International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2025: 7.089

A Rare Case of Primary CNS Lymphoma Presenting as a Stroke Mimic

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Abstract: ***Primary central nervous system lymphoma (PCNSL) is a rare and potentially aggressive malignancy that can present with variable clinical and radiological features, occasionally mimicking acute stroke. We report a 48-year-old deaf and mute hypertensive man who presented with sudden-onset right-sided upper and lower limb weakness and leftward deviation of the angle of the mouth, with progression over 24 hours. An initial diagnosis of stroke was made. Based on imaging and further testing a differential diagnosis of CNS lymphoma vs demyelination was considered. Biopsy confirmed primary CNS Lymphoma – DLBCL subtype. This case highlights the importance of considering PCNSL in atypical stroke presentations and the need for timely tissue diagnosis.***

Keywords: Primary CNS lymphoma, diffuse large B-cell lymphoma, stroke mimic, brain biopsy, sudden weakness

1. Introduction

Primary CNS lymphoma is a rare and elusive disorder with variable symptoms, signs, clinical courses and images findings that can mimic stroke syndromes. Early diagnosis, typically confirmed through imaging and biopsy, is crucial for effective treatment. Here we report a case of Primary CNS lymphoma presenting as a stroke mimic.

2. Case Report

48/M, deaf and mute, hypertensive presented with sudden onset weakness of right upper and lower limbs, deviation of angle of mouth to left that progressed over 24 hours. An initial diagnosis of stroke was made. CT showed a central hyperdense lesion with surrounding edema in left frontoparietal region and hypodensity in right periventricular region. A space occupying lesion was suspected and MRI was done. MRI showed ill-defined T2/FLAIR hyperintense, infiltrating white matter lesions in bilateral periventricular region with diffusion restriction and mild contrast enhancement. All routine blood and CSF work up including HIV, NMO/MOG antibodies, OCBs, CSF malignant cytology was negative. USG scrotum, neck, axilla and inguinal region were negative for any lymph node enlargement. DDx - Multiple sclerosis vs lymphomatous infiltration

Patient was given pulse IVMP for 5 days → limb power improved by 1 grade. Brain biopsy was planned but attenders were not willing. Discharged with oral steroid and advised review → lost to follow up.

4months later, he presented with right focal seizures and altered sensorium. On examination, there was spasticity in all limbs, flexion posturing of right upper limb, right sided weakness. Repeat MRI brain showed increase in previous lesions. Brain biopsy was done suggestive of Diffuse Large B Cell Lymphoma. Imaging was done to exclude other sites of pathology. Palliation was started keeping in view patient's general condition.

