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ABOUT COVER

Editorial Board Member of *World Journal of Radiology*, Alberto Tagliafico, MD, Assistant Professor, Department of Health Sciences (DISSAL), University of Genova, Genova 16138, Italy. alberto.tagliafico@unige.it

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Distinctive magnetic resonance imaging features in primary central nervous system lymphoma: A case report

Li-Hong Liu, Han-Wen Zhang, Hong-Bo Zhang, Xiao-Lei Liu, Hua-Zhen Deng, Fan Lin, Biao Huang

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Li-Hong Liu, Han-Wen Zhang, Xiao-Lei Liu, Hua-Zhen Deng, Fan Lin, Department of Radiology, The First Affiliated Hospital of Shenzhen University, Health Science Center, Shenzhen Second People's Hospital, Shenzhen 518036, Guangdong Province, China

Han-Wen Zhang, Hong-Bo Zhang, Biao Huang, The Second School of Clinical Medicine, Southern Medical University, Guangzhou 510282, Guangdong Province, China

Hong-Bo Zhang, Department of Radiology, Sun Yat-Sen University, Shenzhen 518000, Guangdong Province, China

Biao Huang, Department of Radiology, Guangdong Provincial People's Hospital, Guangdong Academy of Medical Sciences, Guangzhou 510000, Guangdong Province, China

Corresponding author: Han-Wen Zhang, MD, Doctor, Department of Radiology, The First Affiliated Hospital of Shenzhen University, Health Science Center, Shenzhen Second People's Hospital, No. 3002 Sungangxi Road, Shenzhen 518036, Guangdong Province, China.
zhwstarcraft@outlook.com

Abstract

BACKGROUND

Primary central nervous system lymphoma (PCNSL) is a rare malignant tumor originating from the lymphatic hematopoietic system. It exhibits unique imaging manifestations due to its biological characteristics.

CASE SUMMARY

Magnetic resonance imaging (MRI) with diffusion-weighted imaging (DWI), perfusion-weighted imaging (PWI), and magnetic resonance spectroscopy was performed. The imaging findings showed multiple space-occupying lesions with low signal on T1-weighted imaging, uniform high signal on T2-weighted imaging, and obvious enhancement on contrast-enhanced scans. DWI revealed diffusion restriction, PWI demonstrated hypoperfusion, and spectroscopy showed elevated choline peak and decreased N-acetylaspartic acid. The patient's condition significantly improved after hormone shock therapy.

CONCLUSION

This case highlights the distinctive imaging features of PCNSL and their importance in accurate diagnosis and management.

Key Words: Primary central nervous system lymphoma; Primary central nervous system

lymphoma; Diffusion-weighted imaging; Perfusion-weighted imaging; Magnetic resonance imaging; Case report

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Core Tip: Primary central nervous system lymphoma (PCNSL) is a rare tumor of the central nervous system with distinctive imaging features. This case report highlights the imaging manifestations of multiple PCNSL lesions using diffusion-weighted imaging, perfusion-weighted imaging, and magnetic resonance imaging. Accurate diagnosis is crucial for appropriate management. Further research and larger studies are needed to enhance the understanding and diagnostic accuracy of PCNSL.

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INTRODUCTION

Primary central nervous system lymphoma (PCNSL) is an exceptionally rare and aggressive malignancy originating within the central nervous system. Despite its infrequency, PCNSL presents unique diagnostic and therapeutic challenges, rendering it a subject of profound clinical importance[1]. In recent years, advanced imaging techniques, including diffusion-weighted imaging (DWI), perfusion-weighted imaging (PWI), and magnetic resonance (MR) spectroscopy, have greatly enhanced our ability to comprehend the disease and its distinctive manifestations[2].

While previous studies have provided valuable insights into the broader understanding of PCNSL, there is a critical gap in characterizing the imaging features on an individual case level[3]. This study endeavors to bridge this gap by conducting an in-depth examination of DWI, PWI, and MR spectroscopy findings in a single PCNSL case. By scrutinizing these imaging modalities within the unique context of this individual case, we aim to contribute to the comprehensive understanding of PCNSL's imaging features and their potential clinical implications[4].

The primary research problem addressed in this study is to delineate the specific magnetic resonance imaging (MRI) imaging manifestations of PCNSL within the scope of a single case analysis and to comprehend their diagnostic and clinical significance. To accomplish this, we have conducted a detailed analysis of DWI, PWI, and MR spectroscopy findings in the context of this singular PCNSL case.

