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Case Presentation

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Primary T Cell Central Nervous System Lymphoma: Case Report and Literature Review

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Abstract

Primary central nervous system lymphoma accounts for 5% of primary intracranial tumors, most of which are derived from B lymphocytes. Diffuse large B cell lymphoma makes up the majority of primary central nervous system lymphoma. It accounts for more than 90% of all primary CNS lymphomas. Less than 5% of patients suffer from primary central nervous system T-cell lymphoma. Primary T cell central nervous system lymphoma is rare and should be took into account the possibility of getting it for patients whose clinical symptoms progress rapidly and imaging findings show single or multiple intracranial lesions. Here we report a case of multiple intracranial space occupying lesions, which was removed by microsurgical resection. Its pathologic diagnosis was T cell PCNSL. Because of its infrequency and variable display possibility, T cell PCNSL should be considered and included in the differential diagnosis at the time when we encounter multiple intracranial space occupying.

Keywords: Primary T cell central nervous system lymphoma; Lymphoma; Multiple intracranial space occupying; Tumor of the nervous system

Introduction

Lymphomas of the Central Nervous System (CNS) include primary and secondary. Primary central nervous system lymphoma accounts for 0.2% to 2% of the total [1]. Primary central nervous system lymphoma accounts for 5% of

primary intracranial tumors, most of which are derived from B lymphocytes. Diffuse Large B Cell Lymphoma (DLBCL) makes up the majority of primary central nervous system lymphoma. It accounts for more than 90% of all primary CNS lymphomas [2]. Less than 5% of patients have primary central nervous system T-cell lymphoma (TPCNSL) [3]. Primary T cell central nervous system lymphomas are rarely seen in immunocompetent patients, and there are few reports in the literature [4,5]. Here we report a case of multiple intracranial space occupying lesions, which was removed by microsurgical resection. The pathologic diagnosis was T cell PCNSL. Because of its infrequency and variable display possibility, T cell PCNSL should be considered and included in the differential diagnosis at the time when we encounter multiple intracranial space occupying.

Case Presentation

A 43-year-old woman came to our hospital with speech problems and impaired vision, and accompanied by paroxysmal nausea and vomiting. She had no recent history of infection or fever. After admission, we gave the patient a neurological examination. The neurological examination revealed motor aphasia, urinary incontinence and weakness of the right limbs and trunk. Peripheral Lymphadenopathy and Hepatosplenomegaly were not detected. Magnetic Resonance Imaging (MRI) of the brain revealed that intracranial lesions were located in the left frontotemporal parioccipital insula, right basal ganglia region and right frontal occipital lobe. The tumor signal showed multiple patchy, T1-weighted and T2-weighted signal shadows were slightly longer, and the tumor boundary was not clear. Fluid-Attenuated Inversion Recovery Imaging (FLAIR) showed slight high-intensity abnormal signal, and Diffusion-Weighted Imaging (DWI) showed restricted diffusion of patchy cortical areas. Enhanced magnetic resonance imaging showed gyri like enhancement, deepening of the adjacent sulci in some lesions, scattered edema signals in the periphery, compression and deformation of the left ventricle, and shift of the midline structure to the right **(Figure 1)**. Magnetic resonance spectra showed that NAA peak decreased and Cho peak increased. The CHO /NAA ratio was approximately 2.97 **(Figure 2)**. The differential diagnosis included lymphoma, encephalitis, inflammatory, gliomatosis cerebri, vasculitis and demyelinating disease.