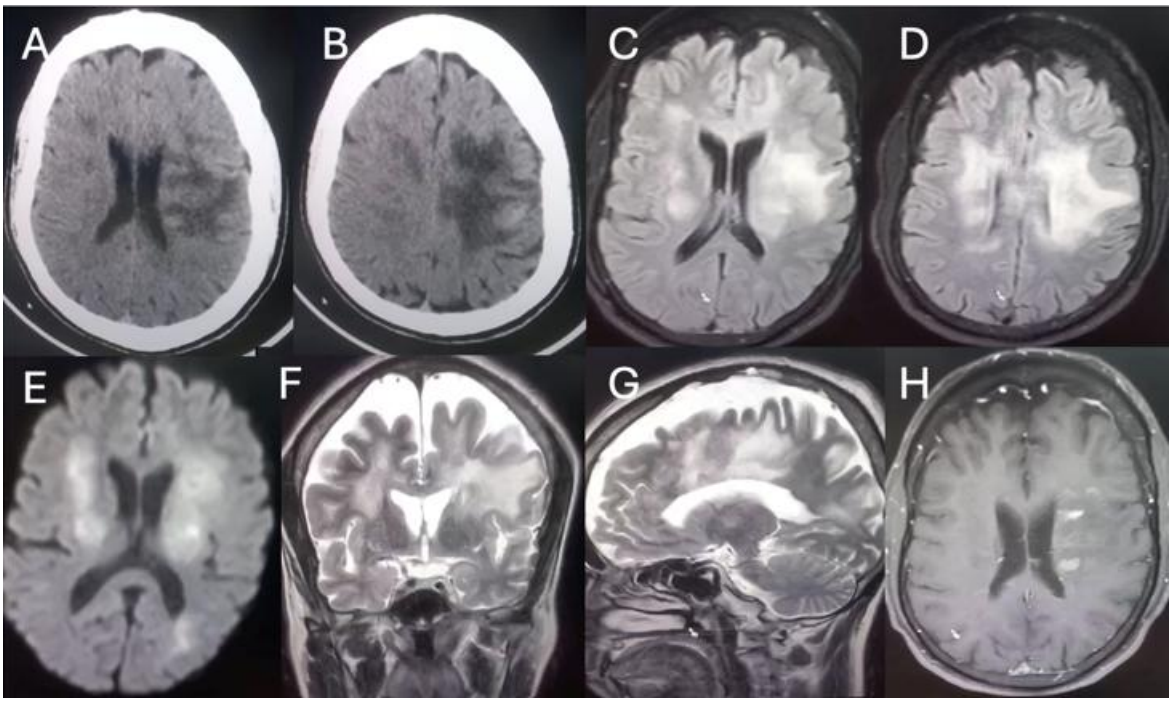

Figure 1: Brain imaging (first presentation): A, B- Plain CT brain showing subcortical, periventricular hypodensities bilateral (left>right) frontoparietal white matter with areas of hyperdensity within. C, D- FLAIR imaging showing subcortical and

periventricular frontoparietal white matter hyperintensities left>right, with U-fiber involvement. E- DWI imaging showing diffusion restriction of the lesions. H-T1C MRI showing areas of patchy contrast enhancement

