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Large T cell granular lymphocyte leukemia with secondary HLH mimicked severe SLE

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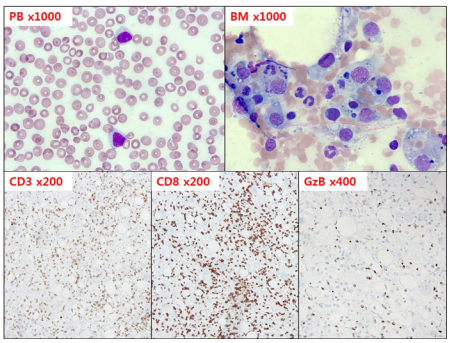
Introduction: Large granular lymphocyte (LGL) leukemia is a rare chronic lymphoproliferative disorder known for the indolent clinical course. Here, we report a highly unusual case of LGL leukemia with secondary hemophagocytic lymphohistiocytosis (HLH) that mimicked systemic lupus erythematosus (SLE). **Case Description:** A 46-year-old previously healthy female presented with fever, dyspnea, and left pleuritic chest pain for 3 months. Erythematous rash was found on her face and chest, and lymph nodes (LNs) were palpable in both axilla. Laboratory test results were as followings: Hb 7.1g/L, WBC 6,890/μL, platelet 92 x 10^3/μL, ANA titer 1:80, and spot urine PCR 2.0g/g. Chest CT showed pericardial and pleural effusion. She met 5 of 11 ACR criteria of SLE. However, some features were unfitted to SLE, including normal ESR compared to high CRP, the absence of lymphopenia, and almost normal C3/C4 levels. Other hidden conditions mimicking SLE were speculated, and therefore LN biopsy was performed. High-dose steroid therapy was initiated (day 1) for symptom relief after the biopsy. Despite the use of steroids, her symptoms were only minimally improved, and the bicytopenia persisted. On day 3, serum ferritin level was elevated to 230,075ng/ml and TG was 344mg/dL, suggestive of the concomitant HLH. LN biopsy revealed atypical lymphoid hyperplasia. Emerging evidence pointed to hidden hematologic malignancy, leading to bone marrow (BM) exam on day 6. On BM aspiration, large granular lymphocytes with cytoplasmic granules and abundant cytoplasm were counted up to 34% of total nucleated cells. BM biopsy showed extensive infiltration of CD3(+), CD8(+), and granzyme B(+) cells and histiocytes with hemophagocytic activity (Figure). The final diagnosis was adult T-cell LGL leukemia with secondary HLH. She was discharged on day 26 as symptoms and cytopenia were gradually improved. Her disease has been well-controlled with oral cyclophosphamide with tapering of steroid. **Discussion:** LGL leukemia can present as autoimmune disorders. In this case, it mimicked severe SLE and challenged the initial diagnosis. Atypical features of SLE should be further evaluated for hidden LGL leukemia



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Clinical Impact of Pretreatment Albumin to Globulin Ratio in Patients with DLBCL Treated with R-CHOP

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Background/Aims: We evaluated the clinical implications of the albumin to globulin ratio (AGR) in patients with diffuse large B-cell lymphoma (DLBCL) treated with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone (R-CHOP). **Methods:** Data of 232 patients with DLBCL treated with first-line R-CHOP from 2004 to 2017 were reviewed retrospectively. Patients with AGR values ≥ 1.22 and<1.22 were assigned to the high and low AGR groups, respectively. Treatment response, treatment-related toxicity, and survival were compared according to the AGR. **Results:** The complete response rate was significantly lower in the low AGR group than in the high AGR group (59.1% vs. 81.3%; p<0.001). Treatment-related mortality was also more frequent in the low AGR group than in the high AGR group (14.0% vs. 4.3%; p=0.009). The low AGR group (median overall survival [OS]=26.87 months; 95% confidence interval [CI]=4.19-49.55) showed a significant decrease in OS compared to the high AGR group (median OS=148.83 months; 95% CI=76.26-221.41; p<0.001; Figure.1-A). Progression-free survival (PFS) also decreased significantly in the low AGR group (median PFS=14.29 months; 95% CI=2.58-26.01) compared to the high AGR group (median PFS=148.83 months; 95% CI=76.21-221.45; p<0.001; Figure.1-B). In a multivariate analysis, low AGR was an independent poor prognostic factor for OS and PFS. **Conclusions:** Pretreatment AGR was useful for predicting treatment response, treatment-related toxicity, and prognosis in patients with DLBCL treated with R-CHOP. Further large prospective studies will be necessary to validate our findings.