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CASE REPORT OF REFRACTORY LARGE B-CELL LYMPHOMA WITH IRF4 REARRANGEMENT

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INTRODUCTION

Large B-cell lymphoma with IRF4 rearrangement (IRF4+ LBCL) presents in young adulthood with involvement of the head and neck region. There is insufficient data on the association of concomitant cytogenetic abnormalities with prognosis.

AIM

We aimed to present a case of refractory IRF4+ LBCL..

CASE

A 20-year-old male patient with no known history of chronic disease or medication, with right cervical swelling which had been growing slowly for 4 months. Physical examination revealed bilateral hypertrophic tonsils with prominent right side. Laboratory analysis revealed no significancy in complete blood count, biochemical analysis and peripheral blood smear. The biopsy of the palatine tonsil was performed.

Microscopic examination demonstrated follicular infiltration of neoplastic lymphoid cells with medium-sized blastic morphology. Immunohistochemical examination revealed strong expression of pan-B markers (CD20 CD79a, PAX-5) as well as CD10, bcl-2, bcl-6 and MUM-1. Ki-67 proliferation index was 75% and myc expression was negative with immunohistochemistry. NGS analysis resulted as IRF-4 mutation, as well as BABAM2::JAK2 fusion and bcl-2 mutation that were supportive for IRF4+LBCL.

Positron emission tomography-computed tomography (PET/CT) performed for staging revealed a 3.9x2.8 cm asymmetric hypermetabolic soft tissue (SUVmax:19.2) at the level of the right palatine tonsil, and hypermetabolic activity (SUV max 14.2) at the level of the left palatine tonsil. R-CHOP chemotherapy protocol was initiated. At the end of 4 cycles, a partial response was observed; and the treatment was completed to 6 cycles. However, at the end of treatment, PET/CT showed an increase in the size and metabolic activity of the primary lesion. Repeat biopsy was resulted the same morphology. Salvage chemotherapy followed by autologous stem cell transplantation was planned.

CONCLUSIONS

In our knowledge, age has been defined as prognostic parameter for IRF-4+LBCL, Thus, more complex molecular abnormalities have been reported in the adult age group. JAK2 fusion abnormalities are associated with treatment resistance in myeloid and lymphoblastic leukemias, but there is no data on their contribution to the clinical course in IRF4+ LBCL. In our case with primary refractory course, it was considered that additional JAK fusion abnomality may have a negative effect on the clinical course.

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