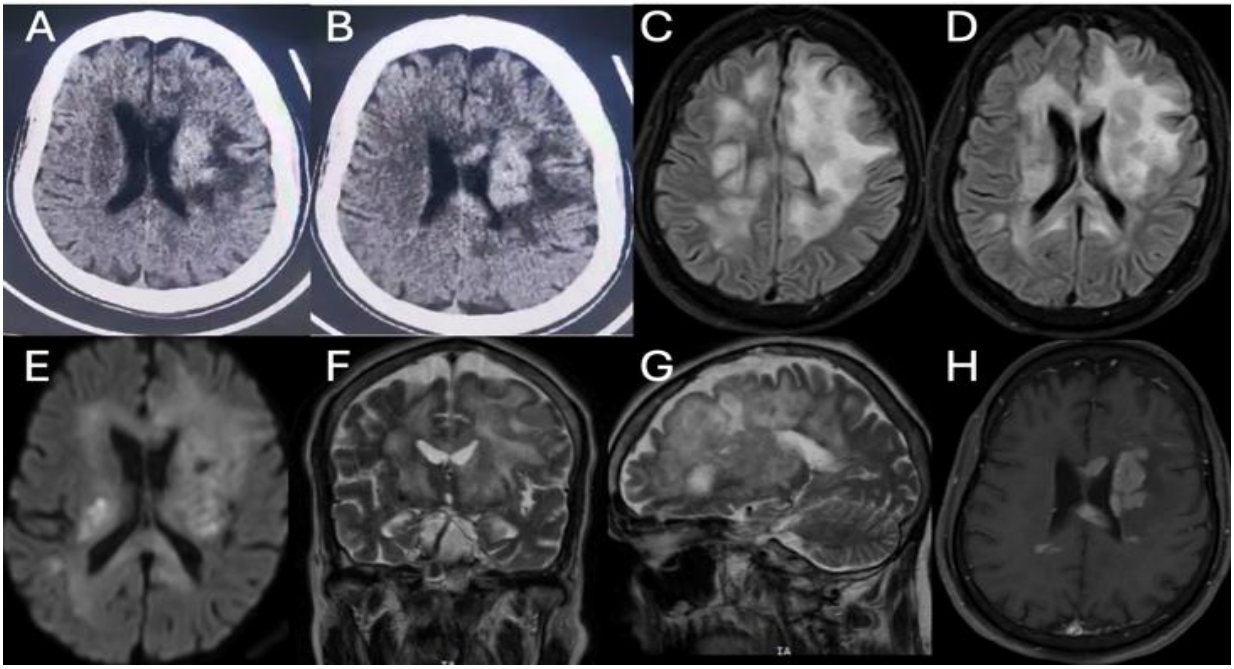

Figure 2: Brain imaging (second presentation) – A, B- Plain CT brain showing periventricular white matter hyperdensity involving corpus callosum (genu and splenium) with surrounding hypodense edema. C, D – FLAIR imaging showing hyperintensities in the frontoparietal periventricular>subcortical white matter including splenium and genu of corpus callosum with areas of hypointensities within. Note that the lesions have increased compared to initial imaging. E- diffusion restriction of the lesions. H) intense contrast enhancement of the same

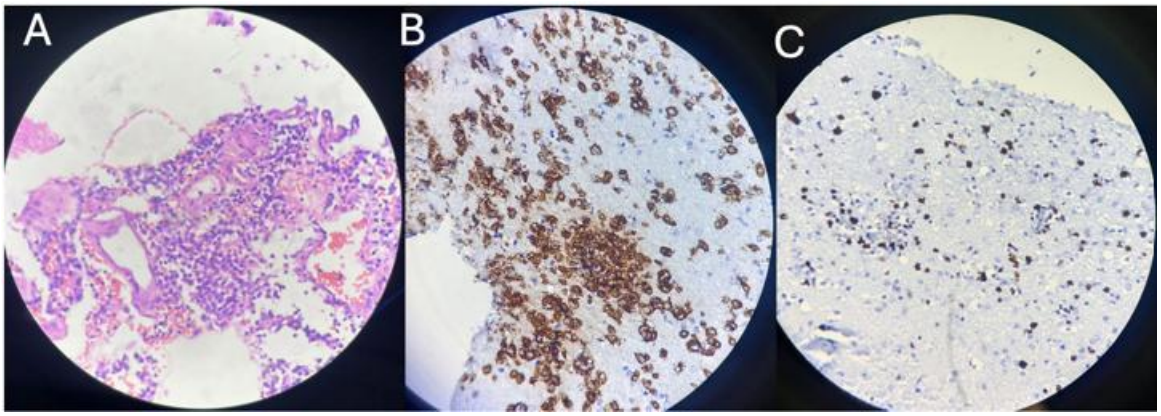

Figure 3: A) Biopsy of the lesion on HPE showing sheets of immature lymphocytes. B, C) On immunohistochemistry, cells were CD20+, with focally high Ki62 activity suggestive of Non-Hodgkin Lymphoma – Diffuse Large B-Cell Lymphoma subtype

3. Discussion

Clinical Presentation

Primary CNS lymphoma (PCNSL) is a rare and aggressive form of extranodal non-Hodgkin lymphoma confined to the central nervous system. It accounts for approximately 1–2% of all primary CNS malignancies and is most commonly diagnosed in immunocompetent individuals aged 60 years and older. The clinical manifestations of PCNSL are diverse and can mimic other neurological disorders, often leading to diagnostic challenges[1].

Patients commonly present with focal neurological deficits, including hemiparesis, aphasia, and visual disturbances, which occur in approximately 70% of cases. Neuropsychiatric symptoms, such as personality changes, confusion, and cognitive decline, are observed in about 43% of patients. Headache, nausea, vomiting, and signs of elevated intracranial pressure are present in approximately 33% of cases. Seizures are less common, occurring in about 14% of patients. In this case, the patient exhibited right focal seizures, altered sensorium, spasticity in all limbs, and right-sided weakness, which are consistent with the typical presentation of PCNSL.

