Repeated Epstein-Barr virus-associated hemophagocytic syndrome in a lupus nephritis patient.

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A 55-year-old Japanese female was diagnosed with systemic lupus erythematosus (SLE) and developed nephrotic syndrome. She was diagnosed with lupus nephritis by a percutaneous renal biopsy. She was treated with intravenous steroid pulse therapy twice, but it proved to be ineffective. She achieved a complete remission after intravenous cyclophosphamide pulse (CPAIV) therapy. Thereafter, her lupus nephritis was well controlled and demonstrated only a low activity. However, she suffered Epstein- Barr virus (EBV)-associated hemophagocytic syndrome (HPS) twice, and in each case she was treated with anticancer drugs and achieved a complete remission. This was a rare case of lupus nephritis who showed repeated EBV-associated HPS.