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Primary Central Nervous System T-cell Lymphoma with Unusual Clinical and Radiologic Findings

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Primary central nervous system lymphomas (PCNSLs), especially T-cell lymphoma, are quite rare and their clinical manifestations and radiologic findings have not been well known yet. We report herein on a 61-year-old man who presented with psychiatric symptoms and rapidly progressive cognitive deterioration without any definite focal neurological deficits. Brain magnetic resonance imaging (MRI) showed diffuse cortico-subcortical involvement with subtle gadolinium enhancement. Primary CNS T-cell lymphoma was confirmed on the histopathologic examination of the brain biopsy specimen.

Key Words: *Primary central nervous system lymphoma, T cell*

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INTRODUCTION

Most of the primary central nervous system lymphomas (PCNSLs) originated from B-cells and their imaging characteristics have been well documented[1-3]. On the other hand, the incidence of the T-cell type PCNSL (T-PCNSL) is less than 5% of all the PCNSLs in Western counturies[1, 4], and the incidence is 8.5-14.3% in the Eastern countries[5, 6]. Thus, their clinical manifestations and radiologic characteristics has been less well documented. In this paper, we describe one patient with T-PCNSL who presented with psychiatric symptoms and unusual radiologic findings.

CASE REPORT

A 61-year-old man was admitted with prominent behavioral psychiatric changes, including wandering, irritability, anxiety, insomnia, confusion, slurred speech and difficulty in walking and balance. Those behavioral changes ensued rapidly over 40 days, and eventually he became unable to follow verbal commands. The neurologic examination revealed confusion, cognitive impairment, dysarthria and dysphagia, but definite hemparesis or myoclonus was not noted. He had a poor oral food intake for about two months and he had lost over ten kg of weight. On admission,, he showed the restlessness, irritability, aggressiveness and the tendency to go out. The orientation for person and place was preserved. The spontaneous speech was relatively spared, but the content of language was meaningless and not understandable. The K-MMSE was not performed due to the poor cooperation and comprehension. Confusion with visual hallucination and purposeless motions was observed at night. His restlessness and irritability have been calmed down over time, finally he has become unresponsive, abulic and disoriented for place and person about 3 months later. The brain T2-wighted, FLAIR and diffusion-weighted magnetic resonance imaging (MRI) demonstrated diffuse hyperintensities in the left cerebellum, the bilateral temporal, frontal and occipital cortexes, and the subcortical structures such as the bilateral dorsomedial thalamus and caudate nucleus (Fig. 1A). Subtle enhancement was shown in these lesions after contrast injec-

