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Original Research Paper

Neurosurgery

LYMPHOMATOSIS CEREBRI RARE CASE SCENARIO

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ABSTRACT Central nervous system lymphoma is an infrequent variant of Lymphomatosis. It is an infrequent type of CNS lymphoma characterized by lymphoma cells diffusely infiltrating the brain parenchyma without forming a mass or distorting the cerebral architecture. It is an unusual form of Non-Hodgkin lymphoma, majority being diffuse large B-cell lymphomas. I am presenting a case about 50-year woman with no vascular risk factor, who had a stealthy progressive dementia & right-side weakness Since July 2018.

KEYWORDS : Lymphomatosis cerebri, Infiltration, White matter dementia

Introduction-:

Primary central nervous system lymphoma (PCNSL) most often presents like a solitary, isolated lesion greatly less well recognized is the diffuse intrusion of the cerebral white matter by individual lymphoma cells without formation of a discernable cohesive tumor mass, which is termed Lymphomatosis cerebri (LC), on cerebral magnetic resonance imaging (MRI), it appears as diffuse leukoencephalopathy without contrast enhancement, often causes diagnostic confusion, with suspected diagnoses ranging from Binswanger's disease to encephalomyelitis. Needle biopsy tends to result in acquisition of a very small specimen, leading to pathological misdiagnosis.

We presenting a case about woman who had Headache on /off since last 2 years and weakness in Right hand grip due to LC. Although her neuroimaging features mimicked those of other leukoencephalopathic disorders, neoplastic disorders such as LC should be included in the list of differential diagnoses.

Case report

This 50-year-old woman with no vascular risk factor, who had a stealthy progressive dementia & right-side weakness Since July 2018. At initial presentation, she was alert and complained of general fatigue, dizziness, and Headache on /off since late 2015, and Rt. hand grip weakness and slurring of speech since 2016. Developed anxiety and insomnia Relatives noted some personality change in the form of emotional liability, they consulted local neurophysician. Investigated with MRI Brain done in 2016 which revealed evidence of slight hyperintensity on T1, iso to hyper intense lesion with perilesional edema in b/l cerebellar parenchyma on T2 flair And contrast there was a patchy enhancement. (Figure 1,2). She was admitted in our hospital. On examination, she was apathetic and obeyed only simple commands. She was unable to stand. There was slight leukocytosis and a 0.15-mg upgrading of C-reactive protein (CRP). After serum examinations, with collagen vascular profile, human immunodeficiency virus, and treponemal serological report, were all negative. Plasma soluble interleukin-2 (IL-2) receptor level was elevated to 566 U/ml (normal range 145–518 U/ml). Cerebrospinal fluid (CSF) analysis showed a protein level of 31 mg/dl (normal 30–45 mg/dl) and a normal increased cell count. On cytology, few lymphocytes but no obvious atypical cells were found. No bacteria were detected in the CSF.

Patient underwent stereotactic biopsy from lt periventricular area. Histopathological examination reveled showed mainly lymphocytes the vessels and also few singly scattered cells in the glial parenchyma. HE 20X : showed mainly lymphocytes surrounding the vessels and also few singly scattered cells in the glial parenchyma.(Figure-3,4)

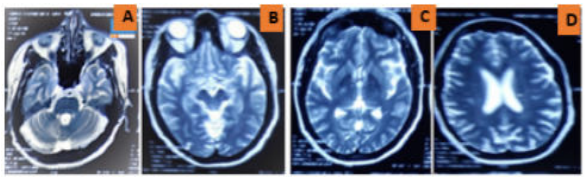


Fig. 1 Computed tomography images taken at initial presentation were considered to be normal

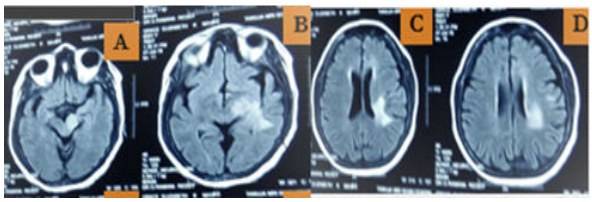
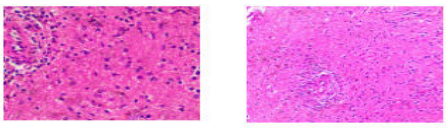


Figure 2- T2 flair sequnces showing left mid brain and left periventricular edema.

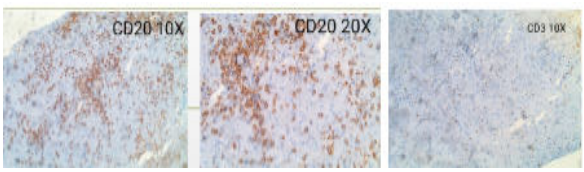
Pathological findings: -

The histopathology examination reveal the following finding.

(Figure 3)



HE 20X : showed mainly lymphocytes the vessels and also few singly scattered cells in the glial parenchyma.



(Figure 4) -

Almost all the tumor cells were CD20 POSITVE. CD3 10X also showed positivity in scattered tumor cells. Mib1 was also high (40%) in the tumor cells. BCL6 and BCL2 are positive and negative for EBV.

Discussion: -

PCNSL accounts for 3% of primary central nervous system tumours. It typically presents as multifocal T2 hyperintense lesions that both enhance after gadolinium administration and demonstrate mass effect.[1,4] LC is a rare type of CNS lymphoma characterized by lymphoma cells diffusely infiltrating the brain parenchyma without forming a mass or distorting the cerebral architecture. It is an unusual form of Non Hodgkins lymphoma, majority being diffuse large B-cell lymphomas. MR imaging typically reveals diffuse white matter disease variably involving bilateral cerebral hemispheres, periventricular region, basal ganglia, thalami, or the brainstem.

A case study in 1999 coined the term 'lymphomatosis cerebri' to describe an exceedingly rare variant of PCNSL characterized by diffuse parenchymal infiltration of lymphomatous cells.[5]

The most common presenting symptoms are cognitive decline (59.5%), gait disturbances (54.8%) and behavioral changes (50%). But the clinical presentation of these patients was easily mistaken for other, more common, conditions such as infectious, inflammatory, vascular, toxic, or neurodegenerative etiologies that can cause white-matter injury and bilateral hemispheric lesions on MRI.[2,3]. Conventional MRI without contrast enhancement showed extensive, diffuse hyperintense lesions involving bilateral cerebral hemispheres on both T2-weighted FLAIR sequences. Most patients show patchy contrast enhancement. MRS consistently presented a pattern of marked decrease of NAA/Cr, increase of Cho/Cr, increased Lip/Cr and Lac/Cr. Most are hyperintensity on DWI and hyperintensity on ADC. The most common regions involved were the white matter of both hemispheres, in the frontal and deep periventricular regions including corpus callosum, and the lesions extended into the gray matter such as basal ganglia, thalamus and cortex.[1,4]

Classification lesions by distribution
1. deep,
2. lobar,
3. infratentorial categories

PCNSL having response rate of 90% and median survival of 60 month with high dose methotrexate-based chemotherapy. But prognosis of Lymphomatosis cerebri is worse than of primary lymphoma.

Conclusion: -

LC variant of PCNSL is a rare entity, and it should be added to the differential diagnosis of cognitive decline, with unusual MRI findings. Prompt recognition of these imaging finding may lead to early diagnosis of LC and brain biopsy with improved prognosis.

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