CASE REPORT

A Case of Epstein–Barr Virus-associated T/B Cell Lymphoproliferative Disease Successfully Treated with Rituximab, Cyclophosphamide, Vindesine and Prednisone

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ABSTRACT

Epstein–Barr virus (EBV)-associated lymphoproliferative diseases (LPDs) cover a wide range of lymphocytes disorders spanning B, T and natural killer cells. The classification of EBV-associated LPD is evolving. We report a case of a 63-year-old male patient with EBV-associated T/B cell LPD diagnosed by pathological biopsy of lymph nodes, which is a new type of EBV-associated LPD that has not been reported previously. Because of its rarity, standard treatment has not been established. This patient showed rapid remission of the disease after combination chemotherapy with rituximab, which provided evidence supporting the use of rituximab and chemotherapy combination in EBV-associated T/B cell LPD treatment.

Keywords: Epstein–Barr virus, lymphoproliferative disease, rituximab.

INTRODUCTION

Epstein–Barr virus (EBV)-associated lymphoproliferative diseases (LPDs) cover a wide range of lymphocytes disorders spanning B, T and natural killer (NK) cells, which has been classified in the 4th edition World Health Organization classification of tumours of haematopoietic and lymphoid tissues. The classification of EBV-LPD is evolving, including Burkitt lymphoma, age-related EBV+B-cell LPD, extranodal NK/T-cell lymphoma of nasal type, aggressive NK-cell leukemia, classic Hodgkin lymphoma, immunodeficiency-associated lymphoproliferative disorders, EBV-associated T- and NK-cell LPD, systemic EBV+T-cell LPD of childhood, and hydroavacciniforme-like lymphoma (1–3).

However, to our best knowledge, EBV-associated T/B cell LPD has not been reported previously. Here, we report a rare case of EBV-associated T/B cell LPD successfully treated with R-CVP (rituximab, cyclophosphamide, vindesine and prednisone).

CASE REPORT

A 63-year-old Chinese man was admitted to the Sixth Medical Center of Chinese PLA General Hospital at Beijing because of a cough and lymphadenectasis of neck in December 2013. Ultrasound revealed a 4.2 × 2.1 × 2.6 cm enlarged lymph node in his right neck. Contrast-enhanced computed tomography revealed enlarged lymph nodes in the para-aortic, mediastinal and abdominal areas. The EBV DNA load in the peripheral blood mononuclear cells (PBMCs) was 1260 000/106 PBMC. Biopsy of the enlarged lymph node in his right neck was made. Immunohistochemistry demonstrated that the cells were positive for EBV-encoded small RNA in situ hybridization, CD3, CD8, CXCL13, Bcl2, partially positive for CD20, CD4 and PD1. Thus, the histopathologic diagnosis of EBV-associated T/B cell LPD was made. The patient received four cycles of R-CVP chemotherapy and achieved complete response (CR). Then, we consolidated two cycles of R-CVP after CR and terminated the therapy. The patient was still alive with disease free until this paper was submitted.

DISCUSSION

To our knowledge, this is the first report of a patient with EBV-associated T/B cell LPD in long-term CR following R-CVP therapy, with no use of high-dose chemoradiotherapy in combination with haematologic stem-cell transplantation.

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Therapeutic approaches on EBV-associated LPD include high-dose immunoglobulin, interleukin-2, antiviral agents, interferon-α or interferon-γ, corticosteroids, rituximab, haematopoietic stem-cell transplantation, and EBV-specific T cells (4). However, these therapies generally have not been successful and relapses were common. Here we report a rare case with EBV-associated T/B cell LPD, which is a new type of EBV-associated LPD that has not been reported previously. Because of its rarity, standard treatment has not been established.

Rituximab is a chimeric mouse/human monoclonal antibody that specifically targets the CD20 antigen on the surface of normal and malignant human B cells, which has been widely used in B-cell lymphomas (5, 6). Rituximab in combination with CHOP (cyclophosphamide, doxorubicin, vincristine and prednisone) has already improved survival in elderly patients with aggressive non-Hodgkin's lymphoma (7). In terms of its safety and efficacy, our case provided evidence supporting the use of rituximab and chemotherapy combination in EBV-associated T/B cell LPD treatment.

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AUTHORS' NOTE

The authors confirm that there are no conflicts of interest.

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