

Case Report

A Rare Histopathological Phenotype of Primary Central Nervous System Lymphoma

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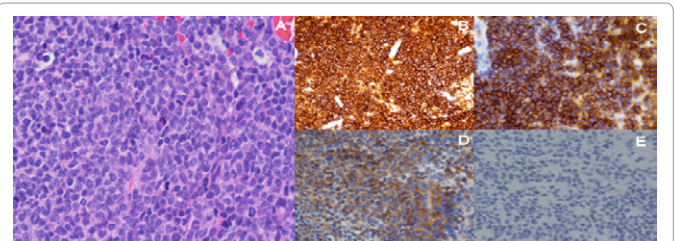
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Case Summary

A 42-year old male presented with a new generalised severe dull aching headache, vomiting, and bilateral papilloedema. MRI brain revealed a mass lesion cuffing the lateral ventricles (Figure 1). He developed pyramidal tract signs, cerebellar dysfunction, aphasia and seizures. Laboratory testing was negative for human immunodeficiency virus serology and bone marrow biopsy was unremarkable. Brain biopsy showed a blastichematolymphoid malignancy (Figure 2). Imaging for evidence of systemic disease outside the central nervous system was negative. Intravenous corticosteroids were administered but the patient deteriorated and his family opted for palliation. He died four weeks after his initial presentation.

Discussion

Primary CNS Lymphoma (PCL) is an aggressive disease, most commonly occurring in immunosuppressed individuals, particularly in the context of HIV infection. PCL can involve the brain, meninges, eyes or spinal cord [1,2]. Brain involvement, known as primary cerebral lymphoma, may manifest as solitary or multiple intracranial lesions, or as diffuse leptomeningeal or periventricular lesions. Periventricular lesions in the brain present with heterogeneous features including headache, focal neurological deficits, personality changes, raised intracranial pressure and seizures. Approximately 98% of PCL are CD20+ B-cell lymphomas, usually of the diffuse large-cell, immunoblastic, or lymphoblastic subtypes [3]. This case is unusual as it occurred in a seemingly immunocompetent individual and followed an aggressive clinical course. Furthermore, biopsy (Figure 2) revealed a



(A) Monomorphous blastichematolymphoid infiltrate (Haematoxylin and eosin 400x). Immunohistochemistry demonstrates immunoreactivity of tumour cells for (B) CD43, (C) CD56 and (D) CD4. (E) CD123 is negative.

Figure 2: Brain biopsy.

rare non-B cell lineage with an unusual immunophenotype suggesting a blastic hematolymphoid malignancy, which was not lineage specific and may be seen in T cell lymphoma, Natural Killer cell lymphoma, or myeloid sarcoma.

Some cases of PCL respond to methotrexate based chemotherapy and radiotherapy, however, treatment is rarely curative. This case demonstrates a characteristic presentation and MRI features of primary CNS lymphoma. However, it is unusual due to atypical immunohistology, (non-B cell origin), absence of apparent immunodeficiency and rapid deterioration leading to death within one month.

Author Contributions

MSB conceptualised the report, ZI drafted the report and prepared the manuscript, QL prepared the pathology slides and revised the draft, SB supervised the imaging and revised the draft.

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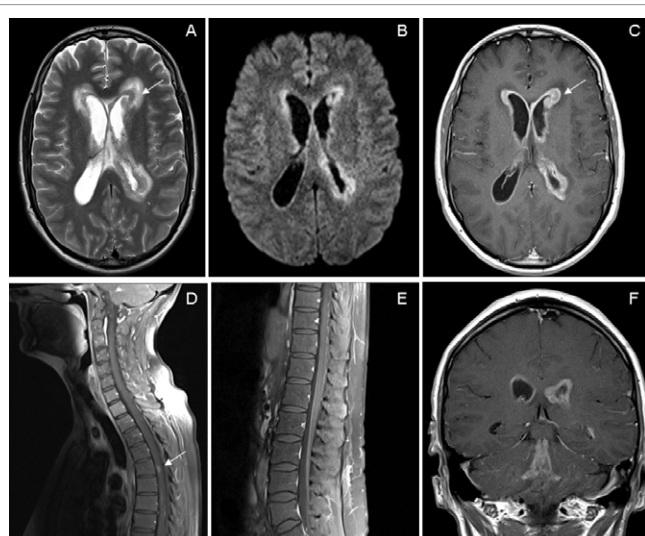
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T2 weighted axial image demonstrates periventricular mass (A, arrow) with restricted diffusion (B). Periventricular enhancement and leptomeningeal enhancement seen in the brain and spinal cord (C-F).

Figure 1: MRI brain and spine with contrast.