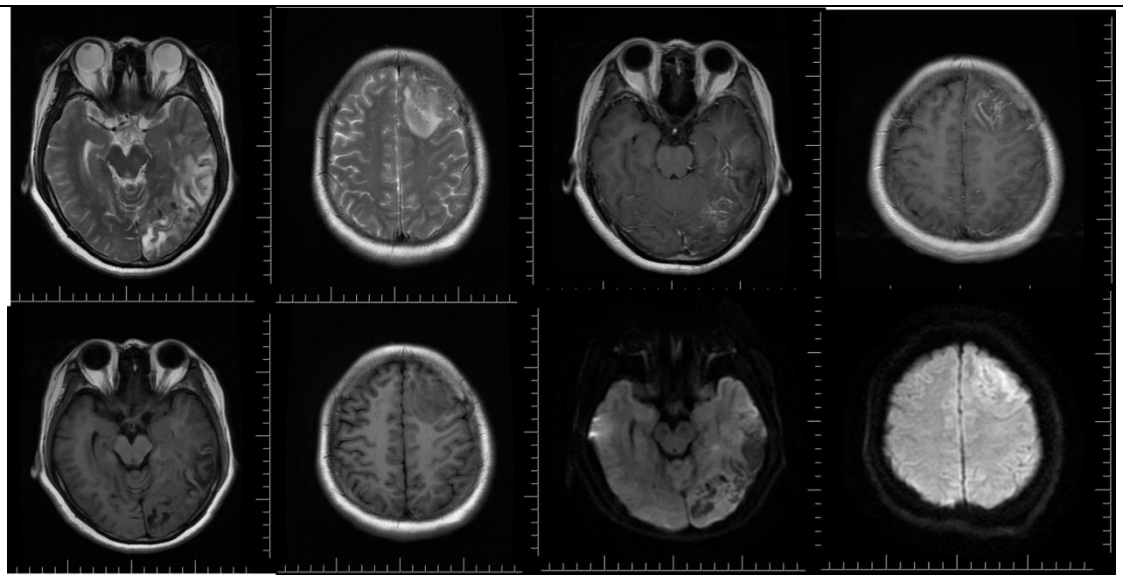

Figure 1: Magnetic resonance imaging (MRI) shows multiple and patchy lesion. T1-weighted and T2-weighted signal shadows are slightly longer. Restricted diffusion of patchy cortical areas can be found in diffusion-weighted imaging (DWI). Enhanced magnetic resonance imaging showed gyri like enhancement.

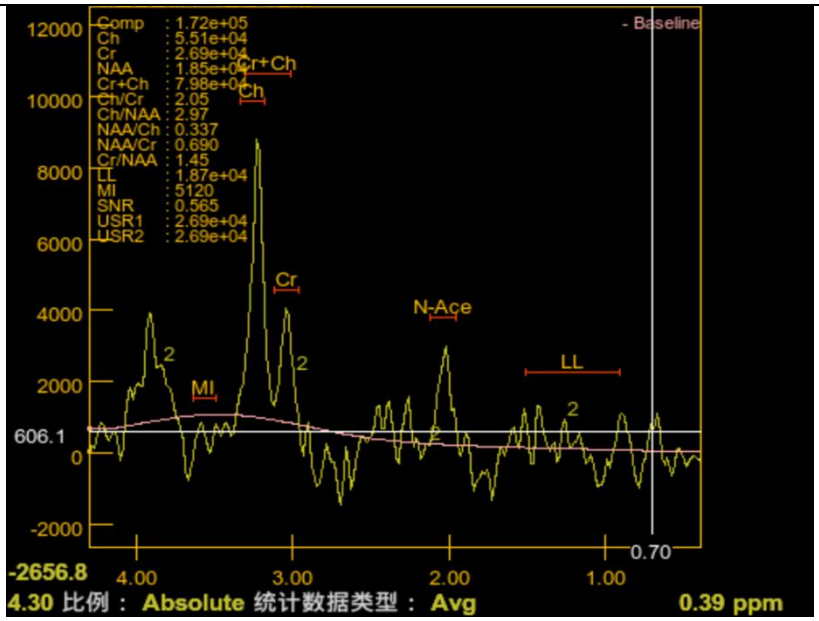

Figure 2: Magnetic resonance spectra showed that NAA peak decreased and Cho peak increased. The CHO /NAA ratio is approximately 2.97.

Microsurgical resection for the lesion was performed, and histopathologic examination revealed an infiltrate of lymphoid cells with angioinflammatory changes and scattered liquefied necrotic lesions. Immunohistochemically, we found these histiocytes were positive for CD2(+), AE1/AE3(-), LCA(+), CD20(-), CD79α(-), CD3(+), CD43(+), CD5(+), CD4(+), CD8(-), TIA-1(+), Perforin(-), CD56(-), CD10(-), Bcl-6(+), MUM-1(+), Bcl-2(+), CD30(-), c-myc(5%+), MPO(-), GFAP(-), NF(-), S-100(-), Syn(-), p53(+), Ki-67 (High expression). Molecular pathology revealed T cell rearrangement **(Figure 3)**. A diagnosis of Primary T cell central nervous system lymphoma was made. One month after surgery, the postoperative recovery was good. The hematology department provided further chemotherapy for the intraoperative pathology of the patient.