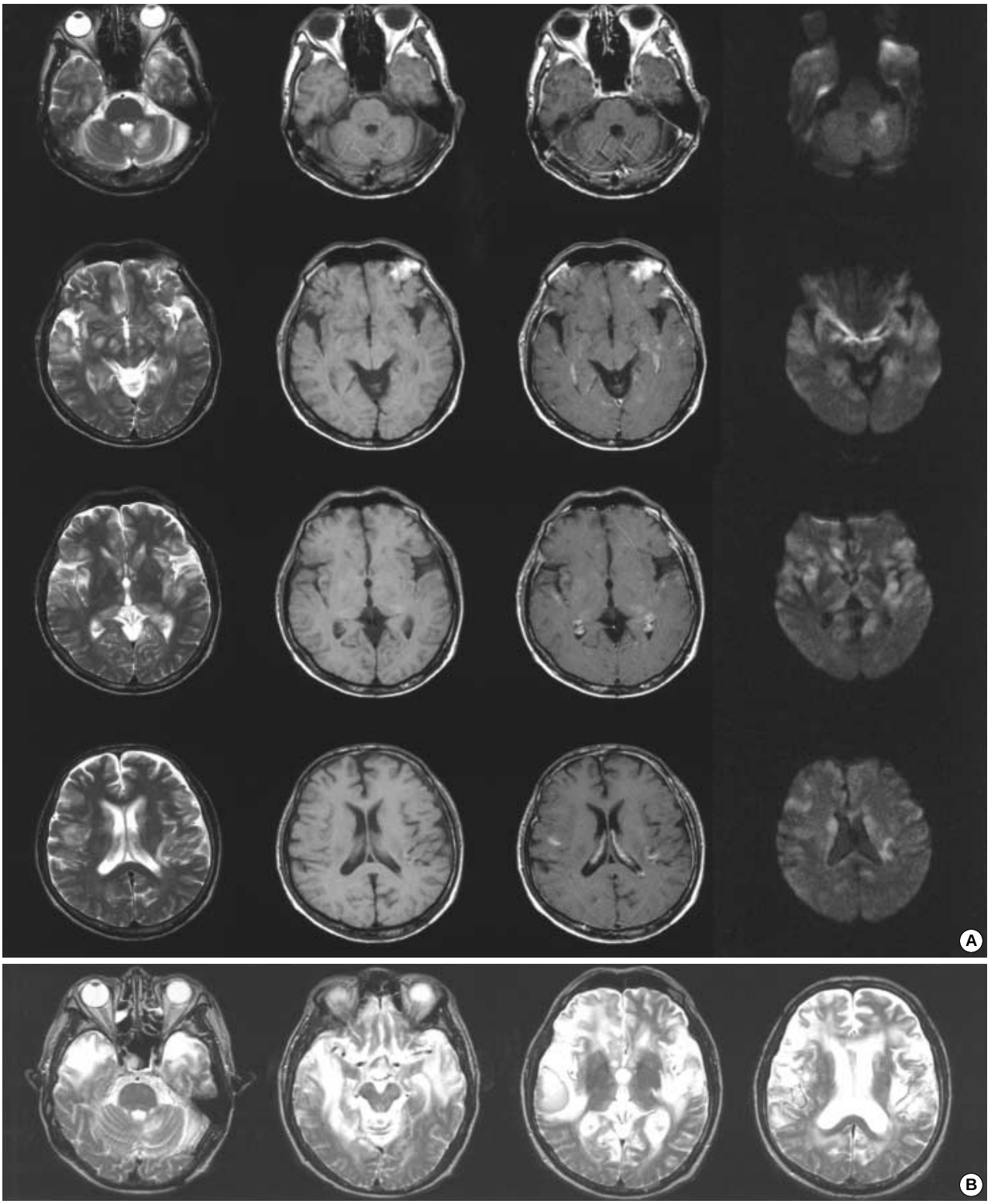

Fig. 1. T2, T1 weighted axial, T1 enhanced axial and diffusion weighted MR images. (A) High signal intensities on the T2 weighted and diffusion images, and hypo- or iso-signal intensities on T1 weighted images are shown in both temporal lobes, the frontal lobes, the occipital lobes, the caudate nucleus, thalamus and deep dentate nucleus of the cerebellum, and the lesions are mostly in the gray matter. Subtle patchy enhancement was shown after contrast enhancement. (B) On the follow-up MRI after 3 months, the previous lesions became more aggravated and involved the subcortical white matter as well, with stronger enhancement.

