Primary Central Nervous System Lymphoma in an immunocompetent patient: An Unusual Case of Prolonged Response to Steroids

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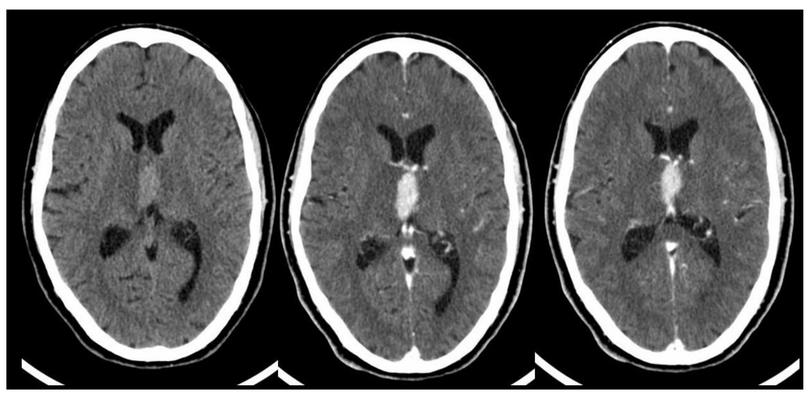
Introduction

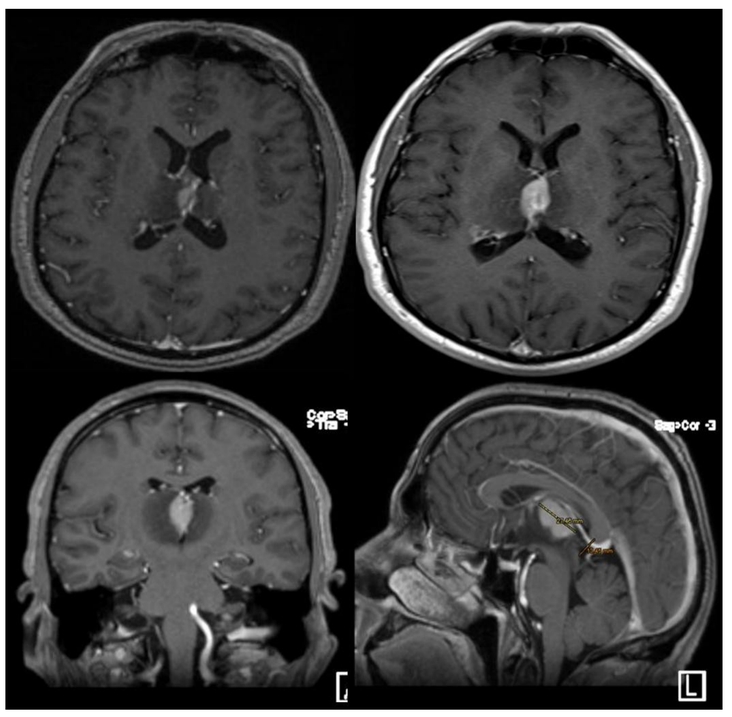
PCNSL causes approximately 3%-4% of all primary brain tumors. PCNSL is defined as lymphoma in the central nervous System (CNS) without primary tumor elsewhere. The incidence rates of PCNSL are increasing among immunocompetent patients.
Early diagnosis of CNS lymphoma is crucial for proper management in both immunocompetent and immunocompromised individuals and is more likely if a tumor is observed on imaging.

Case Report

A 63-year-old man was referred to our emergency room because of nausea with associated acute confusional state. Neurological examination revealed slow response, postural instability without rigidity or tremor in any of the four extremities, and normal sensation.
CT acquisition showing, between I and II ventricles and part od III ventricles, a mild hyperdense of 2 x 1 cm complex lobulated mass in the right frontal region associated with oedema.
MRI images showing inhomogeneous hyperintensity of 27 x 13 x 13 mm in the thalamic, periaqueductal, crura cerebri and colliculi midbrain; the lesion infiltrated casting subependymal region of the third ventricle and was associated with ovalar hyperintensities and perilesional oedema.
Laboratory tests were within normal limits.