CASE PRESENTATION

Chief complaints

A 79-year-old female patient with a previously unremarkable medical history presented with a sudden onset of unexplained dizziness accompanied by projectile vomiting, characterized by the ejection of gastric contents. She also reported a sense of heaviness and weakness in her limbs.

History of present illness

Initial evaluation at another medical facility revealed the presence of multiple intracranial space-occupying lesions. These lesions were detected through a computed tomography scan, which indicated the involvement of the cerebellar hemisphere, corpus callosum's splenium, and the left parietal lobe.

History of past illness

No special notes.

Personal and family history

No special notes.

Physical examination

Physical examination revealed no abnormalities.

Laboratory examinations

The platelet specific volume was slightly elevated. The monocyte count was mildly elevated. The bacterial content in the urine test increased [4225.40, reference value: 0-4000 (/μL)].

Imaging examinations

Upon admission, the patient underwent a comprehensive evaluation, including MRI with DWI, PWI, and MR spectroscopy (MRS). The MRI results confirmed the presence of multiple space-occupying lesions, characterized by low signal intensity on T1-weighted imaging, uniform high signal intensity on T2-weighted imaging, and prominent enhancement on contrast-enhanced scans (Figure 1). DWI further revealed diffusion restriction, while PWI demonstrated hypoperfusion in all the identified lesions. Additionally, spectroscopy (MRS) depicted an elevated choline peak and decreased N-acetylaspartic acid. Notably, MRS also revealed the presence of a Lip peak within the lesion (Figure 2). These combined imaging features strongly suggested the possibility of PCNSL.

FINAL DIAGNOSIS

Pathological examination of the intracranial lesion confirmed the presence of an aggressive B-cell lymphoma. Immunohistochemical analysis demonstrated the following profile: CD21 (-), CD10 (-), CD20 (+), CD3 (background T-cells +), CD30 (-), PAX-5 (+), Bcl-6 (+), MUM-1 (+), Ki-67 (approximately 80%), CD5 (-), CD23 (-), Bcl-2 (-), CyclinD1 (-), CD79A (+), C-MYC (approximately 30%+), P53 (10%+ with varying intensity), GFAP (glial cells +). Importantly, Epstein-Barr virus-encoded small RNA (EBER) was not detected. These findings confirmed the diagnosis of diffuse large B-cell lymphoma of the non-germinal center B-cell (non-GCB) subtype, supporting the diagnosis of PCNSL.

TREATMENT

Treatment and response

In response to the initial presentation and MRI findings, the patient underwent a diagnostic trial of steroid therapy. This treatment led to a significant improvement in the patient's neurological condition, although it was accompanied by the emergence of certain neuropsychiatric symptoms. Subsequent administration of medications, including lorazepam and olanzapine, resulted in a notable improvement in the patient's neuropsychiatric symptoms. Follow-up MRI examinations indicated a reduction in enhancement in the lesions located in the hippocampus and left parietal lobe, with the other lesions remaining stable or showing slight reductions (Figure 3).

OUTCOME AND FOLLOW-UP

The patient subsequently received standardized lymphoma immunotherapy and chemotherapy regimens, and her prognosis remains favorable.

