CASE REPORT

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Follicular Lymphoma With Leukemic Phase: Case Report

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Abstract:
Follicular lymphoma (FL) originating from germinal center B cells constitutes 35% of all non-Hodgkin lymphomas. Patients usually present with asymptomatic peripheral lymphadenopathy (LAP) at the time of diagnosis and approximately 70% of them have bone marrow involvement. Leukemic phase of FL is a rare condition and associated with a poor prognosis.
In this report we presented a 29-year-old male patient with a diagnosis of leukemic phase of FL which is a rare condition. We initiated R-CHOP chemotherapy and he is still being followed in complete remission after 6 course.
Leukemic phase is a rare clinical presentation of FL and it can be confused with chronic lymphocytic leukemia. Flowcytometric examination is very valuable in diagnosis. Leukemic phase of FL is associated with poor prognosis independent of FLIPI score. Therefore detecting circulating leukemic cells in patients with FL is necessary.

Keywords: Case Reports, Leukemic Infiltration, Lymphocytosis, Follicular Lymphoma, Prognostic Factors

Introduction

Follicular lymphoma (FL) originating from germinal center B cells constitutes 35% of all non-Hodgkin lymphomas and 70% of slow progressive (indolent) lymphomas. Patients usually present with asymptomatic peripheral lymphadenopathy (LAP) at the time of diagnosis and approximately 70% of them have bone marrow involvement (1). A hallmark of the disease is the chromosomal translocation t (14.18) contributing to overexpression of the antiapoptotic protein BCL2. Despite commonly advanced stage at presentation, median survival now exceeds 15 years and is improving with the incorporation of anti-CD20 monoclonal antibodies and better supportive care into therapy (2). The leukemic phase of FL is rare and associated with a poor prognosis (3). In this report, we presented a case we followed up with the diagnosis of the leukemic phase of FL.

Case Report

A 29-year-old male patient presented with complaints of abdominal pain and neck swelling for a week. Cervical, axillary lymphadenopathy and hepatosplenomegaly were found on physical examination. The laboratory results were as follows: White Blood Cell:109 ×109/L, Absolute Neutrophil Count: 8,9 ×109/L, Hemoglobin:12.2 g/dl, Platelet:132 ×109/L, Lactate dehydrogenase:289 U/L. Lymphoid cells with a condensed chromatin and narrow cytoplasm were observed in the peripheral smear (Figure 1). Hepatosplenomegaly, cervical, axillary and intraabdominal lymphadenopathies were found in ultrasonography. In peripheral flowcytometric examination; CD45, CD10, CD19, CD20, CD22, CD79A, FMC7 were positive and CD5, CD23 were negative. Bone marrow biopsy was performed. Bone marrow aspiration smear was in hypercellular appearance and about 70% large lymphoid cells with a condensed chromatin and narrow cytoplasm were observed (Figure 2). Immune morphological findings in bone marrow pathology were consistent with follicular lymphoma infiltration (Ki67 index 20%). In FISH analysis, 55% fusion was observed in the IGH/BCL2 [t (14;18)] gene. Axillary lymph node biopsy was performed. The pathology result was interpreted as grade 2 follicular lymphoma. Thoracic-abdominopelvic CT was performed. Conglomerated lymphadenopathies in mediastinal, axillary, intraabdominal localization detected and spleen size was 30 cm, liver size was 18 cm. The patient was considered to be stage 4, grade 2

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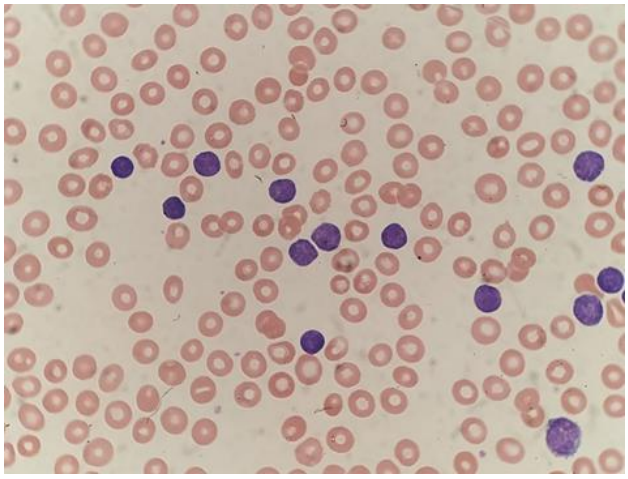

