

Magnetic Resonance Imaging Characteristics of Primary Central Nervous System T-cell Lymphoma

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Primary central nervous system lymphomas (PCNSLs) are rare non-Hodgkin tumors defined as lymphomas of the central nervous system (CNS) without primary tumor elsewhere. It was reported that PCNSLs represented only 3–7% of primary brain tumors and 1–5% of all lymphomas.^[1,2] Most of PCNSLs are B-cell lymphomas, while T-cell PCNSL (T-PCNSL) is extremely rare, the majority of the reported T-PCNSL cases are clinically sporadic that focus on the treatment, and its imaging features have rarely been described.^[1,3,4] Herein, we report a case of T-PCNSL and comprehensively summarize its magnetic resonance imaging (MRI) characteristics. It is the first time in China to focus on the MRI diagnosis of T-PCNSL.

A 29-year-old female, complaining of headache and progressive numbness and weakness in her right extremities for 20 days, was referred to our hospital. Neurological examination revealed reduced muscle strength in her right limbs. Laboratory tests were reported within normal limits. Past medical history was negative for considerable pathologies. HIV and Epstein–Barr virus (EBV) status of the patient were also negative.

Brain MRI was performed at 3.0T (GE Discovery MR750), which revealed an irregular mass of approximately 4 cm × 5 cm in the left frontal subcortical region. The mass was accompanied with prominent perilesional edema, causing homolateral ventricle narrowing without obvious midline shift. The mass showed slightly hypointensity on T1-weighted images (T1WIs) [Figure 1A,a] and heterogeneously hyperintensity on T2-weighted images (T2WIs). In addition, T2WI displayed an extremely

high signal core and a slightly hyperintense periphery, linear and reticular hypointensity was found within the central area on T2WI [Figure 1A, b–d]. Gadolinium-DTPA-enhanced T1WI revealed prominent rim enhancement on the periphery, with a gap facing the cortex, resulting in an “open-ring” pattern [Figure 1B]. Diffusion-weighted imaging (DWI) showed remarkable high signal on the periphery of the mass which also presented a notch similar to the enhancing pattern; and apparent diffusion coefficient (ADC) map confirmed the presence of restricted diffusion, with a low average ADC value (range, 0.81–0.85) [Figure 1C, a and b]. To evaluate the metabolic information of the tumor, single-voxel ¹H-MR spectroscopy (MRS) was performed subsequently. The result revealed an evident lactate and lipid peaks, increased choline/creatine (Cho/Cr) ratio (range, 2.7–3.5) and reduced N-acetylaspartate/creatine (NAA/Cr) ratio (range, 1.3–1.5) in the enhanced peripheral region of the lesion [Figure 1C, c and d].

Because of the progressively aggravated limb asthenia, surgical resection and pathological study were performed subsequently after MR examination. Microscopically with hematoxylin and eosin staining, atypical small lymphocytes

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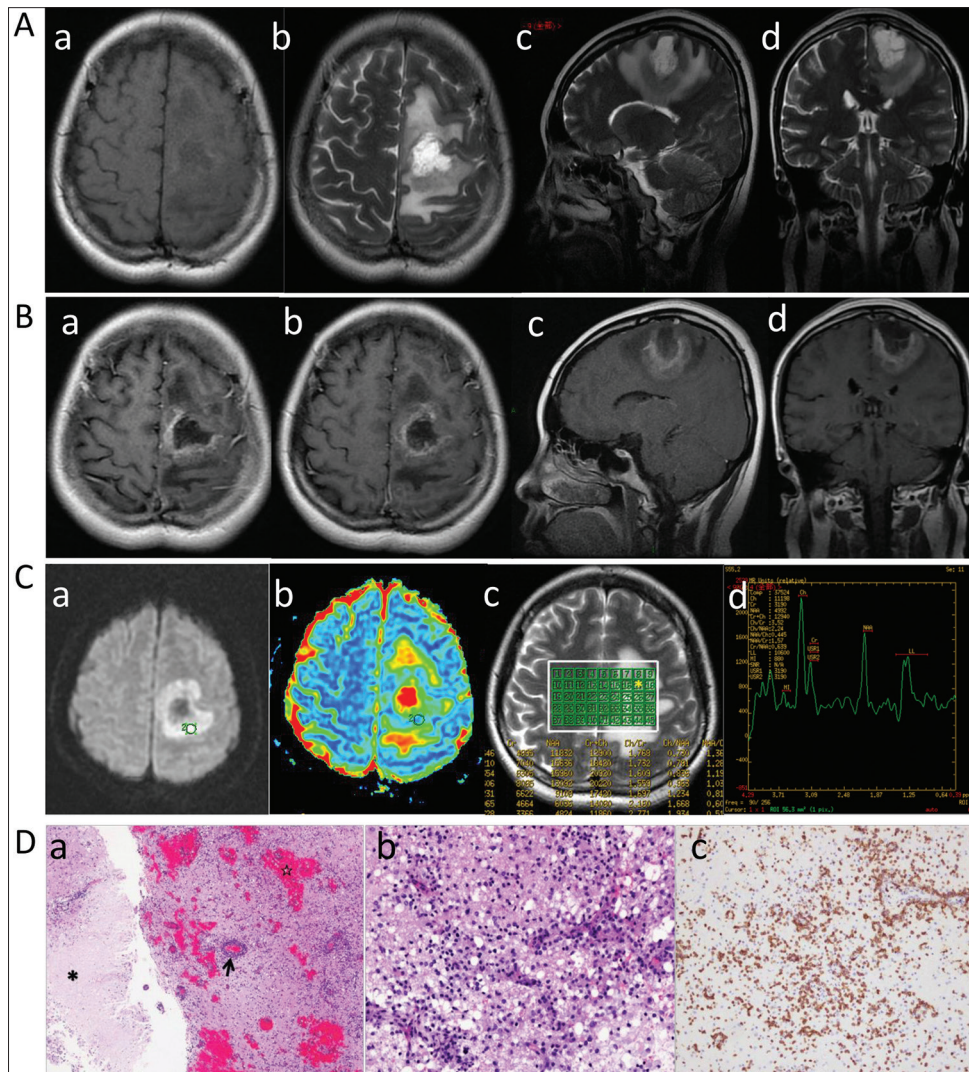