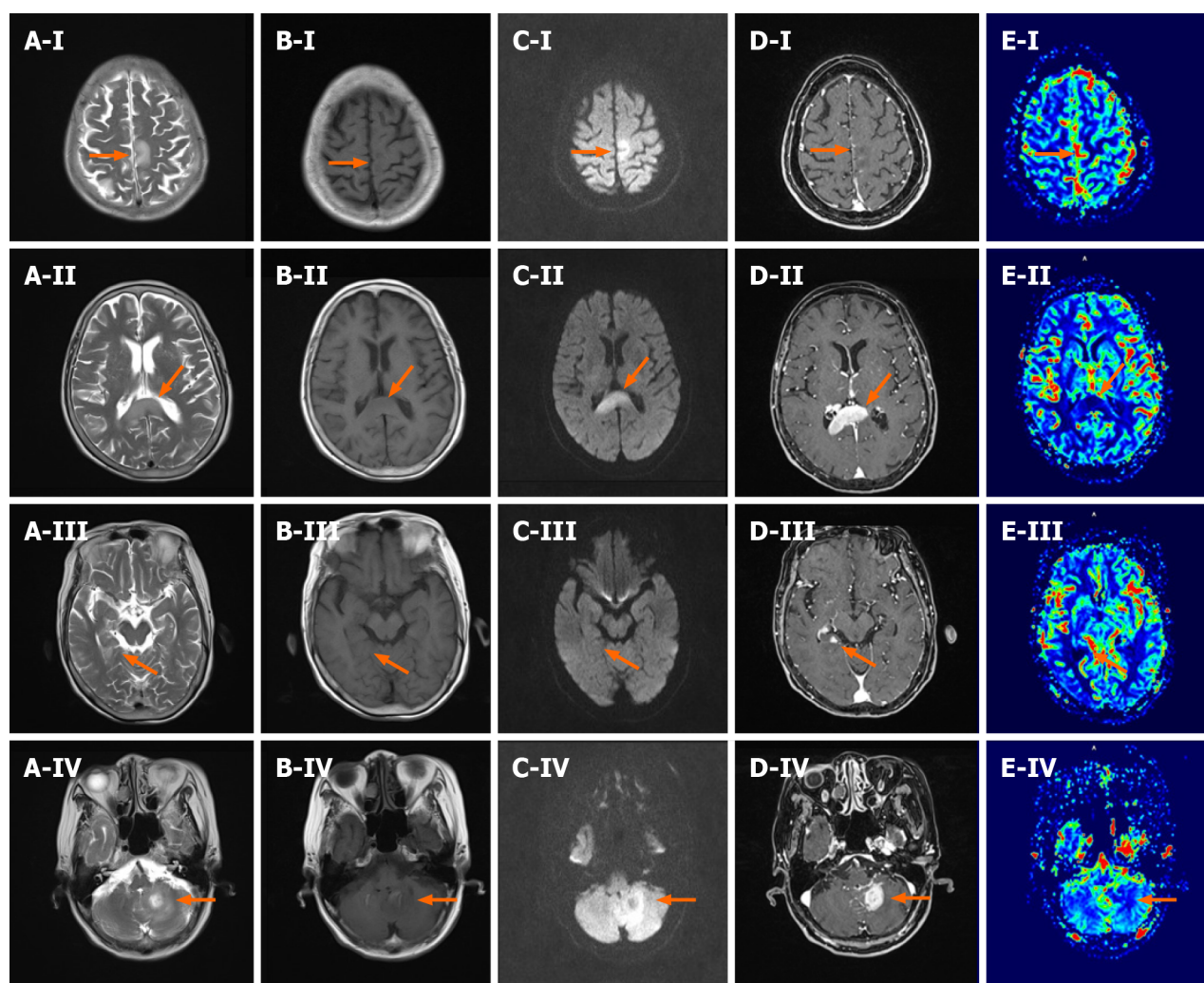
DISCUSSION

There is no lymphoid tissue present within the central nervous system, making brain lymphomas relatively rare. Currently, the origin of intracerebral lymphoma can be attributed to the following factors: Firstly, reactive lymphocytes enter the brain tissue during intracerebral infection and undergo malignant transformation through various mechanisms. Secondly, activated peripheral lymphocytes transform into tumor cells and migrate to the brain through the bloodstream, resulting in tumors primarily located around the ventricles, basal ganglia, and frontoparietal lobes. Thirdly, undifferentiated pluripotent stem cells surrounding blood vessels in the brain may serve as the source of intracerebral lymphoma. Histologically, intracerebral lymphomas exhibit predominant sleeve-shaped growth, infiltrating the surrounding brain parenchyma, and demonstrating multicentric growth within the tumor[5].

PCNSL typically presents as supratentorial lesions, with predilection sites in the cerebral hemisphere, corpus callosum, basal ganglia, and thalamus[6]. The imaging findings of PCNSL are closely related to its pathological features. In this case, the lesions were confined to the brain tissue, involving both supratentorial and infratentorial areas, which is relatively rare[7].

