factor has shown to reduce the duration and severity of neutropenia. Our study aims to assess a reduction in acute mortality with the use of GCSF as primary prophylaxis in NHL and a reduction in hospital admission days with febrile neutropenia

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