The progression of symptoms over a four-month period underscores the aggressive nature of the disease. This case highlights the importance of considering PCNSL in the differential diagnosis of patients presenting with new-onset

neurological symptoms, especially when imaging findings are atypical for common stroke etiologies.

Imaging Findings

MRI is the gold standard for imaging in PCNSL. Lesions are typically located in the supratentorial region, with a predilection for the periventricular white matter, basal ganglia, and corpus callosum. On MRI, PCNSL lesions are often isointense to hypointense on T1-weighted images and isointense to hyperintense on T2-weighted images. They usually demonstrate homogeneous enhancement after gadolinium administration, with surrounding edema and restricted diffusion on diffusion-weighted imaging[2].

In this patient, repeat MRI revealed an increase in the size and number of lesions, which is characteristic of PCNSL progression. The absence of significant necrosis or hemorrhage further supports the diagnosis of PCNSL, as these features are less common compared to other CNS pathologies.

The imaging findings in this case were initially suggestive of a stroke, which is consistent with reports of PCNSL presenting as a stroke mimic. For instance, a 75-year-old patient with PCNSL presented with left hemiparesis and a hyperdense lesion on CT imaging, initially diagnosed as a stroke. However, further imaging and biopsy confirmed the diagnosis of PCNSL. Similarly, another case reported a 56-year-old man with acute left-sided weakness and imaging findings suggestive of a stroke, later diagnosed with PCNSL following biopsy[3].

Diagnostic Approach

The diagnosis of PCNSL requires a high level of suspicion and is confirmed through histopathological examination. Stereotactic brain biopsy is the preferred method for obtaining tissue samples, as it provides a definitive diagnosis in over 90% of cases. It is essential to avoid corticosteroid administration prior to biopsy, as steroids can alter the histopathological appearance and delay diagnosis.

In this case, brain biopsy confirmed the diagnosis of Diffuse Large B-Cell Lymphoma (DLBCL), a subtype of PCNSL. Additional imaging, including contrast-enhanced whole-spine MRI and fluorodeoxyglucose positron emission tomography (FDG-PET) scanning, was performed to exclude systemic involvement. These studies are crucial, as systemic lymphoma with secondary CNS involvement can mimic primary CNS lymphoma.

Differential Diagnosis

The differential diagnosis for PCNSL includes high-grade gliomas, cerebral abscesses, toxoplasmosis, and progressive multifocal leukoencephalopathy. Imaging characteristics, such as homogeneous contrast enhancement and restricted diffusion, can help differentiate PCNSL from these conditions. However, histopathological examination remains the gold standard for definitive diagnosis.

In this case, the initial presentation and imaging findings were suggestive of a stroke, which is consistent with reports of PCNSL presenting as a stroke mimic. For instance, a 75-year-old patient with PCNSL presented with left hemiparesis and a hyperdense lesion on CT imaging, initially diagnosed as a stroke. Similarly, another case reported a 56-year-old man with acute left-sided weakness and imaging findings suggestive of a stroke, later diagnosed with PCNSL following biopsy[3].

Management and Prognosis

Treatment of PCNSL typically involves high-dose methotrexate-based chemotherapy, often in combination with other agents such as cytarabine. Whole-brain radiation therapy may be considered in certain cases, particularly for patients who are not candidates for chemotherapy or those with relapsed disease. The prognosis for PCNSL has improved with advances in treatment, but the disease remains associated with high morbidity and mortality.

In this patient, the initiation of chemotherapy was planned following the confirmation of diagnosis. The patient's response to treatment and overall prognosis will depend on factors such as age, performance status, and the extent of disease at diagnosis.

4. Conclusion

PCNSL is a rare and aggressive malignancy that requires a high index of suspicion for diagnosis. Early recognition and appropriate management are crucial for improving patient outcomes. This case highlights the importance of considering PCNSL in the differential diagnosis of patients presenting with new-onset neurological symptoms and the role of advanced imaging and histopathological examination in confirming the diagnosis.

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