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A CASE OF AUTOIMMUNE LYMPHOPROLIFERATIVE SYNDROME WITH PANCYTOPENIA AND SPONTANEOUS ICH

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Introduction: The autoimmune lymphoproliferative syndrome (ALPS) and ALPS-like syndrome are variable clinical conditions characterized by lymphoproliferative disease, autoimmune cytopenias, and susceptibility to malignancy. When patients have the same clinical findings but do not have abnormalities of FAS-mediated lymphocyte apoptosis are given the diagnosis of the ALPS-like syndrome. The major determinants of morbidity and mortality for patients with ALPS are the severity of the autoimmune disease, complications such as post-splenectomy sepsis and the presence of lymphoma. Therefore, early detection and treatment could be critical to prevent fatal deterioration in ALPS. Here we report a case of ALPS presenting FUO with pancytopenia and spontaneous ICH. **Case presentation:** A 79-year-old woman presented with Fever and bicytopenia (anemia, thrombocytopenia) that developed three weeks previously. She had spontaneous ICH 7 weeks ago. She had neurosurgical drainage. During the rehabilitation, she developed fever and thrombocytopenia. Despite the treatment of antibiotics, ampicillin/sulbactam and ertapenem, fever and cytopenia continued. One year before this admission, she was hospitalized several times for recurrent fever with thrombocytopenia. In the CT imaging, her neck nodes were enlarged, which were excised for biopsy, and the results were atypical T-cell lymphoproliferative lesion. She had had a bone marrow biopsy, and the results were normocellular marrow with a normal distribution of nucleated cells. The follo.w-up laboratory test showed Hb 9.6 g/dL, PLT 79 K/μL, D-Coombs (+; weakly), Polyclonal gammopathy on PEP(S), IgG 2309 mg/dL and Vit-B12 4896.25 pg/mL and 2.97% of dual negative (DN) T cell count. Under the diagnosis of ALPS-like syndrome, we treated with high dose steroids and mycophenolate mofetil, vital signs and blood cell counts were stabilized. **Conclusions:** ALPS is a rare disease that shares overlapping features with infection or hematologic disorder such as lymphoma. When a patient with autoimmune phenomena including hemolytic anemia and ITP does not respond to the treatment, the possibility of ALPS also should be considered.