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Acquired hemophilia an associated with Epstein–barr-virus-associated t/natural killer-cell lymphoproliferative disease: a case report

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Acquired hemophilia A (AHA) is a rare bleeding disorder caused by autoantibodies against factor VIII (FVIII). Hematological malignancies, especially lymphoid malignancies, are known to be underlying causes of AHA; however, thus far, there is no report of AHA associated with Epstein–Barr-virus-associated T/natural killer-cell lymphoproliferative disease (EBV-T/NK-LPD). Here, we present a case of AHA that developed during treatment for EBV-T/NK-LPD. A 69-year-old man visited our hospital because of general fatigue. Blood examination showed pancytopenia, and computed tomography revealed whole-body lymphadenopathy, but there were no findings indicating hematological malignancy from bone marrow aspiration and cervical lymph node biopsy. The level of EBV DNA in peripheral blood was extremely high, and he was diagnosed with EBV-T/NK-LPD. EBV-T/NK-LPD improved with prednisolone (PSL) administration. Seventeen months after starting treatment, the patient complained of back and right leg pain. At that time, he had been treated with low-dose PSL, and EBV-T/NK-LPD was well controlled. Imaging revealed hematoma of the right iliopsoas muscle. Prolonged activated partial thromboplastin time was the only abnormal finding in a screening coagulation test. FVIII coagulant activity was below detection limit, and FVIII inhibitor level was increased. From these results, he was diagnosed with AHA. A higher dose of PSL was administered, and, after 1 month of treatments, FVIII activity gradually increased, and FVIII inhibitor level became undetectable. Activated partial thromboplastin time also normalized, and complete remission was achieved and maintained for 13 months with low-dose PSL. During the treatments, EBV-T/NK-LPD was well controlled. It is speculated that proliferating lymphocytes interfere with normal immune functions and that abnormal autoantibodies are produced from those lymphocytes in patients with LPD. Therefore, we speculate that EBV-infected and proliferating monoclonal NK cells might have modulated the immune system and produced autoantibodies against FVIII, thus causing AHA in this patient with EBV-T/NK-LPD.

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