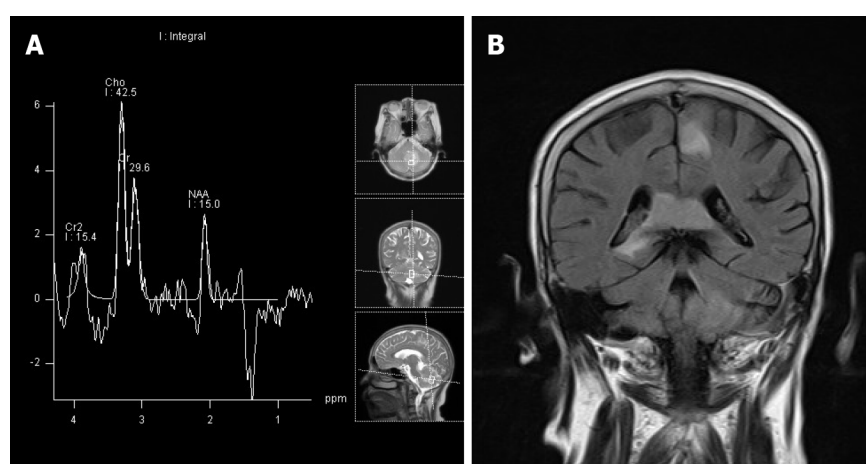
On conventional MRI, PCNSL shows iso-hypointensity on T1-weighted imaging and iso-hypointensity on T2-weighted imaging[8]. DWI demonstrates diffusion restriction due to densely arranged tumor cells. According to Lin *et al*[9], their study suggests that combining DWI ADC value with T1WI enhanced scan can aid in the differentiation of glioblastoma from PCNSL. PWI reveals hypoperfusion, reflecting the hypo-vascular nature of PCNSL[10]. Contrast-enhanced scans show uniform enhancement when the tumor invades adjacent brain parenchyma and disrupts the blood-brain barrier (BBB). In our previous study, we compared high-grade glioma (HGG) with lymphoma using dynamic contrast-enhanced (DCE) imaging, and found that lymphoma has more obvious damage to the BBB, resulting in transfer constant(K^{trans}) values even higher than HGG[11]. The characteristic imaging features, such as the "fist sign," "sharp horn sign," and "butterfly wing sign," (usually occurs in the corpus callosum) are associated with the tumor's angiophilic growth.

MRS plays a crucial role in the evaluation of PCNSL. Elevated choline peak, decreased N-acetylaspartic acid, and a towering lipid peak are commonly observed[12]. The towering lipid peak is highly specific for the diagnosis of PCNSL, attributed to the accelerated turnover of lymphocytes and macrophages. PCNSL should be differentiated from



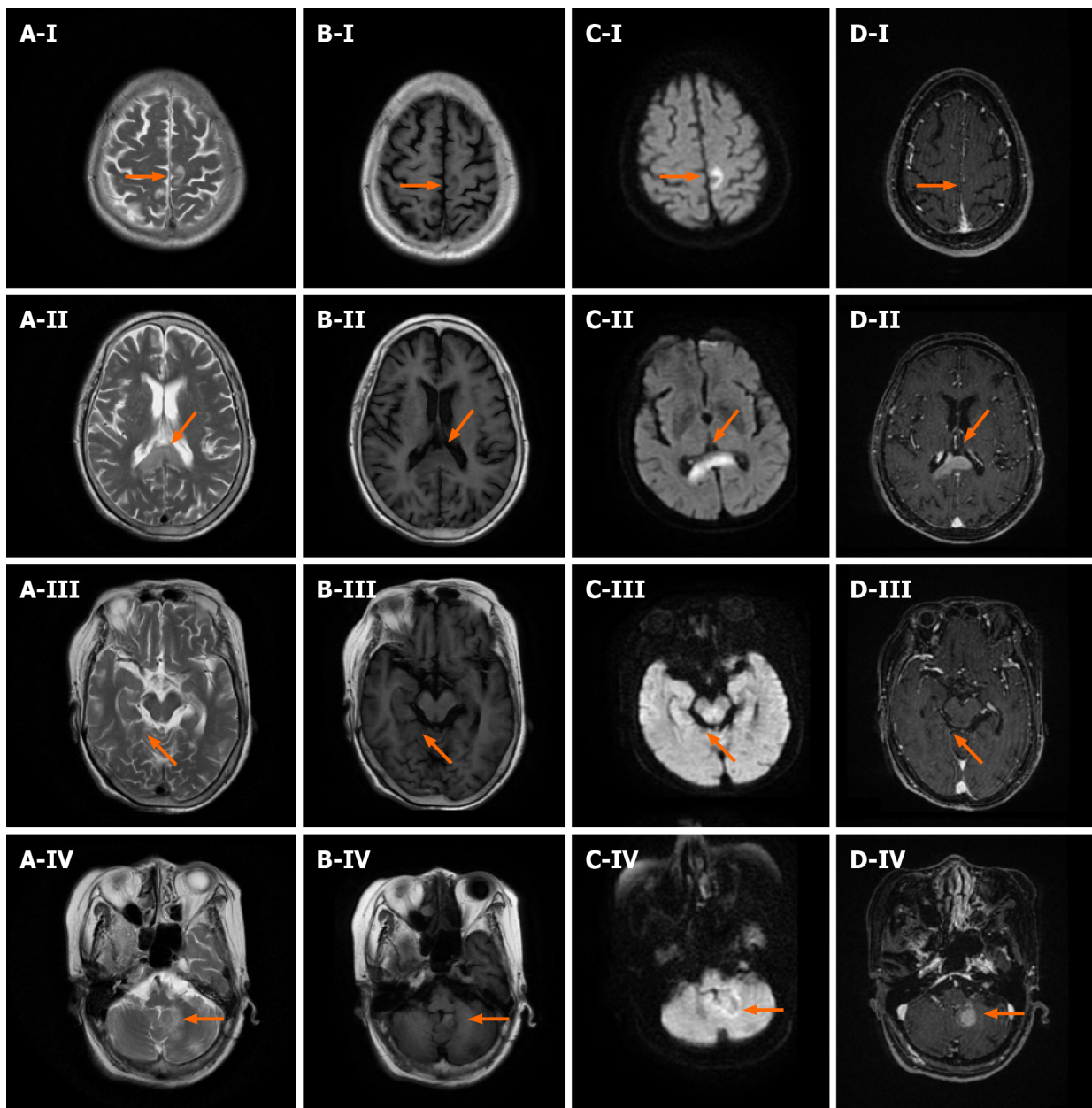
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Figure 1 Images of the patient prior to corticosteroid pulse therapy. A: T2WI; B: T1WI; C: Diffusion-weighted imaging; D: T1-weighted enhanced scan; E: Dynamic susceptibility contrast perfusion-weighted imaging cerebral blood volume; I: parietal lobe lesions; II: corpus callosum lesions; III: hippocampal lesions; IV: Left cerebellar hemisphere lesions.