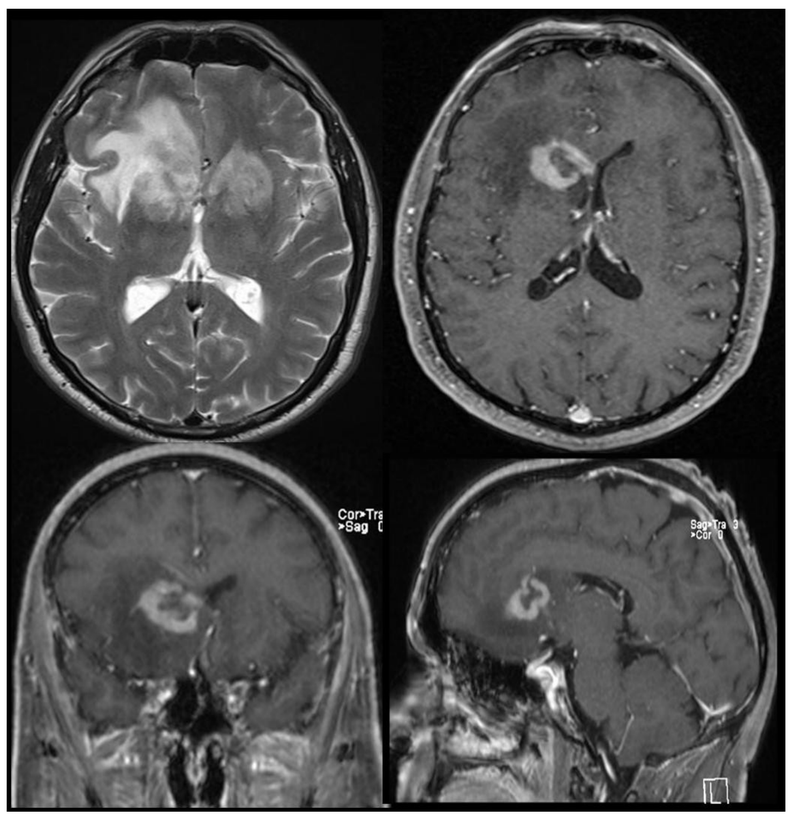


The patient was extensively investigated. Primary central nervous system lymphoma (PCNSL) was then suspected. HIV serum test was negative. The patient refused biopsy, then he was treated with steroids and complete clinical and radiological remission was achieved.
In the following two years, without any treatment, he did not show any pathological evolution and the MRI follow-up showed no recurrence of disease for two years when he developed an acute confusional state.
CT and RMN highlighted the presence of frontal right lobe lesions pf 33 x 26 x 25 mm associated with ovalar hyperintensities and perilesional oedema.
Cerebrospinal fluid examination demonstrated white cell count 1/□L, glucose 63.7 mg/dL (blood glucose 110mg/dL), proteins 67.7 mg/dL (n.v: 15–45 mg/dL), lactate 1.6 mg/dL (n.v: 0.6–2.2 mg/dL), and IgG index 0,90 (n.v: <0,5).
The CSF cytology examination showed lymphoma cells.
Stereotactic biopsy of the right frontal lobe was performed showing medium-sized lymphoid cells with a perivascular pattern; histological diagnosis of diffuse large B-cell lymphoma was obtained.



Conclusion

Primary CNS lymphoma, particularly when affecting young and immunocompetent subjects might present diagnostic difficulties at onset.
Treatment with steroids can lead to a regression of lesions in up to 40 % of patients, thus deferring the definite histological diagnosis.
Our case shows some peculiar features: a long disease history (overall, 2 years) with an unusual and protracted response to steroids.



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