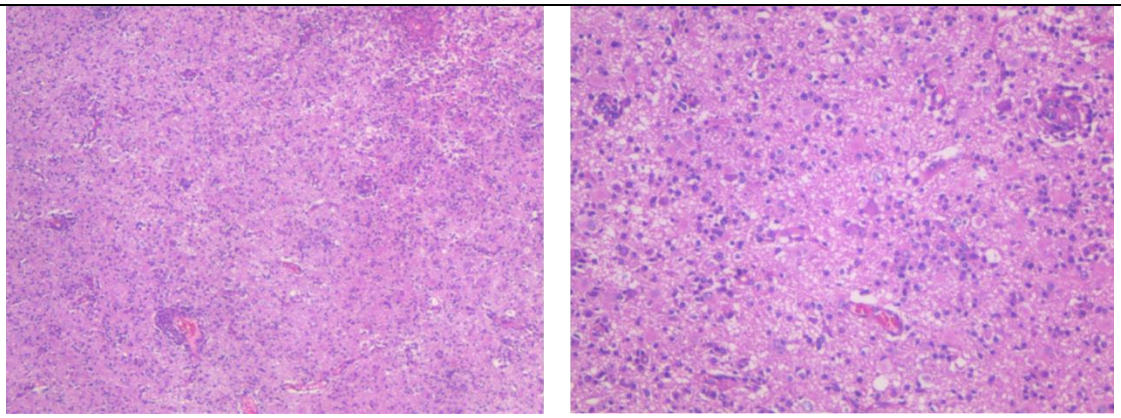

Figure 3: Histopathologic examination (100×) reveals an infiltrate of lymphoid cells with angioinflammatory changes and scattered liquefied necrotic lesions. Immunohistochemically, these histiocytes are positive for CD2(+), AE1/AE3(-), LCA(+), CD20(-), CD79α(-), CD3(+), CD43 (+), CD5(+), CD4(+), CD8(-), TIA-1(+), Perforin(-), CD56(-), CD10(-), Bcl-6(+), MUM-1(+), Bcl-2(+), CD30(-), c-myc(5%+), MPO(-), GFAP(-), NF(-), S-100(-), Syn(-), p53(+), Ki-67 (High expression) and molecular pathology revealed T cell rearrangement.

Discussion

Primary Central Nervous System Lymphoma (PCNSL) is similar to systemic lymphoma in cell morphology. PCNSL is a rare, non-Hodgkin's lymphoma of the central nervous system without systemic lymphoma manifestations. It accounts for 0.2% to 2% of the incidence of lymphoma **[1]**. Primary central nervous system lymphoma accounts for 5% of primary intracranial tumors. Most of these are derived from B lymphocytes. Diffuse Large B Cell Lymphoma (DLBCL) makes up the majority of primary central nervous system lymphoma. T-cell-derived primary central nervous system lymphoma accounts for only 7% of PCNSL. There is a Japanese case involving 121 patients with PCNSL. These patients were observed for 10 years and only 9 of them had TPCNSL, accounting for 7.4% of the total **[6]**. A 15-year follow-up observational study in Slovenia showed that there were only 2 TPCNSL cases (3.4%) out of 59 PCNSL patients at the center **[7]**. There are also some scattered case analysis reports in China, but most of them are B-cell-derived CNS lymphoma. A large multicenter study on TPCNSL has

shown that TPCNSL mostly involves structures above the tentorial area of the cerebellum, often with single or multiple lesions. The affected structures are usually the area around the lateral ventricle and can infiltrate into the corpus callosum and basal ganglia **[8,9]**.We roughly speculated that the multiple lymphoma lesions in this patient might be related to the fact that the lymphoma cells of the patient were undifferentiated T cells with high malignant degree. In this case, the patient was finally confirmed to be T-cell-derived central nervous system lymphoma by brain biopsy, and no involvement of peripheral lymph nodes or lymphatic organs was observed. It is a CD2 positive T-cell primary central nervous system lymphoma. The diagnosis of PCNSL needs to be differentiated from other diseases with multifocal lesions of the central nervous system, such as demyelinating disease of the central nervous system (multiple sclerosis, acute disseminated encephalomyelitis, etc.), AIDS-related lesions of the central nervous system, intracranial tuberculomas, metastatic intracranial tumors, meningoencephalitis, etc. The patient in this paper was initially diagnosed with encephalitis in another hospital, and the diagnosis of encephalitis could not be ruled out at the beginning of admission due to the patient's imaging findings.

Conclusion

Primary T cell central nervous system lymphoma is rare and should be took into account the possibility of getting it for patients whose clinical symptoms progress rapidly and imaging findings show single or multiple intracranial lesions. It is necessary for giving diagnosis to have a pathological biopsy of the position of the lesion in the brain, along with a detailed medical history and physical examination, as well as an appropriate imaging examination.

Acknowledgments

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