Fig. 1. Peripheral Smear; Lymphoid Cells With A Condensed Chromatin and Narrow Cytoplasm

follicular lymphoma in the leukemic phase. The FLIPI score was 3 (high risk). R-CHOP chemotherapy was initiated. There was a complete response after 3 cycles. 6 courses chemotherapy were completed and Rituximab maintenance was initiated. The patient is still being followed in complete remission.

Discussion

Follicular lymphoma is the second most common type of lymphoma and the most common indolent lymphoma. Although the leukemic phase of FL is a rare clinical presentation, it was reported in a previous Japanese study that FL in the leukemic phase was observed more frequently (64%) in Asia compared to western countries (4). Although it can be confused with chronic lymphocytic leukemia in differential diagnosis, it can be distinguished as immunophenotypically. There are case reports and limited case series in the literature. Beltran et al was reported a case series of seven patients. In this study the median age was 57, the patients had a high (4-5) FLIPI score and FL in the leukemic phase was associated with poor prognosis (5). Although there was diffuse LAP at the time of presentation in our case, it has been reported in previous publications that FL in the leukemic phase can be seen with or without lymph node involvement. Al-Nawakil et al. defined 10 FL patients who were in the leukemic phase at the time of diagnosis and 6 of these patients had lymph node involvement, 4 of them had pure FL cell leukemia. Patients without lymph node involvement were associated with a more indolent clinic. However, it was reported that leukemic

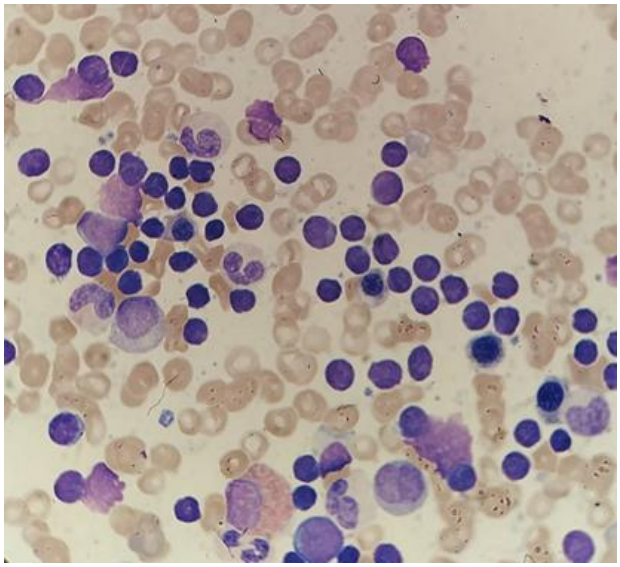

Fig. 2. Bone Marrow Aspiration; 70% Large Lymphoid Cells With A Condensed Chromatin And Narrow Cytoplasm

phase FL may have a different biological behavior and should be considered as a prognostic marker as it may affect the prognosis of these patients (6). There is no standard treatment as few cases have been reported. RCHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone), RCVP (rituximab, cyclophosphamide, vindesine, prednisone), RAEC (rituximab, adriamycin, etoposide, cyclophosphamide) treatments were applied to these patients in the literature (6). We gave our patient R-CHOP chemotherapy, which is a more preferred regimen. Although it is not common, the leukemic phase of FL should be considered in a patient presenting with extensive LAP, HSM, leukocytosis, lymphocytosis as in our case. In addition, the investigation of circulating leukemic cells in patients with FL is important as it may be a prognostic marker.

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