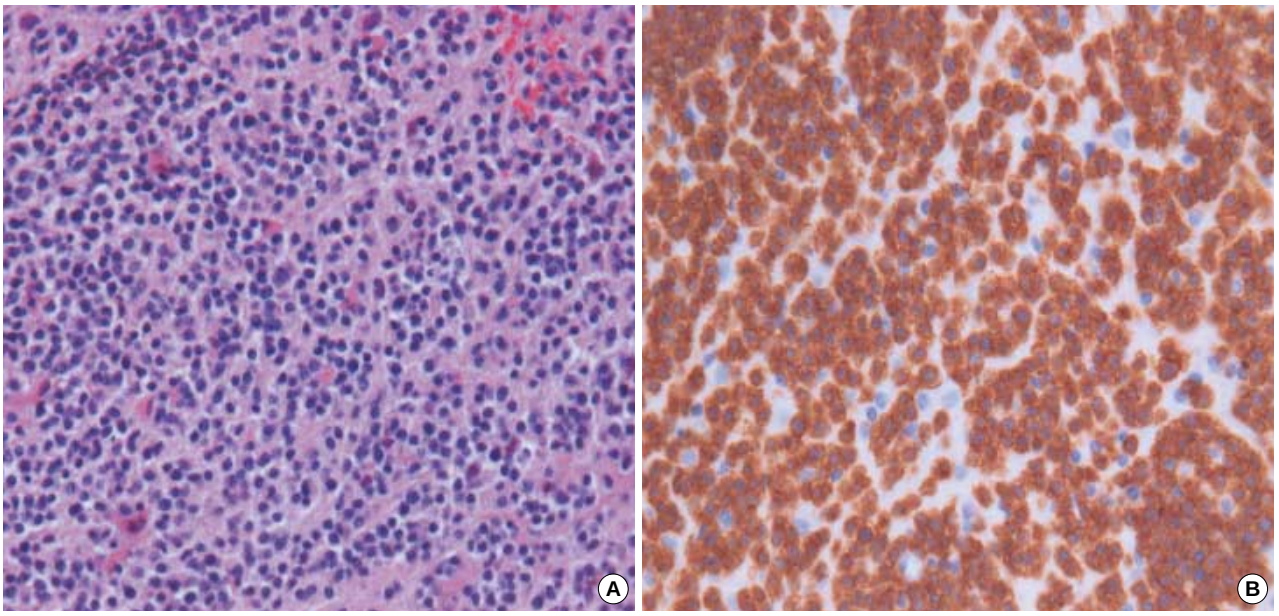

Fig. 2. The pathologic findings. (A) On H-E staining (×100), small to medium sized tumor cells with a high nuclear cytoplasmic ratio are noted. Some tumor cells have very condensed nuclear chromatin and no evident nucleoli, and other tumor cells have finely dispersed chromatin and small nucleoli. (B) On immunohistochemical staining (×100), the tumor cells are diffusely and strongly positive for cytoplasmic CD3, supporting that the tumor is of a T-cell lineage.

tion. On the EEG, generalized slow waves were frequently observed in both hemispheres without the typical periodic sharp waves and slow waves. Except for the positive 14-3-3 protein assay result, the CSF findings were normal on the routine laboratory examination and the cytology examination was negative for malignancy. The other immunologic markers for vasculitis or the connective tissue diseases such as FANA, anti-dsDNA, anti-nRNP, anti SS-A/SS-B, ANCA, cardiolipin antibody, rheumatoid factor and tumor markers were negative. For detecting hidden malignancy, the chest and abdominal CTs were performed and there were no abnormal findings on CT scans.

Considering the rapidly progressive neuropsychiatric symptoms and the multiple diffuse gray matter lesions on brain MRI, we suspected the various possibilities of metastatic cancer, glioblastoma multiforme, PCNSL, encephalitis, vasculitis and Creutzfedt-Jakob disease. Three months later, the follow-up MRI (Fig. 1B) showed more extensive lesions with strong contrast enhancement. A stereotactic brain biopsy in the left parietal cortex was performed for the confirmatory diagnosis. The neuropathologic findings showed tumor cells with a high nuclear/cytoplasmic ratio and diffuse, strong positivity for cytoplasmic CD3, which is compatable with T-PCNSL (Fig. 2). The patient was transferred to the oncology department and he underwent chemotherapy, but his conditions deteriorated and then he expired 6 months later.

DISCUSSION

In general, the common presenting signs and symptoms of PCNSL are a change in mental status followed by nausea, headache, hemiparesis, cerebellar signs, cranial nerve palsies and visual disturbances[2, 3]. Cerebrospinal fluid analysis yields a cytologic diagnosis in fewer than half of patients with PCNSL.

On MRI, B-PCNSL lesions are seen as clearly delineated solitary or multiple masses that appear iso- or hypo-intense on the T1-weighted images and they are mostly hypo-intense on the T2-weighted images[2, 3]. Nearly all the lesions show homogenous enhancement with gadolinium in the immunocompetent patients and they show rim enhancement in the immunocompromised patients[7, 8]. The lesions are commonly located in the supratentorial periventricular areas in the white matter of the frontal or parietal lobes and also on the subependymal regions[7].

In contrast to B-PCNSL, recent studies have revealed that the patients with T-PCNSL are younger and there is a male predominance, but their clinical presentations are similar to B-PCNSL. On brain MRI, these patients generally show one

or more masses with homogenous enhancement and they are frequently located in the infratentorial regions[9]. Unlike B-PCNSL, involvement of the cerebrospinal fluid is very rare in T-PCNSL[10, 11]. Interestingly, one study on Koreans[12] showed that T-PCNSLs are predominantly located in the subcortical lobar hemispheres and they appeared as a solitary mass with mild edema. On MRI, the T-PCNSL lesions have similar signal intensities to those of the B-PCNSLs on the T1- and T2-weighted images, but almost of the cases show rim enhancement after contrast enhancement[11, 12].

In our case, unique features were found on both the clinical and neuroimaging findings. The clinical manifestations are mainly psychiatric and behavioral changes, but not hemiparesis or seizure. Also, the brain MRI showed unusual findings such as multiple diffuse lesions that initially involve both the cortical gray matter and the subcortical midline structures simultaneously. Moreover, the lesions were hyperintense on the T2-weighted images and the DWIs, and the lesions are iso- or hypo-intense on the T1-weighted images with subtle enhancement after gadolinium injection. In other words, there is a remarkable contrast to the previously reported findings of T-PCNSL, in that multiple supra- and infratentorial lesions and initial cortical involvement with only subtle enhancement. When a patient shows rapidly progressing neurocognitive dysfunction and diffuse cortical involvement on MRI, we should consider the possibility of PCNSL as a differential diagnosis.

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