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Figure 2 Notably, magnetic resonance spectroscopy also revealed the presence of a lip peak within the lesion. A: Magnetic resonance spectroscopy; B: Butterfly sign when primary central nervous system lymphoma is located in the corpus callosum.



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Figure 3 Images of the patient following corticosteroid pulse therapy. A: T2WI; B: T1WI; C: Diffusion-weighted imaging; D: T1-weighted enhanced scan; I: Parietal lobe lesions; II: Corpus callosum lesions; III: Hippocampal lesions; IV: Left cerebellar hemisphere lesions.

demyelinating diseases, as their imaging findings may resemble each other[13]. Hormone shock therapy can provide symptomatic relief in both PCNSL and demyelinating diseases, but recovery of symptoms after hormone therapy withdrawal is indicative of PCNSL.

In addition to our contributions to the understanding of PCNSL, it is essential to recognize the limitations of our study. The utilization of a single-case design inherently limits the generalizability of our findings to a broader population of PCNSL patients. Future research should strive to replicate these findings in larger cohorts to establish their broader applicability. Furthermore, our study primarily focuses on the imaging aspects of PCNSL, and future investigations could explore the correlation between imaging features and specific clinical outcomes. These considerations highlight both the strengths and the areas for improvement in our research.

CONCLUSION

This single-case analysis of PCNSL has shed light on the distinctive MRI imaging features of this rare malignancy. By employing advanced techniques such as DWI, PWI, and MR spectroscopy, we have provided a comprehensive characterization of PCNSL's imaging manifestations. However, it is important to acknowledge the limitations of this study. The use of a single-case design restricts the generalizability of our findings to a broader population. Future research should

aim to replicate these findings in larger cohorts of PCNSL patients to establish their broader applicability. Additionally, our study focused on the imaging aspects, and future investigations could explore the correlation between imaging features and specific clinical outcomes. Despite these limitations, our study contributes valuable insights into the unique imaging features of PCNSL and serves as a foundation for further research in this area.

FOOTNOTES

Author contributions: Case studies were provided by Liu LH; Zhang HW wrote the article; Deng HZ searched the literature; Zhang HB analyzed the case; Huang B and Lin F revised and reviewed the manuscript; Huang B (huangbiao@gdph.org.cn), Lin F (foxetfoxet@gmail.com), and Zhang HW contributed equally to this research.

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Country/Territory of origin: China

ORCID number: Han-Wen Zhang 0000-0001-5731-7429; Fan Lin 0000-0003-1595-2736.

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