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Case Report
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Human Immunodeficiency Virus-Negative Plasmablastic Lymphoma: A Case Report and Literature Review

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Abstract

Introduction: Plasmablastic lymphoma is a rare subtype of B cell lymphoma with poor prognosis. The pathogenesis and molecular features of this rare entity was largely unclear.

Case Report: We report a rare case of PBL in a 57-year-old immunocompetent male patient. The patient was admitted to our hospital because of multiple lymph node enlargement detected by computed tomography during the physical examination. The biopsy results confirmed the diagnosis of PBL. This patient achieved partial remission after DA-EPOCH chemotherapy and received ASCT as consolidation therapy. However, disease progression occurred and the patient died 2 months after ASCT.

Discussion: PBL is characterized by rapid disease progression and high mortality rates. More effective treatment regimen needs to be explored.

Conclusion: PBL was a highly invasive rare lymphoma with poor prognosis, it is urgent to develop new targets and agents.

Keywords: Plasmablastic lymphoma; HIV; ASCT

Introduction

Plasmablastic Lymphoma (PBL) is a rare and unique subtype of diffuse large B-cell lymphoma, which is highly aggressive and mainly occurs in Human Immunodeficiency Virus (HIV) positive patients. Delecluse first reported 16 cases of PBL, all of which occurred in the oral cavity, of which 15 cases were accompanied by HIV infection [1]. Later, it was found that PBL also occurred in people with normal immune function [2-4]. Our study demonstrated that patients with PBL were immunocompetent and the prognosis seems to be better than that reported by western countries [2]. The pathogenesis of PBL remains largely unknown and the prognosis was still poor and it is urgent to develop new targets and agents.

Here, we reported a male patient with HIV negative PBL with rapid progression after chemotherapy followed by ASCT.

Case Presentation

A 57-year-old Chinese male presented in November 2021 with multiple lymph nodes enlargement which was detected by computed tomography for lung during physical examination. No systematic symptom was observed and he was negative for HIV test. The patient was referred for 18F-FDG PET/CT examination and extensive 18F-FDG positive mass involving lymph nodes throughout the body (cervical, mediastinal and hilar) and pancreas were detected (Figure 1). No evidence of lymphoma was observed by bone marrow biopsy of the right iliac crest. A biopsy of the supraclavicular lymph node indicated a B cell non-Hodgkin lymphoma with plasmablastic morphology and immunohistochemistry results showed that CD38, Mum-1, CD30, Bcl-2, c-Myc were positive while CD20, CD79a, Bcl-6, CD10, CD138, CD30, CD138, PAX-5, TIA-1, CD4, CD8, CD7, CD43. The ki-67 expression was nearly 100% and EBV-Encoded small RNA (EBER)-1/2 was positive.

Laboratory examination was as follows: White blood cells 8500/μL, hemoglobin 13.5 g/dL, platelets 340,000/μL; the lactate dehydrogenase, serum calcium and immunoglobulin levels were at normal ranges; following the PET/CT and biopsy results, the patient received 5 cycles of DA-

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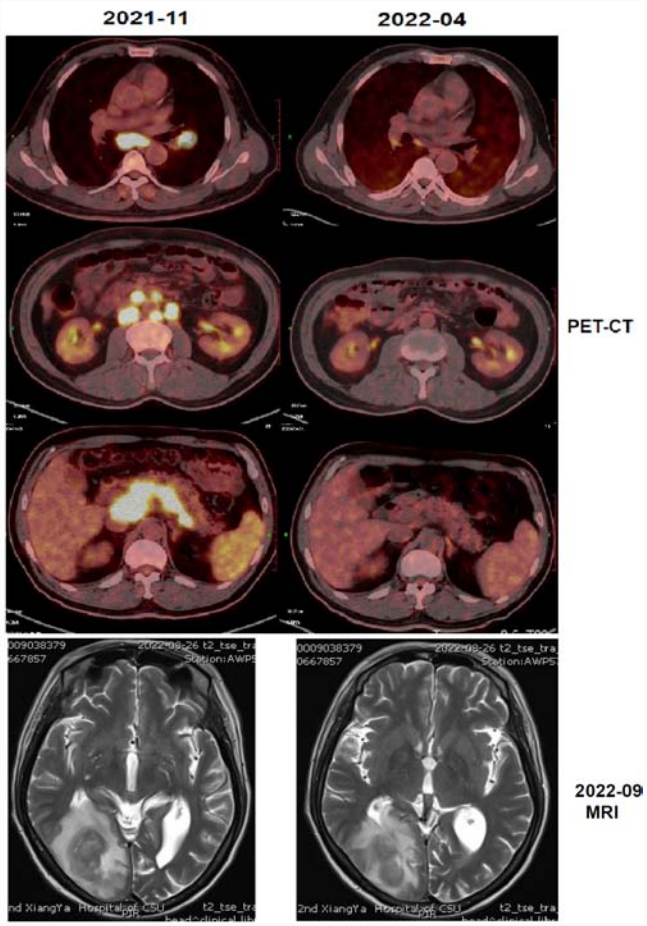

Figure 1: ^{18}F-FDG PET/CT examination and extensive ^{18}F-FDG positive mass involving lymph nodes throughout the body, brain metastases was revealed by MRI two months after ASCT.

EPOCH chemotherapy (cyclophosphamide, doxorubicin, vincristine, etoposide, prednisone). The metabolic activity decreased significantly and the patient achieved partial remission as determined by PET/CT (Figure 1). This patient received Autologous hematopoietic Stem Cell Transplantation (ASCT) as consolidation therapy after chemotherapy. However, disease progression occurred and brain metastases was revealed by MRI two months after ASCT (Figure 1). The patient died around three months after ASCT and the overall survival time was around 11 months.

Discussion

PBL was classified as a special subtype of Diffuse Large B-Cell Lymphoma (DLBCL) in the 2016 World Health Organization (WHO) classification of lymphoid and hematopoietic diseases. Although previous studies showed that PBL mainly occurred in the HIV positive population [5], recent research demonstrated that most of the reported cases of PBL in China were elderly with normal immune function [2].

The most common sites of HIV negative PBL were oral cavity, followed by gastrointestinal tract, lymph node and skin [2,4]. The pathomorphological features of PBL were unique, the tumor cells had immunocytoblast/plasmablastic morphology but showed plasma cell markers, with weakly expressed or no expression of mature B cell markers such as CD20, PAX-5, CD3, CD45. CD56 was expressed in some cases [6].

PBL is characterized by high recurrence and mortality rates and poor overall prognosis. At present, there is no standard treatment regimen for PBL. Surgery and radiotherapy are mainly used for early cases with local involvement. Anthracycline-containing regimens such as CHOP or CHOP-like regimens are commonly used [3,4,7]. However, the duration of remission obtained by chemotherapy is short, and ASCT shows greater feasibility, especially for patients with high risk factors. A retrospective analysis of 9 HIV-negative PBL patients from Moffitt Cancer Center in the United States was conducted. Four patients received ASCT as consolidation therapy after complete remission (first complete remission) as first-line therapy. The survival time was 13.3 months and 36.5 months, respectively [6]. Allogeneic hematopoietic stem cell transplantation for HIV-negative PBL has rarely been reported. The patient in our study received ASCT after achieving PR, however, brain metastasis occurred and the survival time was 11 months.

Conclusion

HIV-negative PBL is an aggressive disease and most of the patients are in late stage. ASCT may be a better treatment option for HIV-negative PBL patients. However, since PBL is rare, it is difficult to achieve single-center research, and prospective and multi-center collaboration is still needed.

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