

## A Case of T-cell Large Granular Lymphocytic Leukemia Presented as Pulmonary Hypertension

국립암센터 내과<sup>1</sup>, 국립암센터 진단검사의학과<sup>2</sup>

조은<sup>1</sup>, 심효은<sup>2</sup>, 정종현<sup>1</sup>

**Introduction:** T-cell large granular lymphocytic (LGL) leukemia is a rare form of chronic lymphoproliferative disorders, characterized by chronic inflammation and autoimmunity. It is diagnosed with evidence of an expanded clonal T-cell LGL population. LGL leukemia is known to be associated with various diseases such as autoimmune diseases, solid or hematopoietic neoplasms, and pulmonary hypertension.

**Case report:** A 49-year-old woman was referred to the Hematology Clinic of National Cancer Center for further evaluation of cytopenia. The previously healthy patient presented acute dyspnea which required admission to the intensive care unit. The complete blood count showed a white-cell count of 1,500 per microliter, a hemoglobin level of 11.2g per deciliter, and a platelet count of 94,000 per microliter. After the initial work-up, she was diagnosed with pulmonary hypertension and IgG-positive autoimmune hemolytic anemia (AIHA). Imaging studies revealed splenomegaly without increased FDG uptake and an enhancing lesion in the perianal area of which punch biopsy proved moderately differentiated squamous cell carcinoma. Peripheral blood smears showed large granular lymphocytes, and bone marrow biopsy revealed multinodular CD3+ lymphoid aggregates. TCR gene rearrangement was confirmed by the PCR assay of the bone marrow biopsy. The patient was diagnosed with anal cancer and T-cell LGL leukemia. She received radiotherapy for anal cancer and started weekly oral methotrexate and prednisolone as the first line treatment for T-cell LGL leukemia after the completion of planned radiotherapy. Although her disease was persistent with methotrexate and prednisolone for six months, pancytopenia was resolved with oral cyclosporine. Nowadays, her disease remains stable for about two years with oral cyclosporine.

**Conclusion:** This case highlights various initial presentations and associated diseases of T-cell LGL leukemia such as AIHA, anal cancer, and pulmonary hypertension. Especially in patients with cytopenia with uncertain causes who present other serious conditions, T-cell LGL leukemia should be considered as a possible diagnosis.