Figure 1: (A) T1-weighted image (a) and T2-weighted image (b-d); (B) axial (a-b), sagittal (c) and coronal (d) gadolinium-DTPA-enhanced T1-weighted images showing prominent “open-ring” enhancement; (C) diffusion-weighted imaging (a) and apparent diffusion coefficient (b) map showing restricted diffusion with a notch sign; ¹H-MRS (c and d), where * indicated the region of interest; (D) pathological findings (a) (H and E, original magnification $\times 40$) showing perivascular cuff (arrow), broad areas of necrosis (*), and microbleeds (\star); (b) (H and E, original magnification $\times 40$) showing small atypical lymphocytes with astrocytic response; (c) immunohistochemical image showing strongly positive staining for CD3 (original magnification $\times 200$).

infiltrated the brain tissue and formed perivascular cuffs [Figure 1D]. Immunohistochemically, these atypical lymphocytes express LCA, CD2, CD3, and CD5, but lack CD20, PAX-5, CD10, and CD56. *In situ* hybridization did not find any EBV-encoded RNA in the tumor cell nuclei. The molecular genetic study further showed T-cell receptor gene rearrangement with a clonal appearance.

The following systematic examination ruled out involvement of the tumor elsewhere. The final clinical diagnosis was primary peripheral T-cell lymphoma of CNS.

T-PCNSL is rare and highly malignant tumor type of CNS, associated with poor prognosis.^[1] Immunodeficiency is a risk factor for T-PCNSL, and the predilection age ranges from 55 to 57 years.^[3] Other than surgical resection for other CNS malignant tumors such as glioma and solitary metastasis, a combination of chemotherapy and radiotherapy is the most preferred treatment for T-PCNSL. In addition, the therapeutic

regimen of T-PCNSL is also quite different from that of B-PCNSLs. Therefore, it is of great importance to make a definite diagnosis of T-PCNSL in the first visit, to avoid unnecessary operation and start appropriate treatment as soon as possible. However, even histopathological biopsy, which was considered the golden criteria of diagnosis, may miss the tumor tissue because of the limited size of the specimen. Instead, MRI can provide more details about the tumor, including a comprehensive evaluation of the invading extent.

So far, the imaging manifestation of T-PCNSL was just sporadic reported abroad, and there has not been any report in China. Due to the inadequate understanding of T-PCNSL, the diagnostic accuracy remains unsatisfied. Kim and Kim^[4] suggested that T-PCNSL may have a predilection of subcortical location, with occasionally observed leptomeningeal involvement, while B-PCNSL usually presented as multiple or solitary intra-axial mass contacting a

CSF surface. T-PCNSL shows mildly hypointensity on T1WI and slightly hyperintensity on T2WI with mild to moderate edema. DWI consistently manifests hyperintensity with an extremely low ADC, representing the high cellularity.^[1] After administration of contrast agent, most lesions show moderate to marked contrast enhancement, with fist-like, flame-shaped, or butterfly-shaped patterns. In addition, necrosis and hemorrhage were more common in T-PCNSL than in B-PCNSLs, especially in immunocompetent patients, resulting in heterogeneous or rim-like enhancement.^[4,5] ¹H-MRS is the only noninvasive strategy to explore the biochemical information of brain lesions and has been investigated for the evaluation of PCNSLs but lack of the detailed characteristic for T-PCNSL.^[1]

The tumor in our case had a subcortical location, with both of the DWI and contrast enhancement demonstrating an “open-ring” pattern. Compared pathological study suggested that this imaging appearance was caused by the infiltration of tumor cells along the vessels and avascular necrosis, which may be a typical feature of T-PCNSL. In addition, the predominance of lipid peaks combined with high Cho/Cr ratios and reduced NAA was demonstrated in the ¹H-MRS study of our case, which was in agreement with the previous report.^[1] Thus, we considered that ¹H-MRS can also contribute to the diagnosis.

The main differential diagnoses of T-PCNSL mainly include other pathological subtypes of PCNSLs and systemic lymphomas involving the CNS and high-grade gliomas. In our opinion, if the tumor has a subcortical location, and any MR evidence of necrosis or hemorrhage, particularly in an immunocompetent patient, T-cell origin should be considered. Meanwhile, a systemic investigation is called for ruling out elsewhere involvement before making the “primary” diagnosis. Compared with heterogeneously garland-like-enhanced

high-grade gliomas, T-PCNSL tends to demonstrate an open-ring enhancement.

Pathologically, prominent tumor cell infiltration in a perivascular cuffing pattern is typical for both B- and T-PCNSL, and CD4, CD3, and CD5 are the major immunohistochemistry markers of T-PCNSL.^[3] The expression pattern of our case all indicated a T-cell lineage, and the genetic study further confirmed the diagnosis.

In conclusion, T-PCNSL has some distinct MRI features, including a predilection of subcortical location and a tendency toward necrosis and hemorrhage. We speculate that these signs may increase the accuracy of MRI diagnosis.

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Conflicts of interest

There are no conflicts of interest.

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